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Axillary arch: Clinical significance in breast cancer patients

Deep Lamichhane, Sanjit Kumar Agrawal, Sumit Mukhopadhyay, Rosina Ahmed

ABSTRACT

Introduction: Langer’s axillary arch is a muscular slip extending from the anterior border of latissimus dorsi muscle to tendons, muscles or fascia around the superior part of humerus, lying anterior to the neurovascular bundle. It is the best-known anatomic variant of the axilla, with definite clinical and surgical implications. Case Report: A 55-year-old female presented with a 3x3 cm carcinoma in the superomedial quadrant of the right breast, with no palpable regional lymph nodes. She underwent breast conservation surgery with axillary nodal clearance. During axillary dissection, an unusual muscle slip was identified crossing the axilla, connecting the anterior border of latissimus dorsi to the posterior surface of pectoralis major, anterior to the axillary artery, vein and brachial plexus. Conclusion: Preoperative knowledge is essential to identify such unusual anatomy and to appropriately tackle it to avoid surgical complications and adequate axillary lymph node clearance.

Keywords: Axillary arch, Axillary lymph node dissection, Breast carcinoma

How to cite this article


Article ID: Z01201712CR10857DL

doi: 10.5348/ijcri-2017118-CR-10857

INTRODUCTION

Anatomical variations in the axilla are of great relevance during all surgical procedures performed in this region. This includes minimal axillary procedures such as sentinel lymph node biopsy, where precision in recognizing anatomical landmarks is essential, axillary dissection in case of malignancy and also a number of larger reconstructive procedures and vascular bypass operations [1].

Various accessory muscular slips have been described in the axilla. The best known variant, the axillary arch (AA), is a muscular or fibromuscular slip of variable dimensions extending from latissimus dorsi muscle, which crosses over the neurovascular structures to join the under surface of the tendon of pectoralis major, coracobrachialis or the fascia over the biceps brachii [2, 3].

The presence of the axillary arch has important clinical implications. This variation occurs unilaterally in about 7% of the population, more commonly in females than in males [3].
CASE REPORT

A 55-year-old female presented with a right breast lump of six months duration. Clinical examination revealed a 3x3 cm lump in the superomedial quadrant of the right breast, with no palpable regional lymph nodes. Mammography showed a BIRADS 5 lesion at 1 o’clock, with suspicious axillary lymph nodes on ultrasonography. Core biopsy showed invasive ductal carcinoma (IDC) Grade III, estrogen and progesterone receptor negative, HER-2 positive. She underwent breast conservation surgery with axillary nodal clearance.

During axillary dissection, an unusual muscle slip was identified, which crossed the axilla, connecting the anterior border of latissimus dorsi to the posterior surface of pectoralis major, anterior to the axillary artery, vein and brachial plexus. It was measured 7 cm in length and 1.5 cm in width (Figure 1). The lymph nodes medial and deep to the arch were successfully dissected and the arch was left undisturbed. The procedure and recovery were uneventful.

Pathological assessment confirmed IDC 3 cm, grade III with two lymph nodes positive for malignancy out of 36 dissected. Metastatic staging was normal, and she received adjuvant chemotherapy, trastuzumab and radiotherapy. Postoperative imaging showed a bilateral axillary arch along with postoperative changes in the right breast and right axilla (Figure 2).

DISCUSSION

Langer’s axillary arch, a variation in axillary musculature, was first identified by Ramsay in 1795 and later described in detail by Langer in 1846 [2]. Testu’s classification (1884) described the complete axillary arch, extending between the latissimus dorsi muscle and the tendon of the pectoralis major near its insertion on the humerus, and the Incomplete Arch, extending from the latissimus dorsi muscle to the axillary fascia, biceps brachii muscle, coracobrachialis, inferior edge of pectoralis minor muscle or the coracoid process [2]. In an additional classification, two forms of arch-shaped variations have been described, muscular (type I) and tendinous (type II), with different subtypes based on nerve supply and site of attachment. The term clinical axillary arch was introduced by Jelev, who described it as a site of entrapment for nerves and vessels, and also classified the condition into superficial and deep [2]. Superficial arches cross in front of the vessels and nerves and primarily present with features of intermittent obstruction of veins. Deep arches occur on the posterior or lateral walls of the axilla and cross-only parts of the neurovascular bundle, axillary or radial nerves [2, 4]. The patient we described had a complete, superficial, muscular axillary arch.

The prevalence of this variation appears to be higher in dissected cadavers than found during surgery in the axilla. The prevalence of axillary arch in cadaveric dissection in Japanese, Turkish and Bulgarian populations is 9.1%, 1.9% and 3.6% respectively [5–7]. On the other hand, it has been recognized in only 0.25% of patients during axillary surgical procedures [8]. The difference in prevalence in anatomical and surgical reports may be due to a failure to identify or report the variation when observed during surgery, whereas the specific aim of cadaveric studies is to identify anatomical anomalies [8].

Clinically, axillary arch may be palpable during physical examination as an axillary mass and may be confused with lymphadenopathy or soft tissue tumor. Most patients, similarly to the one described here, are asymptomatic. However, entrapment of the axillary neurovascular bundle by an axillary arch during arm movements has been described, and may cause circulatory insufficiency, chronic pain or paraesthesia [8]. The simple division of the arch is curative in such situations [9].

During axillary surgery, it may be mistaken for the lateral margin of the latissimus dorsi muscle, leading
dissection to be extended superior to the axillary vein, with the risk of injury to the axillary artery and brachial plexus [10]. It can also pose difficulty during sentinel node biopsy as it stretches in the hyper abducted position, shifting the nodes higher [11]. During axillary clearance for breast cancer some groups of axillary nodes such as lateral nodes, may be concealed under the axillary arch, and may be missed during routine dissection [10]. This may lead to inaccurate staging, which would negatively affect adjuvant systemic therapy decisions and also to an increased chance of local recurrence [10]. The possible presence of accessory muscle slips should also be kept in mind while draining axillary abscesses and while constructing latissimus dorsi flaps.

Some authors suggest routine division of axillary arch at the level of the axillary vein to be able to identify anatomical landmarks and to facilitate dissection of lymph nodes. Division might also reduce the possibility of postoperative axillary vein compression and associated lymphedema [10, 11]. The possibility of precipitating lymphedema is higher in patients having latissimus dorsi flaps for breast reconstruction and in this situation it is advisable that the axillary arch should be divided [11]. In this case, we were able to dissect nodes beneath the arch, without the need to divide it and there were no specific postoperative problems.

CONCLUSION

The axillary arch has both clinical and surgical implications, and surgeons must remember its possible presence and must be cautious during axillary dissection. It may be a reason for confusion during routine axillary surgery and can both affect procedural safety and adjuvant treatment decisions.

**********

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Deep Lamichhane – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Sanjit Kumar Agrawal – Substantial contributions to conception and design, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Sumit Mukhopadhyay – Acquisition of data, Drafting the article, Final approval of the version to be published
Rosina Ahmed – Acquisition of data, Drafting the article, Final approval of the version to be published

Guarantor of Submission

The corresponding author is the guarantor of submission.

Source of Support

None

Conflict of Interest

Authors declare no conflict of interest.

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REFERENCES

Walking the thin line between malignancy and infectious disease: A case report

Catarina Couto, Sofia Fraga

ABSTRACT

Introduction: Children infected by human immunodeficiency virus have a higher incidence of malignancy than non-infected children. Case Report: A healthy 13-year-old boy presented with a month long history of a swelling of the right parotid gland and cervical region. On ultrasound, multiple right cervical and intraparotid lymph nodes were noted. A fine-needle lymph node aspiration cytology and flow cytometry excluded a lymphoproliferative disorder, but serologic testing revealed the patient had a human immunodeficiency virus infection. Tuberculous lymphadenitis was considered and the patient was started on antitubercular drugs after a lymph node biopsy was performed. The biopsy eventually revealed a non-Hodgkin lymphoma. Conclusion: Overlapping symptoms at presentation can complicate differential diagnosis of human immunodeficiency virus infection and malignancy.

Keywords: Diffuse large B cell lymphoma, Human immunodeficiency virus, Malignancy

INTRODUCTION

Late diagnosis of human immunodeficiency virus (HIV) infection in children can be associated with a more severe presentation because HIV-related immunosuppression increases the incidence of an acquired immune deficiency syndrome (AIDS).

HIV-infected children have a higher incidence of malignancy than non-infected children, especially if they are not receiving treatment with highly active antiretroviral therapy [1–3]. AIDS malignancies include Kaposi’s sarcoma, non-Hodgkin lymphoma and invasive cervical cancer.

CASE REPORT

An apparently healthy 13-year-old boy presented to the A&E department of his local hospital with a month long history of a painless swelling of the right parotid gland which extended to the right cervical region, with no signs of skin infection, fever, weight loss or any other constitutional symptoms. The patient denied any current medication, animal exposure, recent traveling or ingestion of unpasteurized dairy products or undercooked meats. His personal and family history, as were given at...
the time, were uneventful. An ultrasound was performed which showed a nodular structure within the parotid gland of approximately 2.4 cm in diameter, described as a probable adenopathy. The ultrasound also identified multiple small right laterocervical lymphadenopathies. To better ascertain the etiology of the mass, a magnetic resonance imaging (MRI) scan revealed an increase in the size of the parotid gland and a nodular lesion within the gland with central areas of necrosis. He was started on oral cefradine, a first generation cephalosporin, which he continued for 15 days with no signs of improvement. At this time he was referred to a pediatric oncology center to exclude a possible lymphoma.

On admission to the center, he had a fine-needle aspiration cytology and flow cytometry which were both negative for neoplastic cells. He also had an extensive blood workup performed (Table 1) which showed a thrombocytopenia, a slightly increased erythrocyte sedimentation rate and lactate dehydrogenase and a hypergammaglobulinemia. Multiple viral serologies were also performed, including HIV p24 antigen, which was positive. A confirmatory test for human immunodeficiency virus (HIV) infection was performed and was also positive. We then learned that both parents had been diagnosed with HIV infection when the child was four-year-old and no testing had been done on the boy at the time. After HIV diagnosis, the child was referred to our hospital where he would have HIV follow-up.

On admission, the patient was a healthy looking boy with a body mass index of 21.3 kg/m², he had a swelling of the right parotid gland and cervical region with a slight erythema (Figure 1), palpable right cervical lymphadenopathies and no hepatosplenomegaly. The rest of the examination was normal. His initial viral load was 96,730 cp/mL and his CD4 count was 189 cells/μL (Table 2). He was tested for antiretroviral resistance, which was negative, and had a tuberculin skin test, which was negative. He was started on Atripla® (emtricitabin e+tenofovir+efavirenz) and cotrimoxazole. He was also started on amoxicillin and clavulanic acid, which he maintained for 15 days, because he had some signs of skin infection. After three weeks of antibiotics, the swelling and erythema of the right parotid gland continued to evolve (Figure 2) and he was switched to clindamycin for another 15 days with no response (Figure 3).

At this point tuberculous lymphadenitis was suspected, the patient had a chest X-ray, which was normal, and a cervical computed tomography (CT) scan which showed a right cervical conglomerate of lymphadenopathies with areas of necrotic tissue, multiple small left cervical and bilateral supraclavicular lymphadenopathies (Figure 4). A lymph node aspiration was performed but came back as having insufficient material so the patient had a lymph node biopsy. Due to the clinical suspicion of tuberculous lymphadenitis and despite having a negative acid-fast bacilli stain, he was started on isoniazid, rifampicin, pyrazinamide and ethambutol but there was no improvement (Figure 5).

We then learned the biopsy revealed a diffuse large B-cell lymphoma. He was started on chemotherapy. The culture for *Mycobacterium tuberculosis* was negative and he stopped the tuberculous drugs. The final diagnosis was diffuse large B cell lymphoma, an AIDS-defining disease in a perinatally HIV-infected patient.

The patient completed three months of chemotherapy, without any major complications, and at sixth month follow-up postchemotherapy, he is in remission. He is still on the same HAART regimen and now has an undetectable viral load. Immune reconstitution has been slow and his CD4 count is still under 200 cells/μL (Table 2).

**DISCUSSION**

Mother-to-child transmission (MTCT) is the principal mode of HIV infection in children (92% of HIV-infected children in Portugal have acquired it perinatally) [4]. Since early 2004, nationwide HIV screening during pregnancy (1st and 3rd trimester) has been mandatory in Portugal. Regularly testing for HIV infection in pregnant women is one of the most important preventive measures of MTCT of HIV because it allows for early treatment with
Table 1: Blood workup on admission to the pediatric oncology center

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<tr>
<td>Hemoglobin</td>
<td>13.3 g/dL</td>
<td>13.0–17.0 g/dL</td>
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<tr>
<td>Platelets</td>
<td>132x10^9/L</td>
<td>150–400x10^9/L</td>
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<tr>
<td>Leukocytes</td>
<td>4.640x10^9/L</td>
<td>4.000–10,000</td>
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<tr>
<td>Neutrophils</td>
<td>49% (2.250x10^9/L)</td>
<td>2,000–7,000x10^9/L</td>
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<tr>
<td>Lymphocytes</td>
<td>41% (1,900x10^9/L)</td>
<td>1,000–4,000x10^9/L</td>
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<tr>
<td>Erythrocyte Sedimentation Rate</td>
<td>37 sec</td>
<td>&lt; 20 sec</td>
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<tr>
<td>Lactate dehydrogenase</td>
<td>323 UI/L</td>
<td>125–220 UI/L</td>
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<tr>
<td>Uric acid</td>
<td>5.0 mg/dL</td>
<td>3.5–7.2 mg/dL</td>
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<tr>
<td>Aspartate aminotransferase</td>
<td>52 UI/L</td>
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<td>Alanine aminotransferase</td>
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<td>Alkaline phosphatase</td>
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<td>12–64 UI/L</td>
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<td>Total bilirubin</td>
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<td>0.20–1.20 mg/dL</td>
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<tr>
<td>Glucose</td>
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<td>Blood urea nitrogen</td>
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<td>Creatinine</td>
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<td>Immunoglobulin A</td>
<td>486 mg/dL</td>
<td>63–484 mg/dL</td>
</tr>
<tr>
<td>Immunoglobulin G</td>
<td>2192 mg/dL</td>
<td>540–1,822 mg/dL</td>
</tr>
<tr>
<td>Immunoglobulin M</td>
<td>667 mg/dL</td>
<td>22–240 mg/dL</td>
</tr>
<tr>
<td>B Hepatitis Virus</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Surface antigen (HBsAg)</td>
<td>Negative</td>
<td></td>
</tr>
<tr>
<td>Surface antibody (anti-HBs)</td>
<td>Negative</td>
<td></td>
</tr>
<tr>
<td>Core antibody (anti-HBc)</td>
<td>Negative</td>
<td></td>
</tr>
<tr>
<td>C Hepatitis Virus (anti-HCV)</td>
<td>Negative</td>
<td></td>
</tr>
<tr>
<td>Varicella Zoster Virus</td>
<td></td>
<td></td>
</tr>
<tr>
<td>IgM</td>
<td>Negative</td>
<td></td>
</tr>
<tr>
<td>IgG</td>
<td>Negative</td>
<td></td>
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<tr>
<td>Citomegalovirus</td>
<td></td>
<td></td>
</tr>
<tr>
<td>IgM</td>
<td>Negative</td>
<td></td>
</tr>
<tr>
<td>IgG</td>
<td>Positive</td>
<td></td>
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<tr>
<td>Epstein-Barr Virus</td>
<td></td>
<td></td>
</tr>
<tr>
<td>VCA IgM</td>
<td>Negative</td>
<td></td>
</tr>
<tr>
<td>VCA IgG</td>
<td>Positive</td>
<td></td>
</tr>
<tr>
<td>EBNA IgG</td>
<td>Positive</td>
<td></td>
</tr>
<tr>
<td>HIV antigen p24</td>
<td>Positive</td>
<td></td>
</tr>
</tbody>
</table>
highly active antiretroviral therapy (HAART), which will decrease the viral load and improve immune function and thus reduce the risk of transmission to the fetus and newborn. In Portugal, around 30% of HIV-infected mothers are diagnosed during pregnancy [4]. Despite such a high rate of maternal diagnosis during pregnancy, which is probably explained by the considerable number of women from foreign countries who come to Portugal during pregnancy (44% of HIV-infected mothers are of non-Portuguese nationality) [4], transmission rate has been consistently under 2% since 2005, which highlights the importance of testing during pregnancy.

Some cases of mother-to-child transmission will keep evading early detection, like in the case reported. There are two reasons for late diagnosis of perinatally acquired HIV infection in a child: either the mother is in the window period in the 3rd trimester and, despite being

Table 2: Viral load and immunological testing over follow-up time

<table>
<thead>
<tr>
<th></th>
<th>On HIV diagnosis</th>
<th>1 month later</th>
<th>2 months</th>
<th>4 months (on chemotherapy)</th>
<th>6 months</th>
<th>9 months</th>
</tr>
</thead>
<tbody>
<tr>
<td>Viral load (cp/ml)</td>
<td>96,730</td>
<td>617</td>
<td>267</td>
<td>72</td>
<td>&lt;40</td>
<td>&lt;40</td>
</tr>
<tr>
<td>Lymphocytes (cells/μL)</td>
<td>1,636</td>
<td>1,228</td>
<td>419</td>
<td>700</td>
<td>1,080</td>
<td></td>
</tr>
<tr>
<td>CD4 count (cells/μL)</td>
<td>189</td>
<td>220</td>
<td>110</td>
<td>184</td>
<td>186</td>
<td></td>
</tr>
<tr>
<td>CD4 percentage</td>
<td>11.6%</td>
<td>17.9%</td>
<td>26.4%</td>
<td>27%</td>
<td>19%</td>
<td></td>
</tr>
<tr>
<td>T4/8 ratio</td>
<td>0.16</td>
<td>0.29</td>
<td>0.44</td>
<td>0.44</td>
<td>0.33</td>
<td></td>
</tr>
</tbody>
</table>

Figure 2: Same child after three weeks of treatment with amoxicillin and clavulanic acid.

Figure 3: After three weeks of amoxicillin and clavulanic acid and two weeks of clindamycin, the swelling and erythema evolved.

Figure 4: Cervical computed tomography scan showed a conglomerate of right cervical lymphadenopathies with areas of necrotic tissue, multiple small left cervical and bilateral supraclavicular lymphadenopathies.
infected, tests negative for HIV or the maternal infection occurs postnatally and the child is infected through breastfeeding.

Perinatally infected infants usually progress earlier than older children or adults, because they are infected before complete maturation of the immune system. Newell et al. reported a mortality rate of 35.2% of HIV-infected infants in the first year of life [5]. In the CHER trial, 16% of untreated and 4% of treated HIV-infected infants died during the median follow-up of 40 months [6]. These infants are called rapid progressors. Those who present in childhood or adolescence are called slow progressors. The boy in the case reported is a slow progressor. He had no symptoms suggestive of HIV infection until the lymphoma developed. Using the Centers for Disease Control and Prevention classification system, he has a stage 3 HIV infection (CD4 count of less than 200 cells/mm$^3$ and an AIDS-defining illness, non-Hodgkin lymphoma).

Since the INSIGHT and TEMPRANO studies in 2015 [7, 8], the paradigm for treatment guidelines regarding initiation of HAART in HIV-infected patients has changed and now HAART is recommended for all patients regardless of CD4 counts. Both these studies showed that early HAART was associated with a decline in severe morbidity. Early treatment was already recommended in infants under 12 months of age after the CHER trial proved a reduction of infant mortality by 76% and of HIV progression by 75% in infants who were immediately started on HAART versus those who were deferred until clinical criteria or CD4 percentages were met [6]. Despite there not being randomized controlled trials supporting early versus deferred treatment in children older than one year of age, PENTA has acknowledged that there is evidence of the long-term benefits of early HAART and since 2016 has recommended treatment for all children with HIV infection [9].

Once-daily antiretroviral regimens are available for HIV-infected older children and adolescents. Despite a theoretical advantage compared to twice-daily regimens in regards to better adherence, this was not proven in the Madrid children and adolescents cohort study [10]. Nevertheless simplification of HAART regimens should remain one of the goals of HAART prescription. In this case, we chose Atripla® (emtricitabine+tenofovir+efavirenz) because it is an extensively researched drug with a reasonable safety profile.

The differential diagnosis of chronic localized lymphadenitis in HIV-infected children includes tuberculous lymphadenitis (when in the cervical region, this is called scrofula), non-tuberculous mycobacterial infection, cat scratch disease, HIV infection and lymphoproliferative disorders. A lymphoproliferative disorder was wrongly excluded earlier because this patient had had a fine-needle aspiration cytology and flow cytometry which were both negative for cancer cells. Since he had no history of contact with tuberculosis, we initially considered the HIV as the causative agent, associated with a bacterial skin infection. Considering there was no improvement with two courses of antibiotics and because the cervical CT findings of a large conglomerate of cervical lymphadenopathies with central areas of necrosis as well as supravacular adenopathies were suggestive, tuberculous lymphadenitis was considered as a possible diagnosis.

Testing for active tuberculosis in HIV patients is made difficult by the unreliableness of both the tuberculin skin test and the interferon-gamma release assays in HIV-infected patients [11–13]. Culture of Mycobacterium tuberculosis is still the gold standard for diagnosis of tuberculosis infection, but the long incubation period of six to eight weeks makes it impractical. Though molecular techniques allow for a rapid diagnosis, in this case this method was not available. Screening for Mycobacterium tuberculosis infection was done on admission to the HIV clinic with a tuberculin skin test, which was negative, and a chest X-ray, which was normal. Both the acid-fast bacilli staining and culture of Mycobacterium tuberculosis were negative and the biopsy eventually revealed a non-Hodgkin lymphoma.

HIV-infected children are at a higher risk of developing cancer than their non-infected counterparts. In literature, this ratio has varied from 10.08 (95% CI 5.87–16.14) in

Figure 5: Despite treatment for tuberculous lymphadenitis, the skin infection worsened.
the Pediatric AIDS Clinical Trials Group [1] up to 2288 (95% CI 920–4715) in the report by Evans et al. [2]. The overall prevalence of malignancy in HIV-infected children has been reported as 3.6% in the pre-HAART era [2]. Highly active antiretroviral therapy (HAART) has reduced the incidence of Kaposi’s sarcoma and non-Hodgkin lymphoma, both AIDS-defining conditions [14, 15]. This is related to the fact that HAART restores immune function and therefore, reduces incidence of all AIDS-defining conditions. However, non-AIDS-defining cancers have been on the rise in adults [16, 17], mainly because of the growth and aging of the HIV population. There is some evidence that the rate of non-AIDS-defining cancers may be increasing in the pediatric population as well [15, 18].

Potential explanations for the increased cancer incidence in HIV patients include HIV-related immunodeficiency, increased prevalence of known cancer risk (such as smoking), a direct pro-oncogenic effect of HIV through viral proteins such as tat and VPr, co-infections with known oncogenic viruses (Burkitt lymphoma is associated with the Epstein–Barr virus, Kaposi’s sarcoma with the HHV-8 and cervical cancer with the human papilloma virus) and the activated inflammatory and coagulation pathways [19, 20].

Whereas in Europe and United States of America, the most common cancer in HIV-infected children is non-Hodgkin lymphoma [2], in Sub-Saharan Africa Kaposi’s sarcoma, which is associated with human herpes virus 8 (HHV-8), is more prominent [20]. Other cancers that are frequently associated with HIV infection include leiomyosarcoma and Hodgkin’s lymphoma [7, 20].

HIV-infected children often present with more advanced malignancies than non-infected children [21]. This may be due to a faster proliferation of cancer cells unchecked by a deficient immune system [20, 22]. As it happened in this case, a delay in cancer diagnosis is common, because clinical signs of HIV are often similar to those of cancer [20].

Problems in the management of cancer in HIV-infected children include a higher incidence of opportunistic and/or aggressive infections, possible drug interactions between HAART and cytotoxic drugs and multiorgan dysfunction caused by the virus [23].

Several studies have supported the initiation or maintenance of HAART during treatment with cytotoxic drugs, as HAART has been showed to increase the chance of remission and survival rates [3, 18, 23] though some concerns remain regarding potential drug interactions. Protease inhibitors and non-nucleoside reverse transcriptase inhibitors are known inhibitors or inducers of the cytochrome P450 enzymes [24, 25]. Through induction antiretroviral drugs can decrease the efficacy of cytotoxic drugs and through inhibition they increase their toxicity [24, 25]. Despite this, there are no current guidelines on dose adjustment of HAART or chemotherapy drugs [24, 26]. However, it seems reasonable to substitute zidovudine for another antiretroviral drug because of the added risk of bone marrow suppression.

In the pre-HAART era, the prognosis of malignancy in HIV-infected children was very poor. Out of the seven HIV-infected children with non-Hodgkin lymphoma reported by Evans et al. in 1997, only one had survived at four years of follow-up [2]. This child was one of the only two who had had standard intensive chemotherapy. The median survival time for the remaining children was 6.5 months (2–14 months). Godot et al. [27] reported on the prognosis of B cell lymphomas in HIV-infected children in the HAART era with much better results. Out of 12 children with high grade B cell lymphoma, 8 had survived at a median follow-up of 72 months. Morbidity and treatment related toxicity were similar to what is reported for non-infected children. Studies in adults have suggested that the response rate to chemotherapy and overall survival in HIV-infected patients with non-Hodgkin lymphoma are approaching those seen in the general population when controlled for the same stage of disease [28, 29].

CONCLUSION

Even though preventive measures of mother-to-child transmission of human immunodeficiency virus (HIV) have greatly reduced the incidence of pediatric HIV infection, some infants will continue to escape early detection because of late maternal diagnosis. The diagnosis of malignancy in HIV-infected patients is often made difficult by overlapping symptoms and a delayed diagnosis is common.

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Author Contributions
Catarina Barreiros Couto – Substantial contributions to conception and design, Acquisition of data, Drafting of the article, Final approval of the version to be published
Sofia Fraga – Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor of Submission
The corresponding author is the guarantor of submission.

Source of Support
None

Conflict of Interest
Authors declare no conflict of interest.

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REFERENCES

Berry syndrome: One stage surgical repair in a neonate

Somia Razzaq, Nadeem Aslam, Waris Ahmad, Muneer Amanullah

ABSTRACT

Introduction: Interruption of aortic arch, aortopulmonary septal defect, patent ductus arteriosus and anomalous origin of right pulmonary artery is a rarely encountered constellation of anomalies. Case Report: We present a case of a 14-day-old male neonate with persistent respiratory distress. Echocardiogram and computed tomography angiography confirmed the constellation of Berry syndrome. A one stage surgical correction was successfully performed. Hypoplastic aortic arch was reconstructed using end to end anastomosis. Aortopulmonary septal defect was closed using single patch technique and the right pulmonary artery was re-implanted into the main pulmonary artery. Conclusion: Early clinical recognition with prompt surgical correction is associated with good outcomes.

Keywords: Anomalous right pulmonary artery, Aortopulmonary window, Berry syndrome, Interrupted aortic arch

INTRODUCTION

Berry et al. [1] in 1982 described the constellation of distal aortopulmonary septal defect, aortic origin of right pulmonary artery, interruption of aortic arch, intact ventricular septum and patent ductus arteriosus as a syndrome (Figure 1). Aortopulmonary window has been classified based on the location of the septal defect [1, 2]. Type I comprises proximal communication with normal origin of pulmonary arteries. Type IIA consists of distal defect with straddling right pulmonary artery that originates from the aortopulmonary communication but maintains its continuity with the left pulmonary artery. Type IIB consists of distal communication with origin of right pulmonary artery solely from the aorta. Type III consists of multiple aortopulmonary communications. Of all the published reports Berry syndrome has not been found to have been associated with chromosomal abnormalities except in one patient with trisomy 13 [3].

Prenatal diagnosis of this rare complex congenital anomaly is related with improved outcomes due to prompt initiation of prostaglandin infusion [4]. Detailed anatomic depiction using echocardiogram and other diagnostic procedures are crucial for planning the surgical repair.
modalities with early surgical repair is essential, thereby avoiding pulmonary hypertension associated with pulmonary over circulation and subsequent heart failure [5–7].

A number of reports in literature have described the utilization of either one-stage or two-stage surgical correction of this complex congenital cardiac anomaly. We describe a case of successful one-stage surgical repair in a neonate.

CASE REPORT

A male baby was born by cesarean section with a birth weight of 4.2 kg at 37th week of gestation. Shortly after birth, the neonate developed respiratory distress. The perinatal history was unremarkable. On physical examination the baby was tachypneic with a respiratory rate of 65 breaths/min and heart rate of 136 beats/min. The blood pressures of both arms were equal. The blood pressures of the lower limbs were lower by 15 mmHg. Differential cyanosis was detected by pulse oximetry measurement with upper limb oxygen saturation of 94% and lower limb oxygen saturation of 87%. On auscultation a grade 3/6 systolic murmur was heard at the left sternal border. Femoral pulses were barely palpable. Chest X-ray demonstrated borderline cardiomegaly with subtle pulmonary plethora. Transthoracic echocardiogram revealed a Type A interrupted aortic arch, a 1.5-mm patent ductus arteriosus, a 12-mm Type IIB aortopulmonary window and possibility of anomalous origin of right pulmonary artery from descending aorta (Figure 2). Doppler study showed supra systemic pulmonary artery and right ventricular pressures. A computed tomography angiography confirmed the presence of interrupted aortic arch, anomalous origin of right pulmonary artery from the proximal part of right aortic wall and a patent ductus arteriosus which continued as descending aorta (Figure 3A–C). Prostaglandin infusion was initiated to maintain patency of ductus arteriosus.

Operative Procedure

On the 14th day of life the neonate underwent one-stage surgical correction. The heart was exposed via a median sternotomy. The anatomy was recognized to be consistent with constellation of Berry syndrome (Figure 4). Extensive dissection was done in order to mobilize the arch vessels, ascending and descending aorta and patent ductus arteriosus. The pulmonary arteries were dissected well into the hilar branches. After heparanization, arterial cannula was placed below the innominate artery to leave enough space above the aortopulmonary window to place the aortic cross clamp. A single venous cannula was placed in the right atrium. Cardiopulmonary bypass was initiated and the patient was cooled to 18°C. Aortic cross clamp was applied and crystalloid cold blood cardioplegia was infused. The pulmonary arteries were snugged. Deep hypothermic circulatory arrest was established. Aortic arch vessels were snugged. Aortic cross clamp was removed. The ductus arteriosus was divided and transfixed (Figure 5A). All ductal tissue was excised until normal aortic tissue. The aortic arch was incised and an end-to-end anastomosis was constructed using 7-0 polypropylene (Figure 5B). Anterior aspect of the aortic anastomosis was augmented with bovine pericardial patch (Figure 5C). Aortic cross clamp was reapplied and aortic arch vessels were unsnugged. Cardiopulmonary bypass was reinstated and a second dose of cardioplegia was infused. During the rewarming period a vertical incision over the aortopulmonary window was made and the septal defect was closed using bovine pericardial patch with a 6-0 polypropylene. Aortic cross clamp was removed. The patient spontaneously converted to sinus rhythm.

Figure 1: Berry syndrome consisting of aortopulmonary septal defect, interrupted aortic arch, patent ductus arteriosus and aortic origin of the right pulmonary artery.

Partial occluding clamp was applied to the aorta and the right pulmonary artery was excised from the aorta. The aortic defect was closed with 6-0 polypropylene. The right pulmonary artery was anastomosed to the right lateral aspect of the main pulmonary artery with 6-0 polypropylene hence obtaining right pulmonary artery to main pulmonary artery continuity (Figure 5D). The patient was easily weaned off from cardiopulmonary bypass with minimal inotropic support. The total cardiopulmonary bypass time was 140 minutes, with a cross clamp time of 80 minutes. The circulatory arrest time was 46 minutes. Elective delayed sternal closure was performed 36 hours later.

The patient was extubated on postoperative day-6 and discharged on postoperative day-12. Postoperative echo and Doppler study demonstrated patched aortopulmonary window with no significant residual defect, mild turbulence in the right pulmonary artery with pressure gradient of 20 mmHg, mild tricuspid regurgitation with maximum gradient of 18 mmHg.

**DISCUSSION**

Berry syndrome is a very rare complex congenital cardiac anomaly that is amenable to surgical repair. The pathogenesis of this anomaly is unknown, however, proposed hypothesis states that the failure of formation of aortopulmonary septum leads to anomalous origin of pulmonary arteries from the undivided truncal segment. This anomalous origin of the pulmonary arteries induces a steal phenomenon hence reducing aortic blood flow during embryogenesis resulting in hypoplasia of the aortic arch [1].

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Figure 2: Echocardiography parasternal short axis view showing aortopulmonary window and anomalous origin of right pulmonary artery from ascending aorta.

Abbreviations: AAO: Ascending Aorta, APW: Aortopulmonary Window, MPA: Main Pulmonary Artery, RPA: Right Pulmonary Artery

Figure 3: Computed tomography angiography (A) Axial view showing aortopulmonary window and anomalous origin of right pulmonary artery from ascending aorta, (B) Aortopulmonary window and interrupted aortic arch, and (C) 3D reconstructed image showing interrupted aortic arch.

Abbreviations: AAO: Ascending Aorta, APW: Aortopulmonary Window, LPA: Left Pulmonary Artery, MPA: Main Pulmonary Artery, RPA: Right Pulmonary Artery, DAO: Descending Aorta, IAA: Interrupted Aortic Arch
Berry syndrome has also been reported to be associated with anomalous origin of left coronary artery from the main pulmonary artery [8]. Patients with Berry syndrome present with respiratory distress, metabolic acidosis, anuria, severe congestive heart failure or acute cardiovascular collapse with spontaneous closure of ductus arteriosus in the first few days of life. Early clinical diagnosis with detailed preoperative recognition of these unique anatomic features is therefore essential for good surgical outcomes [2, 3, 5–7]. Long-term follow-up is required after surgical correction as stenosis at the site of anastomosis is a potential complication. The stenosis can be relieved adequately by percutaneous balloon angioplasty [9, 10].

Surgical repair can be done using either two-stage or one-stage approach. Two-stage repair involves reconstruction of aorta with ligation of patent ductus arteriosus followed by complete closure of aortopulmonary window in the next stage [11]. Ghelani et al. advocate the utilization of two staged surgical repair in case of premature infants and small for gestational age infants [4]. However, one stage surgical repair is considered superior, the rate of reoperation varies with development of complications.

Berry et al. reported eight cases out of which two patients underwent successful one-stage surgical repair, with reconstruction of aortic arch using either left subclavian or Dacron graft, closure of aortopulmonary window and reimplantation of right pulmonary artery to the main pulmonary artery [1].

Ding et al. reported a case of one-stage repair using Dacron baffle to obliterate the aortopulmonary septal defect and maintain continuity of right pulmonary artery with the main pulmonary artery with repair of hypoplastic aortic arch using end-to-end anastomosis [12]. The utilization of intra-aortic baffle is associated with obstruction of right pulmonary artery and left ventricular outflow tract. The size and growth potential of the baffle also pose as a problem to be utilized successfully in neonates [9].

Re-routing of right pulmonary artery by making a tunnel, either by suturing of native aortic wall tissue [13, 14] or utilization of glutaraldehyde fixed autologous pericardial patch [7] with simultaneous closure of aortopulmonary window is another reported successful surgical technique. Stenosis at the site of repair is a major complication of this method of repair [9].

Right pulmonary artery arterioplasty with aortic cuff formation is another method of repair in case of Berry syndrome as utilized by Abbruzzese et al. [15]. In this case, the aorta has to be transected which is associated with increased risk of bleeding along with difficult anastomosis [9].

Burke et al. [16] reported a case of successful arterial switch for the repair of Berry syndrome in a neonate. The aorta was separated from the pulmonary artery. The right pulmonary artery was anastomosed to the main pulmonary artery after being excised completely from

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**Figure 4:** Intraoperative finding confirming the presence of constellation of Berry syndrome. Abbreviations: AAo: Ascending Aorta, APW: Aortopulmonary Window, PA: pulmonary artery, RA: Right Atrium

**Figure 5:** (A) After Arterial and venous cannulation the great arteries and pulmonary arteries were snugged. The ductus arteriosus was transected and ligated, (B) The aortic arch was incised and an end-to-end anastomosis was made with the posterior wall of the descending aorta, (C) The anterior aspect of the aortic anastomosis was augmented with bovine pericardial patch, and (D) The complete repair with anastomosis of descending aorta to the ascending aorta and the right pulmonary artery anastomosed to the main pulmonary artery running posterior to the aorta. Abbreviations: AAo: Ascending Aorta, APW: Aortopulmonary Window, BCA: Brachiocephalic Artery, DAo: Descending Aorta, LCA: Left Carotid Artery, LPA: Left Pulmonary Artery, LScA: Left Subclavian Artery, MPA: Main Pulmonary Artery, PDA: Patent Ductus Arteriosus
the right lateral aspect of the aorta. The descending aorta was connected via end-to-end anastomosis to the defect in the ascending aorta. This technique demonstrated easy anterior transfer of pulmonary arteries in neonates. This leads to decreased risk of compression of right pulmonary artery by the aorta and the airway [9].

Park et al. in 2008 [10] demonstrated two cases of single stage repairs. In this technique, the posterior wall of the ascending aorta was utilized to form confluence between the right pulmonary artery and the main pulmonary artery. The anterior half of the right pulmonary artery was repaired with an autologous pericardial patch. Utilization of this patch technique avoids undue tension and stenosis at the site of anastomosis.

We have implemented a surgical technique that demonstrates the reconstruction of hypoplastic aortic arch by end-to-end anastomosis with patch augmentation of anterior aortic wall. This eliminates the risk of postoperative aortic arch stenosis henceforth decreasing the risk of proximal segment dilation and compression of right pulmonary artery and the left main bronchus [9]. The detachment and anastomosis of the right pulmonary artery to main pulmonary artery behind the aorta maintains the geometry of the pulmonary arteries. This technique is preferred in neonates due to potential growth problems associated with patch repair or the utilization of intra-aortic baffles, thereby also reducing the risk of left ventricular outflow tract obstruction [9, 12, 16, 17].

CONCLUSION

Berry syndrome is a rare congenital anomaly. Echocardiography in an important diagnostic tool in the initial evaluation but computed tomography angiogram or cardiac magnetic resonance imaging scan is frequently required. One-stage surgical correction has demonstrated acceptable outcomes and is now considered the procedure of choice. Stenosis at the site of anastomosis is commonly encountered. Reoperations are mainly due to development of aortic or right pulmonary artery stenosis.

REFERENCES

A large amniocele through a fundal uterine defect diagnosed on 2D ultrasound imaging

Joshua Oluwafemi Aiyekomogbon, Ojah S.O., Shinkafi S.M., Agom B.

ABSTRACT

Introduction: Amniocele is herniation of amniotic sac through a uterine myometrial defect. It commonly results from intrauterine procedures such as suction evacuation, dilatation and curettage, manual removal of placenta and other products of conception, etc. Early diagnosis is imperative as that will go a long way to reduce morbidity and mortality. Case Report: A 35-year-old G4P3+0 woman was presented to our health facility at 21st week gestation with a week history of lower abdominal pain. Obstetric ultrasound scan done at presentation revealed a fundal myometrial defect with a large amniocele through it, resulting in apparent oligohydramnios. Her first two deliveries were uneventful, but the third was complicated by post-partum hemorrhage due to retained placenta. This necessitated instrumentation and manual removal in the theatre at a secondary Health institution, four years prior to the index pregnancy. No immediate complication was noticed after the procedure. Two weeks after this diagnosis, precisely at 23rd week gestation, she had laparotomy, aimed at reducing the herniated amniotic sac and repairing the defect. This was not possible due to the narrow neck of the defect. She was then closed back and kept on exclusive bed rest and weekly sonographic evaluation of the pregnancy. A scan done at 31st week gestation revealed ruptured herniated amniotic sac into the peritoneal cavity with fetal distress. Estimated ultrasound fetal weight at this time was 1.76 kg. She had emergency cesarean section with delivery of a live very low birth weight baby whose APGAR score was 7 and 8 at 1 and 5 minutes respectively. The baby was admitted into special care baby unit (SCBU) but died 10 hours thereafter. The myometrial defect and uterine incision were repaired in layers. She did well, hemodynamically stable and was discharged on postoperative day-7. Follow-up visits were satisfactory. Conclusion: This case is presented to emphasize the need for training and retraining of physicians and midwives on active management of third stage of labor to forestall the observed pitfalls. It also brings to the fore the utility of gray scale and color Doppler ultrasound in antenatal diagnosis of uterine defect and placenta accreta.

Keywords: Amniocele, Pregnancy, Retained placenta, 2D Ultrasound

How to cite this article


Article ID: Z01201712CR10860JA

doi: 10.5348/ijcri-2017121-CR-10860
INTRODUCTION

Uterine perforation is a serious complication resulting from intrauterine procedure such as suction evacuation, dilatation and curettage, manual removal of retained placenta and other products of conception, insertion of intrauterine contraceptive device, hysterosalpingography, hysteroscopy and during procedures for in vitro fertilization. Invasive mole is also identified as a possible cause of uterine perforation [1–3]. Spontaneous uterine perforation from pyometra, placenta accreta, and rarely degeneration of a myoma and uterine infarction has also been documented [4, 5].

Retained placenta is one of the most common causes of postpartum hemorrhage and has an incidence of 1:100 to 1:300 births [6, 7]. Placenta adherens, incarcerated placenta and placenta accreta are the common causes of retained placenta [8]. Placenta increta is an abnormal placental implantation in which placenta villi invade into the myometrium, while in placenta percreta, placental villi penetrate through the uterine serosa or adjacent organ, usually the urinary bladder [9, 10]. Krapp et al. [6] introduced the use of color Doppler sonography in the third stage of labor to diagnose abnormal placental invasion, and differentiating it from normal placental separation. Normally, there is cessation of blood flow between placenta and myometrium immediately after birth, but in placenta accreta, a persistent blood flow from the myometrium deep into the placenta is demonstrated sonographically. This helps in differentiating placenta accreta from other causes of PPH. Using this Doppler assessment, management such as manual removal of placenta can be instituted early and by this, maternal morbidity and mortality are greatly reduced. Some biochemical markers such as elevated levels of maternal serum creatinine kinase, alpha fetoprotein, β-human chorionic gonadotropin, cell-free fetal DNA, placenta mRNA and DNA microarray are also used for the diagnosis of abnormalities of placental invasion [10–12].

Cervical dilators, curette and uterine sound are the most common instruments that cause perforation [13] and the most commonly perforated segment of the uterus is the relatively avascular fundus [14] which fits into the pattern experienced in the index case.

CASE REPORT

A 35-year-old Gravida 4, Para 3+0 (G,P,“G”) female presented at our health facility on account of lower abdominal pain of a week duration at 21st week gestation. She had no fever, vaginal discharge or bleeding per vaginum. Her first two pregnancies were uneventful but had postpartum hemorrhage (PPH) due to retained placenta after her third delivery in 2012, i.e., four years prior to the index pregnancy. This necessitated instrumentation (curettage) after failed manual removal of retained placenta in the theatre. She was fine after the procedure. Her blood pressure at this presentation was 120/70 mmHg and pulse rate was normal. Ultrasound of the patient done at presentation showed amniotic sac herniation through a fundal myometrial defect at 21st week of gestation (Figure 1).

The fetus was active with no sign of distress sonographically although, there was apparent oligohydramnios. She was placed on bed rest and weekly sonographic evaluation for the assessment of fetal well being. She had a laparotomy done after two weeks (at 23rd week gestational age), aimed at reducing the herniated sac and repairing the defect which also trapped the fimbrial end of the left uterine tube. Intra-operative picture is shown in Figure 2. It was difficult reducing the sac as the neck of the defect was narrow, and any attempt to force it in may lead to its rupture. Consequently, she was closed back while weekly sonographic assessment for fetal well being and exclusive bed rest were continued. The herniated sac continued to increase as the pregnancy advanced. At 30th week gestational age, the herniated sac dimension had increased to 17x18.8 cm as against the initial dimension of 6x7 cm at 21st week gestation. She noticed a sudden reduction in the size of her gravid abdomen six days after the last scan and this necessitated a repeat ultrasound scan which revealed ruptured herniated sac with features of fetal distress as shown by fetal Doppler scan (Figure 3). Estimated fetal weight was 1.76 kg at this time.

The patient had emergency cesarean section in view of the recent development, and a very low birth weight male neonate was delivered alive through a low transverse uterine incision with APGAR score of 7 and 8 at 1 and 5 minutes respectively. The child was immediately admitted into special care baby unit (SCBU) but he died ten hours thereafter. The myometrial defect (Figures 4 and Figure 5), and the low transverse incision were repaired in layers after freeing the trapped left fallopian tube. She had two units of whole blood transfused intra-operatively due to excessive blood loss. She was discharged home on postoperative day-7 after satisfactory wound healing and hemodynamic stability. The discharge hemoglobin was 11.0 g/dl and subsequent follow-up visits were satisfactory.

DISCUSSION

Uterine perforation is a defect of the uterine layers, with or without the involvement of the serosa [15]. It is a live-threatening condition for the mother and the fetus; active management is therefore imperative. It commonly results from intrauterine procedures such as suction evacuation, dilatation and curettage, manual removal of placenta to mention a few [2, 3]. We report an unusual case of myometrial defect with amniocele due to previous intrauterine procedure for removal of retained placenta four years prior to presentation. The perforation was subclinical and was presumably walled-off by fibrotic tissues.
until the index pregnancy when the defect gave way with subsequent herniation of amniotic sac through it, resulting in severe oligohydramnios. This was diagnosed at 21st week gestation by 2D ultrasound. She was clinically stable with no sign of fetal distress initially.

Most patients with uterine perforation often present with hemorrhagic shock, abdominal pain and fetal distress. The fetus in most cases is compromised with neurological deficit because of hypoxia [16, 17]. This patient just presented with abdominal pain and the fetus was not in obvious distress at presentation despite the apparent oligohydramnios. No hemoperitoneum was noted clinically and radiologically, and she was hemodynamically stable with hematocrit level of 12 g/dl. The general belief is that a herniated amniotic sac into the abdominal cavity is strongly suggestive of imminent uterine rupture [18]. However, uterine rupture does not always occur immediately after sonographic
detection of extrusion of the amniotic sac because a thin layer of myometrium and even uterine serosa may be around the herniated sac which may be difficult to detect sonographically [17]. Magnetic resonance imaging (MRI) scan is required to appreciate this myometrial layer and even its thickness [17]. This was not feasible in our health institution as MRI scan was not available. Serial ultrasound scan was done to closely monitor her for possible rupture instead. This was discovered at 31st week gestation but there was no hemoperitoneum despite the rupture as the uterine perforation was at the fundus which is relatively less vascular when compared to perforation at the cervical region which tends to bleed more. She only noticed a sudden reduction in the size of her gravid abdomen. This discovery, coupled with fetal distress necessitated emergency Cesarean section. This was largely successful except that the neonate could not survive presumably due to birth asphyxia.

The management of amniocele depends on the gestational age and the clinical state of the mother and fetus [17, 18]. Before fetal lung maturity, two management protocols are advocated; these are surgical management and conservative management [17]. Both may be employed in some cases as it was in the index case. Surgical intervention involves termination of pregnancy and repair of the uterine wall defect, while conservative (expectant) management involves close monitoring, serial ultrasound evaluation for fetal well being and when available, serial MRI scan to evaluate surrounding myometrial thickness for possibility of rupture, and exclusive bed rest. The index patient benefitted from both protocols. Her initial surgical management was not aimed at terminating the pregnancy but to preserve it. It was aimed at reducing the sac and repairing the defect antenatally. When that failed, conservative management was instituted. This was however abruptly terminated by sudden rupture of the herniated amniotic sac with early signs of fetal distress at 31st week gestation.

This study is aimed at preventing uterine perforation when managing third stage of labor or when conducting other intrauterine procedures for any reason, as that was the genesis of the maternal morbidity and child mortality observed in the index case. Several reports have suggested increased perforation rates by junior trainees in USA, Singapore and Nigeria [12, 19, 20]. Five-fold increase rate of perforation has been reported by junior staff in USA [12] while 82.5% of perforations were caused by junior staff in Singapore [19]. Some of these cases are even performed by untrained personnel such as Nurses, Midwives, and community health extension workers (CHEW) in Nigeria [20]. Early identification and diagnosis of uterine perforation and subsequent management will significantly reduce morbidity, long-term sequelae and even mortality [21].

CONCLUSION

Amniotic sac herniation through a myometrial defect, a sequelae of intrauterine procedures such as manual removal of placenta, dilatation and curettage and suction evacuation is globally rare but common in developing world as untrained or poorly trained personnel engage in such procedures with negative consequences as observed in the index case. The utility of gray scale and color Doppler ultrasound in antenatal diagnosis of uterine defect and placenta accreta is re-echoed.

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Figure 5: Picture of the second surgical exploration showing the fundal uterine myometrial defect (inverted arrow) prior to surgical closure.

**Author Contributions**

Joshua Oluwafemi Aiyekomogbon – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Ojah S.O. – Substantial contributions to conception and design, Revising it critically for important intellectual content, Final approval of the version to be published

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**Guarantor of Submission**

The corresponding author is the guarantor of submission.
Source of Support
None

Conflict of Interest
Authors declare no conflict of interest.

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REFERENCES
Simultaneous hemopericardium, hemoperitoneum and hemothorax after complete surgical resection of mediastinal cystic lymphangioma: A case report

Alexis Mupepe Kumba, Filippo Banchini, Luigi Conti, Rocco Delfanti, Patrizio Capelli

ABSTRACT

Introduction: Lymphangioma is an atypical non-malignant tumor. It is most commonly observed in pediatric population and typically found in the cervical region. Its prevalence is estimated between 0.01% and 4.5% of all mediastinal tumors. Complete surgical resection is required to prevent recurrence. Case Report: We present the case of a 47-year-old male with massive right pleural effusion. Magnetic resonance imaging showed a massive mediastinal cystic lymphangioma. A radical excision of the thymus and lymphangiomatosis cystic mass was performed by sternotomy. The postoperative course was marked by simultaneous hemopericardium, hemoperitoneum and hemothorax, the origin of which, was not well explained due to its presentation in the distal site of surgery. This required transfusions and new thoracoabdominal surgical exploration. The patient developed pulmonary and wound infections and died of sepsis and multi-organ failure. Conclusion: Our case would be a further demonstration of an unusual complication of simultaneous hemopericardium, hemoperitoneum and hemothorax explained by some authors as a local production of fibrinolysin.

Keywords: Cystic lymphangioma, Hemoperitoneum, Hemothorax, Mediastinum neoplasm, Pericardial effusion

INTRODUCTION

Lymphangioma is an atypical non-malignant tumor, which occurs during the embryonic development of the lymphatic system. It is most frequently observed in the head and neck, but can occur at any location in the body [1–3]. The exact incidence is unknown, but its prevalence is estimated to be between 0.01% and 4.5% of all mediastinal tumors [4–6]. Lymphangiomas have been classified into three types: simplex lymphangioma (capillary lymphangioma), cavernous lymphangioma, and cystic lymphangioma (cystic hygroma) [7]. Since the mediastinum is hypobaric, most lymphangiomas are cystic. [5]. Largely asymptomatic, cystic lymphangioma can present a diagnostic challenge often requiring lymphoscintigraphy and magnetic resonance imaging (MRI) scan in order to be identified. Treated with complete
surgical resection, long-term prognosis is excellent [5, 8]. We report the case of a young man with mediastinal cystic lymphangioma who developed hemopericardium, hemoperitoneum and hemothorax of unknown origin one day after complete surgical resection and later died.

CASE REPORT

A 47-year-old male was transferred to our institute from a nearby hospital for massive right pleural effusion. He presented with tachypnea, hypoxia (PO$_2$ 66 mmHg on room air, PCO$_2$ 33 mmHg, SaO$_2$ 94%), without fever. Respiratory movements were absent in the right pulmonary fields. The abdomen was increased in volume with ascites effusion and a slight defensive reaction. Biological examinations were normal, except for CRP (1.17 mg/dl), white blood cells (16.95x10$^3$/µl) and D-dimer (3200 µg/l). A chest radiograph, on frontal projection, showed a homogeneous water-density mass in the right pulmonary fields and in the paratracheal area (Figure 1).

An additional thoracoabdominal computed tomography scan revealed abundant pleural effusion associated with collapse of the right lung, and the presence of multiple swollen lymph nodes (7 cm maximum axial diameter) partially confluent in the upper mediastinum and cardiophrenic angle. Multiple hypodense structures of about 12 mm maximum axial diameter were observed around the spleen, lymph nodes (2.5 cm of the maximum axial diameter) were also observed in the obturator region bilaterally, along the internal and external iliac chain and other lymph nodes (1.5 cm of the maximum axial diameter) in the paracaval and para aortic regions.

We decided to perform diagnostic thoracoscopy due to the presence of unknown mediastinal mass that revealed chylous effusion along the mediastinal pleura, diffuse thickening and fibrosis of mediastinal tissue dripping chyle. A biopsy of the mediastinal tissue in the Barety lodge until tracheal plane was carried out. Histologic examination of the biopsy sample showed fibroadipose tissue comprising residues of timing structures, dilated vascular spaces provided with flat endothelial coating positive for CD31, CD34 and Podoplanin (D2-40).

Magnetic resonance imaging (MRI) scan revealed a massive lymphangioma extending craniocaudally 22 cm and 9 cm in transverse diameter involving the entire anterior and posterior mediastinum surrounding the lower third of the esophagus and involving the celiac trunk with hypointensity in T1 and low hyperintensity in T2 (Figure 2). Therefore, a radical excision of the thymus and lymphangiomatosis cystic mass was performed by sternotomy and debulking of the mediastinum without opening the abdomen and pericardium (Figure 3).

Final histology pointed to cystic lymphangioma with overlapping outbreaks of chronic and acute suppurative inflammation. The postoperative course was marked by simultaneous hemopericardium, hemothorax and hemoperitoneum which required transfusions and...
new surgical exploration. A thoracoabdominal incision enabled both chest and abdomen exploration at the same time, without objectifying the source of the blood effusion. Packing was performed to achieve hemostasis. Pericardial blood effusion was evacuated by left anterior thoracotomy. As a consequence of this complication, the patient developed pulmonary and wound infections and died of sepsis and multi-organ failure despite VAC-therapy and oriented antibiotics.

DISCUSSION

Lymphangioma is more commonly observed in the pediatric population and is typically found in the cervical region, because of its derivation from a primitive lymphatic sac. Ninety percent of cases are diagnosed by two years of age and are more common in females [1, 4, 6]. In a study of 37 cases with lymphangioma, Rajesh et al. found a mean age of 45 years [1]. Higher prevalence of lymphangioma is seen in patients with chromosomal abnormalities like Down syndrome, Turner syndrome, Edwards syndrome, and Patau syndrome. An acquired variant of lymphangioma can occur in patients due to chronic obstruction of the lymphatics. This is seen more commonly in middle-aged patients with a history of surgery, those undergoing radiation therapies for malignancy, and those suffering from a chronic infection [1]. In the pediatric population, lymphangioma is typically found in the cervical region but, in adults, it commonly presents as a mediastinal mass without a cervical component [4].

In this case, the patient was an adult male with a mediastinal mass. Screening for chromosomal abnormalities was not performed as no morphological sign matches were present. There was no history of surgery neither of chronic infection nor radiotherapy. Most lymphangiomas are asymptomatic [9]. Symptomatic patients are often found to have large lesions. The symptomatic manifestation can be variable depending on the site. Dyspnea, respiratory distress and hemoptysis are the most common symptoms that occur when the lymphangioma compresses the tracheobronchial tree, pharynx, or phrenic nerve, or presents as pleural effusion [4, 10, 11]. The patient presented pleural effusion, dyspnea and abdominal distension. Lymphangiomias can affect all compartments, but anterior and superior mediastinum are those most commonly affected as in our case. Some unusual sites described are the hilum, pericardium, axilla, shoulder joint, retroocular region, esophagus and chest [12]. Imaging techniques such as computed tomography (CT) scan and magnetic resonance imaging (MRI) scan may be used to make a clinical diagnosis and for follow-up. They respectively show homogeneous low density and T2 hyperintensity [12]. The latter was not confirmed in our case with only low hyperintensity in T2.

The final diagnosis is based on a combination of clinical, radiological, and histopathological findings. On CT scan and/or MRI scan, a cystic lymphangioma can be suggested based on multiseptated and loculated mass [12]. The gold standard treatment of lymphangioma is complete surgical resection [1, 2, 13]. However, due to the infiltrative nature of lymphangiomias, this procedure becomes incomplete and hard to use and can only be applied in 10–50% of the cases. Therefore, recurrence represents an important problem, with a prevalence rate between 0% and 27% after total exeresis and between 15% and 53% after partial exeresis. [2, 13, 14].

The patient presented hemopericardium, hemothorax and hemoperitoneum without any obvious cause one day after complete surgical resection. In literature, coagulopathy has been reported in association with lymphangiomatosis, and several potential mechanisms for this association have been advanced. Local production of fibrinolysin by lymphangiomias may be responsible for the coagulopathy [15]. This could explain blood effusion far from surgical site in the patient. In literature surgical complications, which occur in 19–33% of the cases, include formation of hematoma, lymphocele, scar, abscess, infection, wound dehiscence and nerve palsy have been noticed by several authors [14, 16–18].

To the best of our knowledge, simultaneous hemopericardium, hemothorax and hemoperitoneum after mediastinal lymphangioma surgery has not been reported. Various alternative treatments for lymphangiomias have been attempted; they include laser therapy, radiation therapy, chemotherapy and the use of sclerotizing agents [2, 19]. The efficacy of these treatments is highly variable in incidental reports. They can be used as a therapeutic weapon against lymphangiomias, especially in patients with unresectable or life-threatening lesions or in cases in which surgery would be mutilating. Spontaneous regression of mediastinal lymphangioma has been seen, although it remains uncommon [20]. Among sclerotizing agents, OK-432 (Picibanil), a lyophilized mixture of Su-protein of group A Streptococcus pyogenes, incubated with penicillin G, had a response rate up to 92% with minimal side effects and no cicatricial damage to the skin [21].

CONCLUSION

Mediastinal lymphangioma is a rare condition and should be included in the differential diagnosis of patients presenting with pleural effusions. The treatment of lymphangioma is well known to be a complete surgical resection, but this technique is not always possible to perform. Hemopericardium, hemothorax, and hemoperitoneum together are rare but when present, complicate surgery and are difficult to manage. Some alternative treatments have been tried with good outcomes.
REFERENCES


Radiographic changes of the mandible after proton beam radiotherapy for oral cancer: A case report

Masaru Konishi, Yoshikazu Suei, Minoru Fujita, Keiji Tanimoto

ABSTRACT

Introduction: Radiation-induced osteomyelitis or osteoradionecrosis of the jaw bone is one of the most severe adverse effects of radiotherapy sometimes experienced by head and neck cancer patients. We report on the observed radiographic changes after proton radiotherapy for a patient with squamous cell carcinoma in the mandibular bone. Case Report: We report on a case of a deeply infiltrating squamous cell carcinoma in the mandible and treated with the proton beam radiotherapy. We describe the radiographic changes of the mandible after proton beam therapy. Primary lesion disappeared and the obvious resorption of the left mandible was observed. However, the images after proton beam therapy showed continued new bone formation and regeneration of the cortical bone of the mandible. Conclusion: This case illustrates the new bone formation and regeneration of the cortical bone of the mandible after the proton beam radiotherapy. Careful observations in more cases need to verify whether the effect of the proton beam radiotherapy maintaining a given dose while decreasing unnecessary dose to surrounding normal tissue is demonstrated or not.

Keywords: Bone regeneration, Osteoradionecrosis, Proton beam radiotherapy, Radiation-induced osteomyelitis

How to cite this article


Article ID: Z01201712CR10862MK

doi:10.5348/ijcrai-2017123-CR-10862

INTRODUCTION

One of the adverse effects of radiotherapy for head and neck cancer is radiation-induced osteomyelitis or osteoradionecrosis (ORN) of the jaw bone. A lot of clinical and physical factors have been reported to be associated with the risk of ORN. They include patient-, tumor-, and treatment-related variables [1, 2]. Tolerance dose of the mandibular bone was reported from 60–72 Gy [3]. Fujita et al. reported that a total dose of 90 Gy or more and a dose rate of 0.55 Gy/h or higher were associated with a significant increase in the incidence of ORN at the brachytherapy for oral tongue cancer [4]. Oral health status is a risk factor for ORN, and oral health care is an important element in the prevention of ORN.
Good oral conditions before and after radiotherapy for all irradiated head and neck cancer patients are very important to prevent ORN [2, 5]. The multidisciplinary team communications like the consultation of dentists or dental hygienists before the radiotherapy are commonly recommended. The managements of ORN are reported such as a conservative management, ultrasound therapy, hyperbaric oxygen [6, 7]. In any case, once ORN occurred, it often can persist for a long time and be hard to manage.

Owing to the physical characteristics depositing the bulk of the radiation dose in a highly confined area of the proton beam, some lesions became treatable with the proton beam radiotherapy even if it was the treatment-resistant tumors in sensitive location [8]. Numerous studies have reported on ORN resulting from radiotherapy with X-rays. However, few reports exist on ORN resulting from proton beam radiotherapy. We report on the observed radiographic changes after proton radiotherapy for a patient with squamous cell carcinoma in the mandibular bone.

CASE REPORT

A 60-year-old male was referred to the department of oral and maxillofacial surgery with a history of persistent swelling on the left side of his face. He has been presenting with soft swelling of the left cheek and sub-mandibular region for two weeks. He was also experiencing paresthesia of the third division of the trigeminal nerve, and had limited range of motion of the jaw. The lower left second molar had been extracted and the socket was covered with soft granulation tissue. Histopathological diagnosis was based on a biopsy of the gingiva confirmed squamous cell carcinoma.

Panoramic radiography at the initial visit showed a moss-eaten appearance of the left mandible (Figure 1). Computed tomography (CT) scan revealed a well-enhanced mass and bone destruction of the left mandible (Figure 2). Initially, a surgical approach for treatment was considered. However, after 18F-fluorodeoxyglucose positron emission tomography of the region, the case was deemed inoperable as tumor invasion into the cranial fossa and was suspected along the trigeminal nerve. The patient was thus treated with chemotherapy (cisplatin and 5-fluorouracil) and proton beam therapy to 70 Gy in 35 fractions.

A panoramic image one month after proton beam therapy is shown in Figure 3. At that time, there was obvious resorption and deformity in the left mandible. The images of cone-beam computed tomography (CBCT) one month after proton radiotherapy (Figure 4) showed a possible trabecular bony formation at the location of the primary lesion. The cortical bone once adjacent to the lesion was nearly destroyed.

A panoramic image at 10th month post-proton beam therapy (Figure 5) showed a defect where the left mandible (and tumor) had originated existed and the first molar presented as a floating tooth. Computed tomography scan images at 10th month after proton beam therapy (Figure 6) showed continued new bone formation and regeneration of the cortical bone of the mandible.

Figure 1: Panoramic radiograph showing the moss-eaten appearance of the left mandible at first visit.

Figure 2: (A) Contrast-enhanced axial computed tomography (CT) images showing the enhanced soft-tissue mass in the left mandible; (B, C, D)- Axial, sagittal, and coronal CT images of hard tissues demonstrated the bone destruction with bicortical plate and the disappearance of the mandibular canal.

Figure 3: Panoramic radiograph at first month after proton beam radiotherapy showing the resorption and deformity of the left mandible.
The patient experienced extreme occlusal dysfunction as a result of the bone resorption, which was exacerbated at meal times. The patient had a local recurrence one year after proton beam therapy and perished about 19 months after proton beam therapy and 26 months after the first visit.

DISCUSSION

Proton beam therapy is one of the most technologically advanced forms of radiotherapy. Protons differ from traditional megavoltage X-ray radiotherapy owing to the characteristic Bragg peak of a proton beam. The Bragg peak can be described as the deposition of energy at a specific depth in the body as the protons decelerate. The Bragg peak can be placed at any depth in the body (corresponding to a tumor location), depending on the energy of the beam selected. Conventional radiation therapy faces challenges from side effects because of a relatively high entrance dose as well as a non-zero exit dose. By contrast, proton therapy has a substantially lower entrance dose and no exit dose that reduces damage to healthy tissue surrounding a tumor [9]. In addition to differences in dose deposition; protons have different radiobiological properties compared with photons. Specifically, the relative biological effectiveness (RBE) of protons beam is considered to be 1.1, compared with 1.0 for photons. The head and neck squamous cell carcinoma observed in this case was well suited for proton therapy owing to the improved dose deposition and RBE.

The radiographic features of ORN have many similarities to those of chronic osteomyelitis [10]. An early characteristic change due to therapeutic doses of radiation is a well-defined area of bone resorption within the outer cortical plate of the mandible. Late changes are quite variable and may be predominantly osteolytic or osteosclerotic, or a mixture. Typically, the periphery of a lesion is ill-defined, and the cortical bone of the jaws shows irregular resorption. New periosteal bone formation is uncommon. For comparison, the radiographic findings of the present case and relevant published data resulting from X-ray radiotherapy are given in Table 1 [11]. Published data from patients treated

Figure 4: (A–D) Axial, sagittal and coronal cone beam computed tomography images of hard tissues at first month after proton radiotherapy showed the bone formation in a part of the mandible.

Figure 5: Panoramic radiograph at 10th month after proton radiotherapy showing the defect of the left mandible and the first molar floating tooth.

Figure 6: (A–E) Axial, sagittal and coronal computed tomography images of hard tissues at 10th month after proton radiotherapy showed the additional bone formation and reconstruction of cortical bone in the mandible.
with X-ray radiotherapy showed sequestrum formation, and the present case also demonstrated an unclear trabecular bone sequestrum formation in the mandible. However, the presentation of this formation in this case was different than what is typically observed with osteoradionecrosis resulting from X-ray radiotherapy. Sequestrum formations typically occur in the normal bone areas of the jaw as a result of X-ray radiotherapy. In the present case, the new bone formation started in the mandible where bone resorption had occurred because of the lesion. The amount of new bone formation was very small in the present case and showed a sequestrum formation without bone trabeculae upon CT imaging. With time, it may have been possible to more clearly observe the cancellous and cortical bone on CT images and the trabecular pattern might have been observable with long-term follow-up. However, as the patient passed away 19 months after proton beam therapy, we were not able to observe such radiographic changes.

We compared the radiographic findings of ORN resulting from proton beam radiotherapy with those of ORN from X-ray radiotherapy. One of the main factors in choosing a form of radiotherapy (X-rays or protons) is the physiological state of the disease. Conventional radiotherapy was not indicated for the present case because the lesion had infiltrated deep into the mandibular bone. Osteoradionecrosis resulting from X-ray radiotherapy typically occurs secondary to the treatment of the primary lesion. In the present case, as the primary lesion had extensively infiltrated the mandibular bone, it was assumed that the mandibular bone was concurrently absorbed with the reduction of the lesion resulting from proton beam radiotherapy. Future investigations should consider the radiation-induced changes to bone separately whether or not the primary lesion exists in the bone.

**CONCLUSION**

Most lesions in the bone do not qualify for curative radiotherapy with X-rays. Typically, radiotherapy is used as a palliative treatment for pain relief or reduction of paralysis risk in patients with bone metastasis. Therefore, it is difficult to predict if the bone absorbed because of the presence of a cancerous lesion will or will not experience new bone formation in the area of the destructed lesion. With the advent of proton beam radiotherapy, the number of diseases which can be curatively treated has increased. As more patients with cancers in the jaw bone are treated with protons, careful observation of any radiation-induced effects such as osteoradionecrosis need to be monitored, classified, and disseminated appropriately.

**Table 1: Comparison of the radiographic findings between proton and an X-ray radiotherapy case**

<table>
<thead>
<tr>
<th></th>
<th>Widening periodontal space</th>
<th>Progression of sclerotic change</th>
<th>Progression of osteolytic change</th>
<th>Sequestrum formation</th>
<th>Periosteal reaction</th>
<th>Pathological fracture</th>
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<tr>
<td>Our case (Proton)</td>
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<td>Suei (X-ray)</td>
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<td>33%</td>
<td>100%</td>
<td>81%</td>
<td>6%</td>
<td>36%</td>
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**Author Contributions**

Masaru Konishi – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Yoshikazu Suei – Acquisition of data, Drafting the article, Final approval of the version to be published

Minoru Fujita – Analysis and interpretation of data, Drafting the article, Final approval of the version to be published

Keiji Tanimoto – Analysis and interpretation of data, Drafting the article, Final approval of the version to be published

**Guarantor of Submission**
The corresponding author is the guarantor of submission.

**Source of Support**
None

**Conflict of Interest**
Authors declare no conflict of interest.

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REFERENCES

Supraclavicular course of the left cephalic vein: Rare anatomical variant

Radha Krishna Shetty Kommanda, Muhammed Asif, Shivarama C.H.

ABSTRACT

Introduction: The cephalic vein is formed by dorsal venous arch on the radial side of the upper extremity. Its course was normal till deltopectoral groove. It then communicates with the subclavian vein by crossing superficial to clavicle instead of emptying into the axillary vein. Case Report: The rare variant of supraclavicular course of subclavian vein was observed in routine upper limb dissection. This case report presents variation in the course of left cephalic vein and its abnormal communication with the subclavian vein. Conclusion: Although the supraclavicular course of the cephalic vein is a rare anatomical variant, the knowledge of variations in the origin, course and termination of cephalic vein is important for surgeons, radiologists, and plastic surgeons. It is a preferred vein for suitable central venous access, pacemaker and defibrillator implantation. Further, the normal anatomy of cephalic vein, including its anatomical variation is paramount to avoid complications.

Keywords: Pacemaker, Subclavian vein, Supraclavicular cephalic vein

INTRODUCTION

Dorsal venous arch is formed by longitudinally oriented dorsal and palmar digital veins, which empty into the dorsal metacarpal veins [1, 2]. Radial continuation of the dorsal venous arch forms cephalic vein [3, 4]. At the wrist, the cephalic vein crosses superficial to the anatomical snuff box. It travels upward along the anterior border of the brachioradialis muscle in the forearm. At the level of the elbow in 84% of cases cephalic vein anastomosis with the median cubital vein, which is the tributary of basilic vein [5, 6]. Leaving the antecubital fossa, it ascends in a groove along the lateral border of the biceps brachii till lower two-thirds of the arm, and then it passes in the deltopectoral groove [5]. At the lateral aspect of the deltopectoral groove, the cephalic vein is located superficially in a pad of fat between two muscles [7]. It further courses in the deltopectoral triangle it pierces the clavpectoral fascia and terminates in the axillary vein. The subclavian, femoral, brachiocephalic and cephalic veins are most commonly used for central venous access at the bedside [8]. Further, cephalic vein is suitable for pacemaker and defibrillator implantation, and reported to have a lower incidence of complications than subclavian puncture [9]. Therefore, correct anatomical knowledge of the cephalic vein is of critical importance.
when considering emergency procedures [6]. In this case report, we described anatomical variation of the cephalic vein, where it has joined with the subclavian vein instead of axillary vein by passing over the clavicle.

**CASE REPORT**

The present study, observation was made during routine dissection for undergraduate students in Department of Anatomy, Yenepoya Medical College, Karnataka, India. The venous variation of left cephalic vein was noticed on the left upper extremity in a male cadaver aged 55 years. The left cephalic vein instead of draining into the axillary vein, passes superficial to the clavicle piercing the investing layer of deep cervical fascia of the neck and opens into the subclavian vein (Figure 1). The course of the right cephalic vein on the right extremity was normal (Figure 2).

**DISCUSSION**

Anatomical variations in the course of the cephalic vein is already documented, but the clinical cases reported in medical literature are rare, and include cases with absence or small diameter of the cephalic vein, accessory veins running parallel to the cephalic vein or even preclavicular or supraclavicular anomalous courses [10–15]. The knowledge of the normal venous anatomy of cephalic vein, including its anatomical variation, is paramount to avoid complications. If cephalic vein is used as an access during permanent lead placement the knowledge of the supraclavicular course of the cephalic vein would help to reduce complications related to lead dysfunction, erosion or collateral vascular damage [16]. Some of the clinicians prefer the cephalic cut-down method rather than subclavian or the internal jugular vein puncture for implant a device to venous access for transvenous placement of pacemaker or defibrillator which may avoid risk of pneumothorax, subclavian crush, and other possible complications as well [17]. The cephalic vein may terminate at the internal jugular vein, the external jugular vein, or the basilic vein [18, 19]. Lau et al. reported a supraclavicular course of the cephalic vein. In this case report, the cephalic vein drained into the subclavian vein with an unusual supraclavicular course.

Further, it is important to know about the communication between the cephalic vein and subclavian vein across the clavicle because in case of fracture of clavicle the cephalic vein may bleed profusely and during catheterization it may get punctured to the overlying skin, and structures on the pectoral area leading to damage. Moreover, if guide wire punctures the subclavian vein, there is a risk of damage to vital structures in the supraclavicular fossa [20]. Therefore, the knowledge of the supraclavicular course of the cephalic vein would help to reduce iatrogenic complications such as erosion or collateral vascular damage.
CONCLUSION

Cephalic vein with supraclavicular course is an rare anatomical variant and seen in 0.2% of individuals. The knowledge of cephalic vein variation is of top importance, considering its role in central venous access, pacemaker and defibrillator implantation. The supraclavicular course of the cephalic vein would help to reduce iatrogenic complications such as erosion or collateral vascular damage, avoid profuse bleeding in the fracture of clavicle and puncture of the guide wire to skin.

*********

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Guarantor of Submission
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Source of Support
None

Conflict of Interest
Authors declare no conflict of interest.

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REFERENCES
Acute guttate psoriasis and psoriatic arthritis simultaneously in a 33-year-old male with streptococcal infection

Jianfeng Zheng, Yangfeng Ding

ABSTRACT

Psoriasis is a chronic, inflammatory, immune-mediated skin disease. There are several clinical phenotypes at disease onset, such as guttate, plaque, erythrodermic, pustular phenotypes and psoriatic arthritis. In this report, we describe a 33-year-old Chinese male diagnosed with acute guttate psoriasis who had concurrent psoriatic arthritis. The patient had non-pruritic erythematous guttate scaly papules on his trunk and proximal extremities for two weeks and swelling and pain in the fourth metacarpophalangeal joint of the right hand for four weeks on admission. A diagnosis of guttate psoriasis was quickly made according to the typical eruptions and positive Auspitz sign. And then joint ultrasound revealed that there was tenosynovitis in the fourth metacarpophalangeal joint of the right hand, consistent with the performance of psoriatic arthritis. So a diagnosis of psoriatic arthritis was also made according to the ultrasonic results and a history of psoriasis. At last, the patient had a history of the chronic tonsillitis and antistreptolysin O titer was 797.1 IU/ml at admission. Streptococcal infection was considered as the major causative factor for this patient with guttate psoriasis and psoriatic arthritis, although this was a rare report of acute guttate psoriasis and arthritis psoriasis simultaneously appeared in a patient after the chronic tonsillitis attacked.

Keywords: Guttate psoriasis, Psoriasis, Psoriatic arthritis, Streptococcal infection

How to cite this article


Article ID: Z01201712CR10864JZ

doi:10.5348/ijcri-2017125-CR-10864

INTRODUCTION

Psoriasis is a chronic, proliferative, and inflammatory skin disease, characterized by increased propagation of epidermis with dermal capillaries dilation. There are various types of psoriasis identified by clinical outcomes. Psoriasis is often first detected between the age of 15 and 25 years, and psoriasis arthritis usually develops between the...
age of 30–50 years. Guttate psoriasis is a type of psoriasis in which erythematous guttate scaly papules appear all over the body [1]. Among these patients, 56–100% has recent precedent evidence of streptococcal disease such as the tonsillitis [2]. Psoriasis arthritis is a type of psoriasis in which joints become red, swollen, tender, warm, and stiff under the infiltration of inflammation. Its causative agents include trauma, allergies of medicines, alcohol consumption, skin irritants, and smoking [3]. Here, we present a case of acute guttate psoriasis and arthritis psoriasis simultaneously appeared in a 33-year-old male with a 15-year history of plaque psoriasis after the chronic tonsillitis attacked.

**CASE REPORT**

A 33-year-old male presented with one-month history of swelling and tender in the fourth metacarpophalangeal joint of the right hand and a two-week history of non-pruritic erythematous guttate scaly papules on his trunk and extremities. He denied any recent travel, drugs or medications, environmental changes. However, the patient stated that there was a sore throat, swollen tonsils, fever, congestion, malaise, and fatigue about one month and a half before admission to our hospital and a 20-year history of the chronic tonsillitis. These symptoms were not actively treated because of his busy job. The fourth metacarpophalangeal joint of the right hand suddenly become red, swollen, tender about one month before admission. But the patient did not go to hospital in time. And then some round, erythematous, hyperkeratotic, flat papules with adherent scale began appearing on his trunk about two weeks before admission and had spread to his extremities on the day that he was admitted. He also reported a personal history of chronic plaque psoriasis over 15 years ago which, when the chronic tonsillitis attacked, would relapse. In the last six months, there were only few patches on his extension side of bilateral calves under conditions of actively treatment of psoriasis and preventing the recurrence of tonsillitis.

On physical examination, the author noted 1–10 mm round, erythematous, hyperkeratotic, flat papules with adherent scale on his trunk and bilateral upper extremities and thighs, Auspitz sign (+). Patches on his extension side of bilateral calves and mildly swollen in the fourth metacarpophalangeal joint of the right hand were checked simultaneously, tenderness (+). His mucous membranes, however, were not involved. In addition, there was moderate swelling on the tonsil (Figure 1).

Laboratory data disclosed the following value: white blood cell count 8690/mm³, hemoglobin 16 g/dl, neutrophil absolute 6930/mm³, lymphocyte absolute value 1040/mm³, monocyte absolute value 590/mm³, eosinophil absolute value 110/mm³, basophil absolute value of 20/mm³. Thyrotropin (TSH) 0.783 mIU/l, free triiodothyronine (TT3) 1.3 nmol/l, free thyroxine determination (FT4) 16.86 pmol/l, free triiodothyronine (FT3) 4.28 pmol/l. Alanine aminotransferase 25U/l, γ-glutamyltransferase 39 U/l, aspartate aminotransferase 25U/l, urea 3.50 mmol/l, creatinine 89 umol/l, uric acid 429 umol/l, blood glucose 5.27 mmol/l, triglyceride 2.85 mmol/l, total cholesterol 4.98 mmol/l, high sensitivity CRP 2.20 mg/l, rheumatoid factor 10.0IU/ml, antistreptolysin O 797.1 IU/ml. B cell and T cell and subpopulations: CD3+ cells 79.2%, CD4+ cells 56.3%, CD8+ cells 17.2%, CD4+/CD8+ ratio 3.27; natural killer (NK) cells 1.62%. Ultrasound revealed that there was tenosynovitis in the fourth metacarpophalangeal joint of the right hand, consistent with the performance of psoriatic arthritis (Figure 2).

**DISCUSSION**

Fehleisen first isolated β-hemolytic streptococci in 1883. Lancefield’s classification of β-hemolytic streptococci according to their carbohydrate surface antigens has revealed that group A β-hemolytic streptococci are the most pathogenic to humans [4]. And it was not until 1916 that the association streptococcal tonsillitis and guttate psoriasis was first reported [5]. Much work has involved monitoring titres of antibodies to the streptolysin O exotoxin (ASO titres) as these provide an indication of recent streptococcal infection.

In 1952, Norholm-Pedersen divided a group of 133 unselected patients with psoriasis into three groups depending on their ASO titres and found the highest proportion of patients with guttate psoriasis in the highest ASO group [6]. The close relationship between guttate psoriasis and streptococcal infection has lead some investigators to examine a possible role for...
The presence of streptococcal antigens has not been demonstrated in the joints of psoriasis arthritis patients. Therefore, it is difficult to prove a pathogenic role for such antigens, as secondary infection of psoriatic plaques is common. Some reports have suggested that HIV and hepatitis C virus may play a more significant role in the pathogenesis of psoriasis arthritis [7, 8]. Both of acute guttate psoriasis and psoriatic arthritis are the common phenotypes of psoriasis. But they have obviously different characterizes. Rare reports had revealed that the patients with arthritis were found among the non-guttate patients [9]. In this case, there is no direct evidence to support a role for such antigens in the pathogenesis of psoriasis arthritis. However, there is a distinct tendency for improvement in the symptoms of arthritis after treatment of the tonsillitis.

In an earlier paper, it was described how acute eruption of psoriasis may be produced in phases of immune deficiency and in the presence of bacterial antigen-releasing inflammatory foci, whereas clinical spontaneous remissions are produced in phases of immunologic activity. In this case, the patient had significantly elevated antistreptolysin titer, as well as proliferative responses to streptococcal antigens. But serum immunoglobulin levels (IgM, IgG, IgA) was in normal range. It revealed that our patient was clinically active with eruptions. Furthermore, there is a rise in CD4+ cells and a reduction in CD8+ and natural killer cells in our report, CD4+/CD8+ ratio was considerably increased. Systemic defense essentially depends on the activity of CD8+ and natural killer cells. Animal experiments have shown that natural killer cells play a decisive role in the response to microbial infections as part of nonspecific cellular defense [10]. These results are consistent with immune deficiency. So the dermatosis has been regarded as a T cell mediated autoimmune disease.

CONCLUSION

In this case, we present a rare case of acute guttate psoriasis and psoriasis arthritis simultaneously appeared in a patient with a 20-year history of the chronic tonsillitis. The typical eruptions, the ultrasonic results and a history of psoriasis were helpful to make this diagnosis. Clinicians should be aware of a certain relationship between psoriasis arthritis and streptococcal infection and make more examinations to discover the pathogenesis of psoriasis.

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Author Contributions
Jianfeng Zheng – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Yangfeng Ding – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor of Submission
The corresponding author is the guarantor of submission.

Source of Support
None

Conflict of Interest
Authors declare no conflict of interest.

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REFERENCES


SUGGESTED READING

Takayasu arteritis: A rare presentation as pulseless disease of lower limb with middle aortic syndrome

Harshita Sharma, Sailesh Kumar Bansiwal, Rajesh Manocha, Prabal Rajvanshi, Kathuria Paras

ABSTRACT
Takayasu arteritis also known as pulseless disease is an inflammatory and stenotic disease of medium and large sized arteries characterized by a strong predilection for the aortic arch and its branches. However, it is rarely reported as a cause of middle aortic syndrome. Middle aortic syndrome is characterized by localized or extended narrowing of the descending thoracic or abdominal aorta. Hypertension proximal to the aortic stenosis and relative hypotension distal to it, are characteristic findings in middle aortic syndrome. We present a case of 24-year-old hypertensive male, who presented with abdominal pain, decreased urine output and lower limb claudication, found to have involvement of abdominal aorta, visceral, renal and lower limb arteries on imaging. Though it is a rare cause of hypertension, a high index of suspicion is necessary for diagnosis and early treatment.

Keywords: Hypertension, Middle aortic syndrome, Takayasu arteritis

INTRODUCTION
Takayasu arteritis is a rare chronic inflammatory vasculitis that primarily affects young females in 2nd and 3rd decade. It occurs worldwide with wide geographical variation, mainly seen in Japan, South East Asia, India and Mexico [1]. Takayasu arteritis has an annual incidence of 0.8 per million and prevalence of 4.7 per million. It is characterized by panarteritis with inflammatory cells in vessel wall, leading to stenosis of lumen with or without thrombosis. It usually affects subclavian artery (93%), common carotid (58%), abdominal aorta (47%), renal artery (38%) and other visceral arteries. Carotid intimal thickness, C-reactive protein, and elevated ESR are important markers for disease activity, which are followed-up in Takayasu arteritis [2].

Middle aortic syndrome, also known as coarctation of abdominal aorta, is rarely reported with Takayasu arteritis. It may be congenital, with incomplete fusion or over fusion of the paired embryonic aortas, or acquired, such as with Takayasu or giant cell arteritis, neurofibromatosis, or retroperitoneal fibrosis [3]. This...
condition can also involve renal and other visceral arteries leading to hypertension and other features.

American College of Rheumatology criteria (ACR), imaging and other investigations aid the diagnosis. Medical therapy is given to control hypertension and halt the progress of disease. Surgery is required to revert the hypertension and its complications, if not controlled by medical therapy.

CASE REPORT

A 24-year-old male presented to us with a history of abdominal pain in right lumbar and iliac area, decreased urine output and lower limb claudication for one and a half month. The patient denied any history of fever, vomiting, shortness of breath, chest pain, hematuria or visual disturbances. Inquiry about joint pain, photosensitivity, oral ulcers and recurrent infections were not contributory. The past history was unremarkable.

On examination the patient was conscious and oriented. His blood pressure was not recordable in both lower limbs while bilateral upper limb showed reading of 200/100 mmHg. Pulses of upper limb were regular and did not show any radio radial delay while in lower limbs pulses were not palpable. A head-to-toe examination was unrewarding. A bruit was heard over the infraumbilical region while the examination of other systems was unrewarding. Fundus examination revealed grade two hypertensive changes.

Laboratory data showed hemoglobin 11.8 g/dL (13–16 g/dL) and total leukocyte count 6,680 cells (4000–11000 cells/µl) with 68% neutrophils. Other baseline biochemical investigations were within normal limits including kidney function test, total cholesterol, serum triglycerides, serum HDL levels, 24 hour urinary protein and urine routine microscopy. Anti-nuclear antibody (ANA) test was positive with high titers (1:160) while anti-dsDNA, anti-phospholipid antibody (APLA), protein C, protein S, factor V Leiden, anti-thrombin, homocysteine levels were normal. HIV was non-reactive. High-sensitivity C-reactive protein with value >3 mg/L (low risk <1 mg/L) and erythrocyte sedimentation rate (ESR) with value 98 mm/hr (0–20 mm/hr) were significantly raised while Mantoux test was insignificant.

On chest radiography, no significant abnormality was detected while ECG and echocardiography showed left ventricular hypertrophy (LVH).

Ultrasoundography of abdomen with renal Doppler was done, which revealed shrunken left kidney (size 5x4 cm) and normal sized right kidney. Right renal artery and interlobar arteries demonstrated normal spectral wave forms but left intrarenal flow was decreased. There was large atheroma filled in abdominal aorta distal to superior mesenteric artery (SMA), flow in both renal arteries was showing turbulence at the site of origin of arteries which may be due to atheroma filled in aorta.

Computed tomography angiography of thoracic aorta revealed normal aortic root, ascending aorta, arch of aorta with its branches and descending thoracic aorta (Figure 1) while computed tomography angiography of abdominal aorta corroborated the findings on ultrasonography and showed thrombosis, stenosis, calcification of juxta renal, infrarenal aorta up to aortic bifurcation; complete occlusion of celiac, superior mesenteric, inferior mesenteric, left renal arteries with severe stenosis of right renal artery at origin; reformation of distal visceral arteries (collaterals) and both common iliac arteries through intercostal, lumbar, superior, inferior, epigastric and circumflex arteries (Figure 2). For further evaluation, computed tomography angiography of bilateral lower limb was done which demonstrated stenosis of right anterior and posterior tibial arteries in distal third of leg with stenosis of peroneal artery in distal half, delayed opacification of distal anterior and posterior tibial arteries on left side, suggestive of sluggish distal flow (Figure 3).

Using American College of Rheumatology criteria 1990, patient was diagnosed with type IV Takayasu Arteritis (4 out of 6 criteria were met) with lower limb involvement.

The patient was put on anti-hypertensive: prazosin (5 mg PO 12 hr), amlodipine (5 mg PO 24 hr), clonidine (0.25 mg PO 8 hr) and methyl prednisolone 250 mg/day for 5 days, tapered to maintenance dose of 7.5 mg/day. He received atorvastatin (20 mg PO 24 hr) and anti-coagulation with LMW heparin (6 mg SC q 12 hr) and warfarin (5 mg PO 24 hr) and INR was monitored.
Type III and type IV are also known as middle aortic syndrome (Table 2) [5]. Middle aortic syndrome, either of congenital or acquired etiology, is a rare and important cause of hypertension in young. Hypertension is the cardinal clinical feature in middle aortic syndrome and is present in more than 90% of cases, weak or absent femoral pulses may be appreciated and an audible bruit are typically heard over aorta [6]. The most common anatomic form in middle aortic syndrome of either etiology is interrenal (19–52%), followed by suprarenal (11–40%), infrarenal (19–25%) and diffuse (12%) [3]. Stenosis of the renal arteries is common (60–90%), with less involvement of the celiac and superior mesenteric arteries (20–40%), and infrequent involvement of the inferior mesenteric arteries [3, 7, 8].

High titre of anti-nuclear antibody was attributable to autoimmune mechanism affecting large vessels, which responded to immunosuppressive agents. After excluding other connective tissue disorders affecting large vessels and hypercoagulable states, clinical and radiological assessment aided the diagnosis. Hence, a diagnosis of middle aortic syndrome of Takayasu arteritis origin causing malignant hypertension in a young male patient was made. The rarity of disease has led to paucity of data in literature.

The mainstay of treatment is to decrease inflammation with immunosuppressive drugs and to control hypertension with anti-hypertensive drugs. Surgery is subsequently. Patient responded well to the treatment and has been on irregular follow-up. The patient was referred to vascular surgery department for further management.

DISCUSSION

Takayasu arteritis is a rare, systemic auto-inflammatory disease of young females which usually involves aortic arch and its branches. It affects the vessel walls leading to stenosis or thrombosis of vessels and thus hampers the blood supply of the concerned organs. It can result in weak pulses or loss of pulse in arms, legs and organs. For this reason it is referred as pulseless disease. Takayasu arteritis can have a spectrum of presentation ranging from being asymptomatic to a catastrophic disease presenting as malignant hypertension. As symptoms are non-specific and the disease is so rare that there is often a delay in detecting it.

Tuberculosis has remained an important differential diagnosis which has been ruled out in our case. American College of Rheumatology 1990 criteria (Table 1) is the most widely accepted criteria for diagnosis of Takayasu arteritis which included arteriographic abnormality, best seen on angiography (Table 2) [4]. Type III and type IV are also known as middle aortic syndrome (Table 2) [5].

Middle aortic syndrome, either of congenital or acquired etiology, is a rare and important cause of hypertension in young. Hypertension is the cardinal clinical feature in middle aortic syndrome and is present in more than 90% of cases, weak or absent femoral pulses may be appreciated and an audible bruit are typically heard over aorta [6]. The most common anatomic form in middle aortic syndrome of either etiology is interrenal (19–52%), followed by suprarenal (11–40%), infrarenal (19–25%) and diffuse (12%) [3]. Stenosis of the renal arteries is common (60–90%), with less involvement of the celiac and superior mesenteric arteries (20–40%), and infrequent involvement of the inferior mesenteric arteries [3, 7, 8].

Table 1: American College of Rheumatology criteria for classification of Takayasu’s arteritis [4] (A diagnosis of Takayasu’s arteritis requires that at least three of the six criteria are met)

<table>
<thead>
<tr>
<th>Sr. No.</th>
<th>Criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>Age at disease onset ≤ 40 years</td>
</tr>
<tr>
<td>2.</td>
<td>Claudication of extremities</td>
</tr>
<tr>
<td>3.</td>
<td>Decreased brachial artery pulse</td>
</tr>
<tr>
<td>4.</td>
<td>Blood pressure difference &gt;10 mmHg</td>
</tr>
<tr>
<td>5.</td>
<td>Bruit over subclavian arteries or aorta</td>
</tr>
<tr>
<td>6.</td>
<td>Arteriogram abnormality</td>
</tr>
</tbody>
</table>

Table 2: Angiographic classification of Takayasu’s arteritis (Takayasu conference, 1994) [5] (Involvement of the coronary or pulmonary arteries should be designated as C (+) or P (+) respectively)

<table>
<thead>
<tr>
<th>Type</th>
<th>Vessel involvement</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type I</td>
<td>Branches from the aortic arch</td>
</tr>
<tr>
<td>Type IIa</td>
<td>Ascending aorta, aortic arch, and its branches</td>
</tr>
<tr>
<td>Type IIb</td>
<td>Ascending aorta, aortic arch and its branches, thoracic descending aorta</td>
</tr>
<tr>
<td>Type III</td>
<td>Thoracic descending aorta, abdominal aorta, and/or renal arteries</td>
</tr>
<tr>
<td>Type IV</td>
<td>Abdominal aorta and/or renal arteries</td>
</tr>
<tr>
<td>Type V</td>
<td>Combined features of type IIb and IV</td>
</tr>
</tbody>
</table>

Figure 2: Computed tomography angiography abdominal aorta showing non visualization of left kidney and multiple collateral vessels.

Figure 3: Computed tomography angiography abdominal aorta and lower limbs showing non visualization of left kidney, collaterals between abdominal aorta and common iliac artery with sluggish flow in right and left anterior and posterior tibial arteries.
required if there is uncontrolled hypertension due to renal artery stenosis, extremity claudication limiting activities of daily life, cerebrovascular ischemia, moderate aortic regurgitation and ischemia due to coronary artery involvement. Our patient responded well to high dose steroids along with anti-hypertensive drugs. If left untreated such patients die by age of 35 years [3, 7].

Rare presentation of pulseless lower limbs in a young hypertensive male attributing to middle aortic syndrome of Takayasu arteritis origin are being highlighted in this case study.

**CONCLUSION**

Takayasu arteritis is a chronic systemic autoimmune inflammatory disease of young females which usually involves large vessels, most commonly the aorta. Middle aortic syndrome of Takayasu arteritis origin, which is characterized by coarctation of distal descending thoracic and abdominal aorta, is rarely reported as a cause of hypertension in young. Clinical examination and radiology play an important role in its diagnosis. There is a need to have a thorough workup of a young hypertensive patient, especially to differentiate congenital and acquired causes, as the course of treatment varies accordingly. A high index of suspicion is necessary for diagnosis and early treatment as the mortality is high, if left untreated.

********

**Author Contributions**

Harshita Sharma – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Sailesh Kumar Bansiwal – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

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**Guarantor of Submission**
The corresponding author is the guarantor of submission.

**Source of Support**
None

**Conflict of Interest**
Authors declare no conflict of interest.

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**REFERENCES**

A 68-year-old female with probable multiple system atrophy

Robert T. Kidnie, Jonathan Mowrey, David R. Mantilla, Ryan D. Nicklas, Ramaswamy Rangarajan, Gregor K. Wenning

ABSTRACT

Introduction: Multiple system atrophy (MSA) is a rare, progressive neurodegenerative disease that encompasses elements of cerebellar abnormalities, parkinsonism and autonomic dysfunction. Autonomic dysfunction classically manifests as orthostatic hypotension and is present in all forms of MSA. While MSA can only be definitively diagnosed post-mortem, a probable diagnosis is obtained clinically. There is no cure for MSA and patients are managed symptomatically. Different symptoms vary greatly in their response to pharmacotherapy, which makes management a challenge. Case Report: We present the case of a 68-year-old female with probable MSA. The patient first presented to the emergency department of a community hospital complaining of dizziness when standing from a supine or seated position. On questioning it was learned that she was being followed at another hospital for possible MSA. Her orthostatic symptoms proved refractory to treatment with midodrine, so she was eventually started on fludrocortisone; this greatly reduced her symptoms. Early in her hospital stay, our patient also began experiencing urinary retention. This was effectively managed with catheterization; however, our patient’s hospital stay was prolonged due to a urinary tract infection and physical deconditioning. Conclusion: This case illustrates some of the many challenges associated with both diagnosing and managing MSA. We wish to reinforce the high-level of clinical suspicion required to diagnose MSA and the therapeutic resilience and pharmacologic versatility necessary to manage symptoms.

Keywords: Autonomic dysfunction, Multiple system atrophy, Orthostatic hypotension, Shy-Drager syndrome

INTRODUCTION

We present the case of a 68-year-old female who is being followed at the ataxia clinic of another institution for progressive cerebellar ataxia and a constellation of other neurologic findings that have progressed over the previous ten years. The working diagnosis at that institution is multiple system atrophy; however, until her presentation at the emergency department, she had not experienced any symptomatic orthostatic hypotension.
Current expert opinion posits that although definitive diagnosis of MSA requires the presence of alpha synuclein accumulation in oligodendroglial cells seen on autopsy, a probable diagnosis requires autonomic failure and poorly levodopa-responsive parkinsonism or cerebellar ataxia [1]. Highlighted in this case, then, is the potential challenge associated with diagnosing MSA. Although some elements of MSA such as motor symptoms are resistant to pharmacological therapy, other elements may be managed medically [2]. Our patient’s presenting complaint was orthostatic hypotension, and studies have shown that certain medications do have a role in ameliorating this symptom [3]. However, our patient’s hospital stay was complicated by several factors related to MSA and thus also serves to illustrate some of the challenges associated with management of this disease.

CASE REPORT

A 68-year-old African-American female presented to the emergency department complaining of persistent dizziness over the previous three days. She stated this dizziness was most pronounced when she stood up and that due to a change in medication approximately two weeks prior, she had been drinking less than normal. On questioning, it was discovered that she was being followed at a large research institution for progressive cerebellar ataxia for the previous three years. The working diagnosis at that institution was multiple system atrophy, formerly known as Shy-Drager syndrome.

In addition to her progressive cerebellar ataxia, her past medical history was significant for seizures, eye-movement abnormalities, wide-based gait, dysmetria, postural tremor, mild incontinence, and bilateral paresthesia of both upper and lower extremities. These symptoms had been gradually worsening over the past ten years, however, were not evident on presentation in the emergency department. She was also being managed medically for hypothyroidism and diabetes. Her surgical history was significant for a left hemi-colectomy, secondary to colonic volvulus more than fifteen years ago. Her family history was non-contributory. She had no known allergies and had a 20-pack year history of smoking, but quit approximately 27-years prior. At the time of admission, she was living at home with her husband. She underwent CT scans in the emergency department. Computed tomography scans demonstrated mild generalized atrophy and patchy, non-specific periventricular white matter hypoattenuation (Figure 1). There was, however, also some evidence of olivopontocerebellar atrophy (OPCA) seen on midsagittal CT imaging (Figure 2), which is consistent with MSA-C. Following a workup in the emergency department, the patient was admitted to the telemetry floor for management of dehydration and orthostatic hypotension.

On the telemetry floor, she was started on 5 mg midodrine PO TID, but after two days of poor response to the medication, her dose was increased to 10 mg. On hospital day-4, she was transferred to a sub-acute floor so that her medications could be optimized and she could begin physical and occupational therapy secondary to a chronically deconditioned state. Her hospital stay, however, was prolonged due to number of issues. First, her orthostatic hypotension proved resistant to pharmacotherapy. Despite thigh-high compression stockings, a high-salt diet and rising slowly from bed, our patient continued to experience symptomatic orthostatic hypotension even on 10 mg midodrine PO TID. Second, she experienced rebound hypertension due to the midodrine. This was significant enough to require a reduction in the midodrine to 5 mg TID, on hospital day-11. Throughout this period, the patient also experienced urinary retention with post-void residuals occasionally in excess of 500 mL. This prompted Foley catheterization, however, on hospital day-13, the patient was diagnosed with a multi-drug resistant catheter associated urinary tract infection (CAUTI). This spectrum of complications is common amongst patients with MSA [2].

On hospital day-24 the diagnosis of MSA was made and the decision to add fludrocortisone 0.1 mg PO Qday to her treatment followed. Her orthostatic hypotension responded reasonably well to fludrocortisone and the patient’s rehabilitation continued on the sub-acute floor. On hospital day-35 the midodrine was discontinued. Her mean arterial pressure is presented over the course of her stay in Figure 3.

Figure 1: Transverse cranial computed tomography scan of our patient on admission to the emergency department demonstrated mild generalized atrophy and patchy, non-specific periventricular white matter hypoattenuation.
Despite a slow recovery, the patient’s presenting symptoms gradually improved; over the course of weeks, she regained the ability to ambulate independently and experienced minimal dizziness on standing. Her urinary retention remained, while the other pre-existing neurologic conditions did not surface clinically. However, it was decided that she could be managed on an outpatient basis in conjunction with a neurology follow-up. She was subsequently discharged on hospital day-45.

DISCUSSION

This case serves to underscore the potential challenges of diagnosis and the management of a patient with MSA. Multiple system atrophy is a rare condition, with an incidence rate in the United States of approximately 3 per 100,000 individuals over the age of 50 [4]. Multiple system atrophy encompasses several progressive neurodegenerative disorders, including Shy-Drager syndrome, striatoniagral degeneration, and olivopontocerebellar atrophy [5]. In striatoniagral degeneration (termed, MSA-P) parkinsonian symptoms predominate, while in olivopontocerebellar atrophy (termed, MSA-C) cerebellar ataxia predominates. The predominate feature in Shy-Drager Syndrome is autonomic dysfunction and it is expressed to varying degrees in both MSA-P and MSA-C. These three conditions share a common, although incompletely understood etiology of alpha-synuclein accumulation in oligodendroglial cells [6]. Consequently, the manifestations of MSA in a single patient may incorporate elements from all three subtypes. For instance, our patient presented to the emergency department with orthostatic hypotension, a finding classically associated with Shy-Drager; however, her initial clinical manifestation and the symptom which initiated specialist care was ataxia, the classic finding in MSA-C.

Due to the neurodegenerative etiology of MSA, the disease can only be definitively diagnosed on autopsy. However, an expert panel convened in 2007 and produced a second consensus statement on the diagnosis of MSA that concluded probable MSA required a sporadic, progressive adult-onset disorder including rigorously defined autonomic failure and poorly levodopa-responsive parkinsonism or cerebellar ataxia [1]. The same panel published tables for aiding in the diagnosis of probable MSA (Table 1) and possible MSA (Table 2), as well as additional features supporting possible MSA-P and MSA-C specifically (Table 3) [1]. Our patient’s clinical presentation in the emergency department supported a diagnosis of probable MSA-C. Studies have shown that computed tomography (CT) is of limited value in the diagnosis of MSA; in fact, when the diagnosis of MSA can be made clinically, as was the case in our patient, the role of CT imaging is best suited to rule out intracranial pathologies, such as tumors [7]. For this reason, CT scans was ordered for the patient. Due to the non-specific nature of the CT and clinical diagnosis of MSA-C, we determined that no further cranial imaging was warranted.

While diagnosis of multiple system atrophy is a challenge, so too is the disease’s management. No effective disease-modifying or neuroprotective treatment is currently available for MSA [8]. Indeed, treatment is directed at individual symptoms and responses vary greatly. For instance, the motor symptoms associated with MSA-C and the bradykinesia and rigidity associated with MSA-P are both typically resistant to pharmacologic
Table 1: Criteria for the diagnosis of probable multiple system atrophy (MSA) as established by the Second consensus statement on the diagnosis of multiple system atrophy, 2008.

<table>
<thead>
<tr>
<th>Criteria for the diagnosis of probable multiple system atrophy</th>
</tr>
</thead>
<tbody>
<tr>
<td>A sporadic, progressive, adult (&gt;30 years) ~onset disease characterized by</td>
</tr>
<tr>
<td>• Autonomic failure involving urinary incontinence (inability to control the release of urine from the bladder with erectile dysfunction in males) or an orthostatic decrease of blood pressure within three minutes of standing by at least 30 mmHg systolic or 15 mmHg diastolic and</td>
</tr>
<tr>
<td>• Poorly levodopa-responsive parkinsonism (bradykinesia with rigidity, tremor, or postural instability) or</td>
</tr>
<tr>
<td>• A cerebellar syndrome (gait ataxia with cerebellar dysarthria, limb ataxia, or cerebellar oculomotor dysfunction)</td>
</tr>
</tbody>
</table>

Table 2: Criteria for the diagnosis of possible multiple system atrophy as established by the Second consensus statement on the diagnosis of multiple system atrophy, 2008.

<table>
<thead>
<tr>
<th>Criteria for the diagnosis of possible multiple system atrophy</th>
</tr>
</thead>
<tbody>
<tr>
<td>A sporadic, progressive, adult (&gt;30 years) ~onset disease characterized by</td>
</tr>
<tr>
<td>• Parkinsonism (bradykinesia with rigidity, tremor, or postural instability) or</td>
</tr>
<tr>
<td>• A cerebellar syndrome (gait ataxia with cerebellar dysarthria, limb ataxia, or cerebellar oculomotor dysfunction) and</td>
</tr>
<tr>
<td>• At least one feature suggesting autonomic dysfunction (otherwise unexplained urinary urgency, frequency or incomplete bladder emptying, erectile dysfunction in males, or significant orthostatic blood pressure decline that does not meet the level required in probably multiple system atrophy) and</td>
</tr>
<tr>
<td>• At least one of the additional features given in Table 3</td>
</tr>
</tbody>
</table>

Additional features of possible multiple system atrophy as establish by the Second consensus statement on the diagnosis of multiple system atrophy, 2008.

<table>
<thead>
<tr>
<th>Possible MSA-P or MSA-C</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Babinski sign with hyperreflexia</td>
</tr>
<tr>
<td>• Stridor</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Possible MSA-P</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Rapidly progressive parkinsonism</td>
</tr>
<tr>
<td>• Poor response to levodopa</td>
</tr>
<tr>
<td>• Postural instability within 3 y of motor onset</td>
</tr>
<tr>
<td>• Gait ataxia, cerebellar dysarthria, limb ataxia, or cerebellar oculomotor dysfunction</td>
</tr>
<tr>
<td>• Dysphagia within 5 y of motor onset</td>
</tr>
<tr>
<td>• Atrophy of magnetic resonance imaging scan of putamen, middle cerebellar peduncle, pons or cerebellum</td>
</tr>
<tr>
<td>• Hypometabolism on FDG-PET in putamen, brainstem, or cerebellum</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Possible MSA-C</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Parkinsonism (bradykinesia and rigidity)</td>
</tr>
<tr>
<td>• Atrophy on magnetic resonance imaging scan of putamen, middle cerebellar peduncle, or pons</td>
</tr>
<tr>
<td>• Hypometabolism on FDG-PET in putamen</td>
</tr>
<tr>
<td>• Presynaptic nigrostriatal dopaminergic denervation on SPECT or PET</td>
</tr>
</tbody>
</table>

Abbreviations: MSA: Multiple System Atrophy, MSA-P: MSA with predominant parkinsonism, MSA-C: MSA with predominant cerebellar ataxia, FDG: [18F] fluorodeoxyglucose, PET: Positron Emission Tomography, SPECT: Single-Photon Emission Computed Tomography

Current guidelines for the treatment of orthostatic hypotension in MSA recommend the corticosteroid volume expander fluocortisone (brand name Florinef®, manufactured by Bristol-Myers Squibb Company, Princeton, NJ 08540 USA) and the α1-receptor agonist midodrine. The US Food and Drug Administration also recently approved the synthetic amino acid precursor Droxidopa (Lundbeck Inc. 6 Parkway North, Deerfield, IL 60015) for treatment of these symptoms [13]. This orally administered medication is converted to norepinephrine in the body. The enzyme aromatic amino acid decarboxylase is responsible for this conversion and is so ubiquitous that systemic norepinephrine levels increase even in the absence of failing postganglionic sympathetic neurons [16]. We use the case of our patient’s presenting complaint and the subsequent challenges achieving adequate orthostatic blood pressure control to illustrate MSA’s variable response to pharmacotherapy. Despite IV rehydration in the emergency department, our patient continued to suffer from orthostatic hypotension. Once on the telemetry floor, 5 mg midodrine PO TID was started. Midodrine represents a first-line treatment [7]. Although there are clinical trials in phases II and III currently underway, current management of these symptoms focus on physical and occupational therapy. Modern technology may be adapted to facilitate some activities of daily living. For instance, to address our patient’s dysgraphia secondary to ataxia, we took advantage of her smartphone’s voice-to-text features to make lists and encouraged her to practice typing, which she was still able to do reasonably well. Multiple system atrophy is still progressive and even these solutions will eventually prove inadequate without medical intervention.

Even without a cure, and despite resistance to pharmacotherapy, some symptoms are amenable to medical therapy. The characteristic orthostatic hypotension common to both MSA-P and MSA-C is a result of autonomic dysfunction and unlike the motor symptoms, may respond to pharmacotherapy [9].
pharmacologic therapy in the treatment of orthostatic hypotension [11]. Our patient's average supine to standing orthostatic drop in systolic blood pressure was 58 mmHg and her drop in diastolic blood pressure was 25 mmHg during this period and she remained symptomatic. Given these findings, the decision was made to increase the midodrine dose from 5 mg to the maximum recommend 10 mg TID. Although she initially responded with significant rebound hypertension (169/94 mmHg one day after increasing the midodrine dose), over the following 23 days, the average recorded orthostatic systolic blood pressure drop was 34 mmHg and the average diastolic blood pressure drop is 16 mmHg. This was achieved by titrating the dose of midodrine down from 10 mg to 5 mg again and eventually up to 7.5 mg. The patient’s symptoms improved, but she still complained of occasional dizziness. After the diagnosis of MSA was made, the patient was started on fludrocortisone 0.1 mg PO Qday. Fludrocortisone is another first line therapy used in the treatment of orthostatic hypotension, particularly in cases associated with MSA [12]. In our patient, the average orthostatic systolic and diastolic blood pressure drops were smaller on fludrocortisone: 19 mmHg and 5 mmHg, respectively. These values are presented in Table 4. Of note, the patient also reported ‘much less dizziness’ after starting fludrocortisone and tolerated increasing amounts of physiotherapy. Given our patient’s positive response to fludrocortisone and the much higher cost of droxidopa, the recently FDA approved medication was not considered necessary in this patient’s medical management.

Notwithstanding the challenges of obtaining effective pharmacotherapy to treat orthostatic hypotension in MSA, non-drug therapies have also been shown to improve symptoms. These therapies include abdominal binders, compression stockings, increased salt intake, and taking care to rise slowly from a horizontal or seated position [2]. The latter three therapies were implemented throughout the patient’s hospitalization.

Table 4: Average orthostatic systolic and diastolic decreases in blood pressure during our patient’s hospital stay and in response to various pharmacologic therapies

<table>
<thead>
<tr>
<th>Medication</th>
<th>Systolic blood pressure orthostatic decrease in mmHg (Supine to Standing)</th>
<th>Diastolic blood pressure orthostatic decrease in mmHg (Supine to Standing)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Initial midodrine dose</td>
<td>58</td>
<td>25</td>
</tr>
<tr>
<td>Day 0–2</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Titrating midodrine dose Day 3–26</td>
<td>33</td>
<td>16</td>
</tr>
<tr>
<td>0.1 mg fludrocortisone Day 27–45</td>
<td>19</td>
<td>5</td>
</tr>
</tbody>
</table>

CONCLUSION

Multiple system atrophy (MSA) is a rare and progressive neurodegenerative disorder that encompasses elements of three subtypes: MSA-P with parkinsonian symptoms predominating, MSA-C with cerebellar symptoms predominating and Shy-Drager, which classically presents with autonomic dysfunction and is present in varying degrees in both MSA-P and MSA-C. There is no cure for MSA and at present, it can only be diagnosed definitively at autopsy. Instead, a probable diagnosis is typically made clinically. Current medical management of the disease is aimed at improving symptoms; although some symptoms, such as the motor symptoms of MSA-C and MSA-P, are generally resistant to medical therapy. Although the patient presented to the emergency department with a working diagnosis of MSA from another institution, her case illustrates the challenges of making a diagnosis of MSA, as well as managing the associated orthostatic hypotension. It also highlighted some of the potential complications of prolonged MSA-associated hospitalization, including physical deconditioning and catheter associated urinary tract infections. We, therefore, wish to reiterate that a high-level of clinical suspicion is required to diagnose MSA and that therapeutic resilience along with pharmacologic versatility are key to symptomatic management.

**********

Author Contributions

Robert T. Kidnie – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Jonathan Mowrey – Substantial contributions to conception and design, Revising it critically for important intellectual content, Final approval of the version to be published

David R. Mantilla – Substantial contributions to conception and design, Analysis of data, Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published

Ramanwamy Rangarajan – Substantial contributions to conception and design, Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published

Gregor K. Wenning – Substantial contributions to conception and design, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor of Submission

The corresponding author is the guarantor of submission.

International Journal of Case Reports and Images, Vol. 8 No. 12, December 2017. ISSN: 0976-3198.
REFERENCES


A case of sudden cardiac arrest in a young adult
Zaid B. Al Jebaje, John Elibol, Robbie Wall, Osman Saleem

ABSTRACT

Introduction: Arrhythmogenic right ventricular cardiomyopathy (ARVC) is an inherited myocardial condition that primarily affects the right ventricle. The hallmark characteristic of this disease is continual loss and replacement of normal myocardium with fibrofatty tissue. This replacement of tissue can lead to life-threatening arrhythmias and potentially sudden cardiac death (SCD). Current diagnostic modalities include, electrocardiography, family history, echocardiogram, MRI scan, angiography and myocardial biopsy. Case Report: A 27-year-old athletic female with no known past medical history collapsed while playing frisbee in the park. Upon emergency medical service (EMS) arrival, the patient was unconscious, pulseless, and in ventricular fibrillation. After a successful resuscitation, the patient was transferred to the emergency department and admitted to the ICU. Electrocardiography revealed a QT interval of 460 milliseconds and T-wave inversion in V1, V2, and V3. Transthoracic echocardiogram revealed a left ventricular ejection fraction (LVEF) of 30% along with moderate enlargement and reduced function of the right ventricle. Genetic testing showed the patient was heterozygous for a novel variant of uncertain significance in the DSC2 gene that codes for desmosomal protein desmocollin-2. Management at this time included a wearable defibrillator for 30 days, β-blockers, and abstaining from moderate and severe physical activity. The patient then received a single chamber subcutaneous intracardiac device (ICD) and was counseled on avoiding strenuous physical exertion. Six months later, she received an implantable ICD. The patient’s first degree family members were all offered screenings. Conclusion: This case demonstrates the complex workup involved as well as the therapeutic options for patients with ARVC. This case also highlights the importance of counseling, affected patients and unaffected carriers, as well as screening of first-degree relatives in hopes of preventing serious unwanted outcomes.

Keywords: Arrhythmia, Arrhythmogenic right ventricular cardiomyopathy, Cardiomyopathy, Sudden cardiac arrest, Sudden cardiac death

INTRODUCTION

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is an inherited myocardial condition that
primarily affects the right ventricle. The prevalence of ARVC is approximately 1 in 2000 to 1 in 5000 in the general population and generally affects men more than women at a ratio of 3:1 [1, 2]. The hallmark characteristic of this disease is continual loss and replacement of normal myocardium with fibrofatty tissue [3]. This replacement of tissue can lead to life-threatening arrhythmias and potentially sudden cardiac death (SCD). Despite the name, variations of ARVC can also affect the left side of the heart. In some cases, left-sided arrhythmias as well as left-sided heart failure may occur. Approximately 30–50% of cases of ARVC are considered familial and can be inherited in autosomal dominant or autosomal recessive forms. However, the autosomal dominant form is more common [4]. The pathogenesis is believed to be linked to over 30 genes that code for proteins involved in desmosomal structures between cardiac cells. The most known affected proteins are desmoplakin (DSP) [5], plakophilin 2 (PKP2) [6], desmoglein 2 (DSG2) [7], and desmocollin 2 (DSC2) [8]. These mutations cause a weakening in cell to cell mediated adhesions within the myocardium and lead to fibro-fatty replacement of normal ventricular tissue. Consequently, intense physical activity is restricted in these patients because of the increased risk of arrhythmia and sudden cardiac arrest/death.

What makes this diagnosis challenging is that many of the patients suffering from ARVC are young, athletic individuals, who seem overtly healthy. The most common presenting symptoms can include syncope, cardiac ischemia, arrhythmia, sudden cardiac arrest or sudden cardiac death (SCD). The original diagnostic criteria, which was established in 1994, was revised in 2010 (Table 1). In 1994, a diagnosis of ARVC had to include either 2 major, 1 major + 2 minor, or 4 minor features. When the criteria were revised in 2010, three categories were introduced. They included, definite (2 major or 1 major + 2 minor), borderline (1 major + 1 minor or 3 minor), or possible (1 major or 2 minor) features for the diagnosis of ARVC. The purpose of this revision was to increase the sensitivity of the criteria.

Table 1: Diagnostic criteria

<table>
<thead>
<tr>
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<tbody>
<tr>
<td>2 major, 1 major + 2 minor, or 4 minor</td>
<td>Definite = 2 major or 1 major + 2 minor</td>
</tr>
<tr>
<td></td>
<td>Borderline = 1 major + 1 minor or 3 minor</td>
</tr>
<tr>
<td></td>
<td>Possible = 1 major or 2 minor</td>
</tr>
</tbody>
</table>

I. Global/regional dysfunction/structural alterations

**Major**
- Severe dilatation and reduction of RVEF with/without (or only mild) left ventricular impairment
- Localized right ventricular aneurysms (akineti
  or dyskinetic areas with diastolic bulging)
- Severe segmental dilatation of the right ventricular

**By 2D echo**
- Regional right ventricular akinesia, dyskinesia, or aneurysm
- and one of the following (end diastole):
  - PLAX RVOT ≥32 mm (correct for body size [PLAX/BSA] ≥19 mm/m²)  
  - PSAX RVOT ≥36 mm (correct for body size [PSAX/BSA] ≥21 mm/m²)  
  - fractional area change ≤33%

**By magnetic resonance imaging scan**
- Regional right ventricular akinesia or dyskinesia or dyssynchronous right ventricular contraction
- and one of the following:
  - Ratio of right ventricular end-diastole volume (RVEDV) to body surface area (BSA) ≥110 mL/m² (male) or ≥100 mL/m² (female)
  - RV ejection fraction ≤40%

**By right ventricular angiography**
- Regional right ventricular akinesia, dyskinesia, or aneurysm

**Minor**
- Mild global right ventricular dilatation and/or ejection fraction reduction with normal left ventricular
- Mild segmental dilatation of the right ventricular
- Regional right ventricular hypokinesia

**By 2D echo**
- Regional right ventricular akinesia or dyskinesia
- and one of the following (end diastole):
  - PLAX RVOT ≥29 to <32 mm (correct body size PLAX/BSA ≥16 to <19 mm/m²)
  - PSAX RVOT ≥32 to <36 mm (correct body size [PSAX/BSA] ≥18 to <21 mm/m²)
  - fractional area change >33% to ≤40%

**By magnetic resonance imaging scan**
- Regional right ventricular akinesia or dyskinesia or dyssynchronous right ventricular contraction
- and one of the following:
  - Ratio of RVEDV to BSA ≥100 to <110 mL/m² (male) or ≥90 to <100 mL/m² (female)
  - Right ventricular ejection fraction (RVEF) >40–≤45%
II. Tissue characterization of wall

Major
- Fibrofatty replacement of myocardium on endomyocardial biopsy
- Residual myocytes <60% by morphometric analysis (or <50% if estimated), with fibrosis replacement of right ventricular free wall myocardium in ≥1 sample, with or without fatty replacement of tissue on endomyocardial biopsy
- Residual myocytes 60–75% by morphometric analysis (or 50–60% if estimated) with fibrous replacement of the right ventricular free wall in ≥1 sample, with or without fatty replacement of tissue on endomyocardial biopsy

Minor
- Inverted T-waves \( (V_1, V_2, V_3) \) or beyond; >14 years; in absence of complete RBBB QRS ≥120 ms
- Inverted T-waves in \( V_1 \) and \( V_2 \); >14 years; in absence of complete RBBB in \( V_1, V_2, V_3 \) or \( V_2 \)
- Inverted T-waves in \( V_1-V_2 \); >14 years; in presence of complete RBBB

III. Repolarization abnormalities

Major
- Inverted T-waves \( (V_1, V_2, V_3) \) or beyond; >14 years; in absence of complete RBBB QRS ≥120 ms
- Inverted T-waves in \( V_1 \) and \( V_2 \); >14 years; in absence of complete RBBB in \( V_1, V_2, V_3 \) or \( V_2 \)
- Inverted T-waves in \( V_1-V_2 \); >14 years; in presence of complete RBBB

Minor
- Inverted T-waves in \( V_1 \) and \( V_2 \); >14 years; in absence of complete RBBB or in \( V_1, V_2, V_3 \)
- Inverted T-waves in \( V_1-V_2 \); >14 years; in presence of complete RBBB

IV. Depolarization/conduction abnormalities

Major
- Epsilon waves or localized prolongation (>110 ms) of QRS complex in right precordial leads \( (V_1 \) to \( V_3 ) \)
- Late potentials by signal-averaged electrocardiogram in ≥1 of 3 parameters in absence of QRS duration of ≥110 ms on electrocardiography
- Filtered QRS duration (fQRS) ≥114 ms
- Duration of terminal QRS <40 µV (low amplitude signal duration) ≥38 ms
- Root mean square voltage of terminal 40 ms ≤20 µV
- Terminal activation duration of QRS ≥55 ms measured from nadir of S wave to end of QRS, including R’, in \( V_1 \), \( V_2 \) or \( V_3 \), in absence of complete RBBB

Minor
- Late potentials signal-averaged electrocardiogram (SAECG)

V. Arrhythmias

Major
- NSVT or sustained ventricular tachycardia of LBBB morphology with superior access (negative or indeterminate QRS in leads II, III, and aVF and positive in lead AVL)
- NSVT or sustained ventricular tachycardia of right ventricular outflow configuration, LBBB morphology with inferior access (positive QRS in leads II, III, and aVF and negative in lead aVL) or of unknown axis
- >500 ventricular extrasystoles per 24 hours (Holter)
- ARVC/D confirmed in false discovery rate who meets task force criteria
- ARVC/D confirmed pathologically at autopsy or surgery in false discovery rate
- Pathogenic mutation (associated or probably associated with ARVC/D) in patient under evaluation

Minor
- LBBB sustained or NSVT (ECG, Holter, Exercise tolerance test)
- >1000 ventricular extrasystoles per 24 hours (Holter)
- ARVC/D confirmed in false discovery rate who meets task force criteria
- ARVC/D confirmed pathologically at autopsy or surgery in false discovery rate
- Pathogenic mutation (associated or probably associated with ARVC/D) in patient under evaluation
- History of ARVC in false discovery rate in whom not possible or practical to determine if family member meets task force criteria
- Premature sudden death (<35 years) due to suspected ARVC/D in false discovery rate
- ARVC/D confirmed pathologically or by current task force criteria in second-degree relative

VI. Family History

Major
- Familial disease confirmed at necropsy or surgery
- ARVC/D confirmed in false discovery rate who meets task force criteria
- ARVC/D confirmed pathologically at autopsy or surgery in false discovery rate
- Pathogenic mutation (associated or probably associated with ARVC/D) in patient under evaluation

Minor
- Familial history of sudden death (<35 years) due to suspected ARVC/D
- ARVC/D confirmed in false discovery rate who meets task force criteria
- ARVC/D confirmed pathologically or by current task force criteria in second-degree relative

Source: With permission from Hopkinsmedicine.org
and to increase the likelihood affected family members being identified before a cardiac event. The criterion was also revised in part to change in genetic testing and technological advances in medicine. If a patient survives the initial event, current diagnostic modalities include electrocardiography, family history, echocardiogram, magnetic resonance imaging scan, angiography and myocardial biopsy.

CASE REPORT

A 27-year-old athletic female with no known past medical history collapsed while playing frisbee in the park. Upon emergency medicine services (EMS) arrival, the patient was unconscious, pulseless, and in ventricular fibrillation. Cardiopulmonary resuscitation efforts were initiated and lasted 30 minutes. The patient received 8 DC shocks as well as on-scene intubation. Resuscitation attempts were successful and the patient was brought to the emergency department and transferred to the ICU.

Initial electrocardiography (Figure 1) showed a QT interval of 460 ms and T-wave inversion in V1, V2, and V3. Transthoracic echocardiogram revealed a left ventricular ejection fraction (LVEF) of 30% along with moderate enlargement and reduced function of the right ventricle. The patient was successfully extubated four days after admission. Physical examination at that time was unremarkable. Follow-up transthoracic echocardiogram revealed a preserved LVEF of 55%. Cardiac catheterization was negative for coronary disease. The initial read of the cardiac MRI scan focused specifically on the left ventricle and was unremarkable. Genetic testing showed the patient was heterozygous for a novel variant of uncertain significance in the DSC2 gene that codes for desmosomal protein desmocollin-2, the pathogenic variants of which are found in autosomal dominant forms of ARVC. After the genetic study was performed, cardiac MRI scan with and without contrast (Figure 2) was read for a second time with a focus on the right ventricle. This revealed mild dilatation of the right ventricle, an elevated right ventricular end-diastolic volume index of 111 mL/m$^2$ (normal range 60–100 mL/m$^2$), a decreased right ventricular ejection fraction (RVEF) of 36% (normally 60%), and no evidence of regional wall defects. Electrocardiography changes as well as the mutation in the DSC2 gene are consistent with two of the major criteria of ARVC and support a definitive diagnosis. Management at this time included a wearable defibrillator for 30 days, β-blockers, and abstaining from moderate and severe physical activity. The patient then received a single chamber subcutaneous intracardiac device (ICD) and was counseled on avoiding strenuous physical exertion. Six months later, she received an implantable ICD and has been following-up with our cardiac clinic. The patients first degree family members were all offered screenings.

DISCUSSION

The diagnosis of arrhythmogenic right ventricular cardiomyopathy can be extremely challenging. This is because many patients are young and healthy adults before a symptomatic event ensues, usually precipitated by exercise. Unfortunately, by the time an event occurs, it may be too late because the most common presenting symptoms are cardiac ischemia, syncope, arrhythmia, or sudden cardiac arrest/death. ARVC accounts for 17% of cases of sudden cardiac death in the United States [9]. By definition our case represented a definitive diagnosis of ARVC due to two characteristics of the major criteria being met. This included T-wave inversion in V1–V3 with absence of right bundle branch block and mutation in desmocollin-2, which has a genetic association with ARVC [8, 10]. Restricting strenuous physical activity is imperative for both affected individuals as well as healthy genetic carriers in order to prevent exercise induced cardiac events and progression of disease [11, 12]. Additionally, clinical surveillance every two years and abstinence from intense physical activity is recommended.
in individuals with either unknown genotypes or in individuals who are known carriers of the diseases. Although there is no cure for ARVC, the name of the game is symptom management and prevention of arrhythmia or sudden cardiac death. This can be achieved with medical therapy that includes beta-blockers, diuretics, ace-inhibitors or angiotensin II receptor antagonists. Other therapies include endo/epi cardiac ablation or an intracardiac device. If these regimens prove to be suboptimal in symptom control, cardiac transplant is an alternative option. Ultimately, ARVC is a serious and complex cardiomyopathy that can result in ventricular arrhythmias and sudden cardiac death. The ARVC has an underestimated prevalence and is a challenging diagnosis that requires a high index of suspicion. Although many patients including the one discussed in this case do not show evidence of right ventricular damage, eventually all patients will. As such, diligent follow-up is imperative when treating patients with ARVC. Many aspects of ARVC have yet to be discovered and unfortunately no cure has been found to date.

CONCLUSION

This case demonstrates the complex workup involved in cases of (ARVC) as well as the therapeutic options for these patients. This case also highlights the importance of counseling, affected patients and unaffected carriers, as well as screening of first-degree relatives in hopes of preventing serious unwanted outcomes.

*********

Author Contributions

Zaid B. Al Jebaje – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

John Elibol – Substantial contributions to conception and design, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Robbie Wall – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Osman Saleem – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor of Submission

The corresponding author is the guarantor of submission.

Source of Support

None

Conflict of Interest

Authors declare no conflict of interest.

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REFERENCES


Introduction: Dental implants are becoming the most efficient treatment option for missing teeth. These are because of their ability to osseointegrate directly into the jawbone and then support dental prosthesis. However, a challenged bone condition may prevent the achievement of an optimal implants placement and then cause clinical complications. One of these conditions could be the focal osteoporotic bone marrow defects (FOBMD). Case Report: A 22-year-old female was referred with complaint of missing tooth #35. The void was filled using Bio-Oss® bone grafts. Thereafter, Straumann® ITI Dental Implant (Ø 3.3 mm, Roxolid®, SLActive®) was immediately placed with a torque wrench with adequate primary stability. Four months after placement, the implant was fully integrated and showed good healing. No clinical abnormality was noted. Conclusion: The presence of FOBMD does hinder the mechanical stability of dental implants, although it is not negatively affect implants osseointegration from biological point of view. Therefore, we recommend securing the initial stability of dental implants using bone grafting procedure simultaneous the implant placement.

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Received: 26 September 2017
Accepted: 25 October 2017
Published: 01 December 2017

Keywords: Bone grafts, Dental implants, Focal osteoporotic defects, Primary stability

How to cite this article


Article ID: Z01201712CR10868HA

doi:10.5348/ijcri-2017129-CR-10868

INTRODUCTION

Nowadays, dental implants are becoming the most alternative treatment option for missing teeth. These are because of their ability to osseointegrate directly into the jawbone and then support dental prosthesis [1]. However, a challenged bone condition may prevent the achievement of an optimal implants placement and then cause clinical complications [2]. One of these conditions could be the focal osteoporotic bone marrow defects (FOBMD) [3]. However, in oral implantology, serious complications might occur in the presence of FOBMD, mainly in the posterior mandibular region of the jawbone. This would probably cause an incidence of implant fixture displacement inside the body of the mandible [4, 5]. Such serious complication can occur immediately with the surgical insertion of dental implants or postoperatively because of a deficiency in the primary stability of implants [6, 7].

Clinically, FOBMD is an unusual condition that appears as undefined radiolucent area in the jawbone [3, 8]. It is usually localized, asymptomatic, and associated with previous history of extraction of teeth. They occur more commonly in the middle-aged female patients and
show a predilection for the region of mandibular molars [9]. The radiographic appearance of FOBMD including scattered trabeculae may extend short distances into the defect with irregular borders [10]. The etiology of the FOBMD is still unknown. However, it may be related to an alteration in the repair process of bone trabeculae in the area of trauma or inflammation after teeth extraction [11]. This would also be related to ischemic changes in the bone marrow tissue, which stimulate the development of hematopoietic foci [8]. For the placement of dental implants, bone morphology as well as density plays an important role in the implant stability and then success outcome of implant treatment. Serious complications and accidents related to surgery can include the displacement or migration of implants into the body of jawbones because of poor surgical technique or anatomic variances (i.e., FOBMD).

This paper describes a successful dental implant placement in a focal osteoporotic bone marrow defect located in the posterior left mandible of a 22-year-old female patient.

CASE REPORT

A 22-year-old female was referred to the dental implants clinic, with complaint of missing tooth in the left lower posterior region. Dental history revealed that #35 was badly decayed, non-restorable, and then had been extracted five years ago. Intraoral examination showed healthy mucosa and there was not any sign of infection. Her past medical history was unremarkable. Panoramic radiography of the jaws showed radiolucent area with ill-defined borders associated to the region of #35 (Figure 1). This condition was asymptomatic and no expansion of the mandibular cortical bone was clinically detected. Thus, a provisional diagnosis of focal osteoporotic bone marrow defect (FOBMD) was considered as a differential diagnosis based on age, site, clinical and radiographic findings.

The patient was given the option of possibility of dental implant treatment. Pros and cons of the procedure were explained to the patient and informed consent was obtained. Then, preoperative evaluation included clinical and radiographical assessments of implant size, position of implant, and anatomical landmarks were applied. Under local anesthesia (2% lignocaine with adrenaline 1:80,000), a crestal incision was placed in the edentulous #35 region. A full-thickness mucoperiosteal flap was elevated and reflected. Implant site was marked with round bur. A 2-mm pilot drill osteotomy was done to perforate the crestal cortical bone. Large FOBMD was found isolated with several millimeters in diameter. The void was filled using deproteinized bovine bone grafts (Bio-Oss®, Osteohealth Co, Shirley, NY, USA) (Figure 2). Thereafter, Straumann® ITI Dental Implant System (Institut Straumann AG Waldenburg, Switzerland) was used. Implant fixture (Ø 3.3 mm, Roxolid®, SLActive®) was immediately placed with a torque wrench with adequate primary stability. The selection of Straumann® implant was suitable for bone level treatment in combination with subgingival healing. It has a special anatomical design, which combines a cylindrical shape in its apical region and a conical shape in the coronal region, making this implant particularly suitable for implantation in challenged condition. Roxolid® Titanium-Zirconium alloy material is designed to offer more confidence when placing small diameter implants in the molars region. Also, SLActive® has unique properties of hydrophilicity and chemical activity, which can accelerates implant osseointegration [12].

Flap repositioned and sutured with simple interrupted sutures using resorbable Vicryl material size 4-0 (Vicryl®, Ethicon, Johnson & Johnson, Norderstedt, Germany). Primary closure was achieved. Immediate postoperative radiograph was taken to confirm position of implant fixture (Figure 3). Antibiotics and analgesics were prescribed for seven days. The patient was placed on regular maintenance protocol. Four months after placement, the implant was fully integrated and showed good healing (Figure 4 and Figure 5). No clinical abnormality was noted.
DISCUSSION

Focal osteoporotic bone marrow defect (FOBMD) has been reported as an unexpected radiolucency in the posterior mandible of the middle-aged female patients [8, 13]. Our case diagnosis is consistent. Moreover, the present case shows the fact of the rigorous method for radiographic assessment of the patients and X-Ray prescription as an initial step for treatment planning. In addition, the use of CBCT examination should not be neglected, as it can be an accurate method for the pre-assessment during dental implant treatments [14].

The exact cause of FOBMD has not been confirmed [3]. However, it frequently occurs in an extraction area and might be related to alterations in socket healing and bone trabeculation [8]. Other possibilities might be related to some systemic conditions, e.g., hematologic disorders [11]. In this case, we could speculate that the FOBMD is associated with the impairment of alveolar bone healing after tooth extraction of #35. Also, the patient medical history was normal excluding a possible association with other medical conditions. Previous reports showed that ~70% FOBMD is occurred in the posterior mandible in adult women and only minority of the cases are reported in maxilla [15]. Most of the cases are asymptomatic and have no swelling, found incidentally on routine radiographs. Radiographic appearance of FOBMD shows isolated radiolucency with several millimeters to centimeters in diameter and ill-defined areas [10, 16]. In addition, the radiolucent area of FOBMD is trabeculated randomly and separately distributed. Unlike other odontogenic lesions, FOBMD tend to respect the lamina dura of adjacent teeth without expanding the bone cortex to the corresponding area [17].

Clinically, serious complications in oral implantology might occur in presence of FOBMD. This would probably cause an incidence of implant fixture displacement inside the body of the mandible [4, 5, 18]. The displacement of an implant can occur immediately or within a short period due to poor initial implant stability can result from the anatomical variances. And this because of the bone pattern of FOBMD provides low mechanical stability for dental implants at placement and should be carefully considered [6]. As previously reported, unforeseen accidents of implant displacement could occur especially in the posterior FOBMD, when could not be delineated on a panoramic radiograph [6, 19]. Thus, careful radiographic evaluations may be necessary for female patients with extracted posterior teeth prior to dental implants placement.

For management, it has been suggested that surgical manipulation of dental implants may improve the mechanical pattern of bone at an implant site [20]. As in this case, we recommend augmenting the FOBMD with bone grafting materials prior to implant insertion. For such surgical protocol, it was done similar to the method of immediate implant placement after teeth extraction.
CONCLUSION

Focal osteoporotic bone marrow defect (FOBMD) is generally a rare condition that might be detected in posterior mandible of middle-aged women. When FOBMD presents with unusual radiographic findings, it requires more specific examination of the tissue. The presence of FOBMD does hinder the mechanical stability of dental implants, although it is not negatively affect implants osseointegration from biological point of view. Therefore, we recommend securing the initial stability of dental implants using bone grafting procedure simultaneous the implant placement.

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Author Contributions

Hamdan S. Alghamdi – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor of Submission

The corresponding author is the guarantor of submission.

Source of Support

None

Conflict of Interest

Author declares no conflict of interest.

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Obstructed inguinal hernia containing female reproductive organ: A rare presentation

Diyaree N. Ismael, Zuhair D. Hammood, Fahmi H. Kakamad, Goran A. Qadr

ABSTRACT

Introduction: Inguinal hernia is a common surgical problem; usually the bowel and omentum herniate through the defect. Herniation of the female reproductive organs is extremely rare. The aim of this study is to report a case of obstructed inguinal hernia with herniation of ovary and uterine tube. Case Report: A 14-year-old girl with a history of reducible left inguinal hernia was admitted to the emergency room. She had tender irreducible swelling of left groin associated with nausea for two days. On examination there was irreducible tender left inguinal swelling. Ultrasound examination proved the diagnosis as irreducible left inguinal hernia containing left ovary with complex cystic content. Under general anesthesia, exploration of the left inguinal canal was done. The sac was opened; both ovary and fallopian tube were viable. The ovarian cyst was excised and the sac closed then reduced to the abdominal cavity. Round ligament was preserved and the posterior wall was reinforced by darning procedure. Conclusion: Inguinal hernia with ovarian and uterine tube herniation is a very rare disease. Management is by surgery with careful reduction of the content.

Keywords: Hernia, Ovary, Uterine tube

INTRODUCTION

The inguinal hernia is a relatively common surgical problem in kids with an accounted occurrence of 0.8–4.4% [1]. About 13.7–23% of inguinal hernias of indirect type happen in young females [2]. The proportion of male to young ladies is 6:1.1. Nearly, 68.1% occur on the right side, 23.4% on the left side and 8.5% are bilateral. The accounted frequency is about 71% for youngsters under five years and 30% for youths or ladies in reproductive ages [3]. The most common organs involved in the hernia pathology are omentum and small bowel. Other intra-abdominal organs are infrequently encountered in the inguinal canal such as the vermiform appendix, urinary bladder and fibroid. Very rarely, ovaries and fallopian tubes have been reported as content of inguinal hernia sac which presented challenge to the physician and surgeon [4]. The inguinal hernia containing a reproductive organ...
in young woman usually happens due to a partial closure of the peritoneal processus vaginalis. Unfortunately, the repair of hernia in female child is usually underwent with much less care than in male because of absence of spermatic cord in female, this led to different type of injuries to both ovary and uterine tube [4, 5]. The aim of this report is to discuss an uncommon case of left obstructed sliding inguinal hernia with left ovary containing a large ovarian cyst along with it is uterine tube.

CASE REPORT

A 14-year-old girl a student of secondary school and known case of neglected, reducible left inguinal hernia since two years ago, was admitted to the emergency room with a two-day history of tender irreducible swelling of left groin associated with nausea but no vomiting. On examination; There was 5x6 cm, irreducible, tender left inguinal swelling (Figure 1), the abdomen was soft and the bowel sound was positive. Vital signs were stable apart from tachycardia (114 beats per minute). Urgent ultrasound examination was done which proved the diagnosis as irreducible left inguinal hernia containing left ovary with complex cystic content. After informed consent, she was taken to the emergency operating room. Under general anesthesia with endotracheal tube and single prophylactic antibiotic, exploration of the left inguinal canal was done. The hernia sac and its contents were isolated which were irreducible below the superficial inguinal ring. The hernia was of indirect type and the contents were ovary containing large cyst and the fallopian tube. The sac was opened; both ovary and fallopian tube were viable while the fimbrias were gangrenous (Figure 2). The ovarian cyst was excised and the small gangrenous part of the fimbrias was left, the sac closed then reduced to the abdominal cavity. Round ligament was preserved and the posterior wall was reinforced by Darning procedure (Figure 3). The wound was closed in layers and the patient was kept in hospital for two days. Postoperative period was uneventful. The skin stitches were removed one week later.

DISCUSSION

In female, the inguinal canal normally forms a passage for the round ligament of the uterus, processus vaginalis and labial arteries [5]. The round ligament formed by the distal part of the gubernaculum, while proximal part forms the suspensory ligament of the ovary [1]. The reproductive systems of both females and males share the same steps in the early uterine development. Understanding this fact may help understanding the mechanism of the ovarian herniation through the inguinal canal [6]. The pathophysiology of the development of sliding inguinal hernias of ovary and fallopian tube in female is thought...
to be a homologous to the normal physiology of the testis descent in male [1]. Increased pressure over the hernia may compromise blood supply of its contents, especially venous one, and may lead to venous congestion and subsequently ischemia and infarction. Another risk of vascular compromise is torsion of the ovary as it has a long pedicle [2]. In this case, both the ovary and the tube were viable. Differential diagnoses of the inguinal region swelling in the female are varied, which may include direct inguinal hernia, indirect inguinal hernia, soft tissue tumors (sarcoma, leiomyoma or lipoma), cystic lesions, abscess collection, enlarged lymph nodes, or hydrocele [1]. The diagnosis can be easily confirmed by ultrasonography scan in most cases. However, for better anatomical illustration and details of the hernias and its boundaries as well as contents magnetic resonance imaging (MRI) and computed tomography (CT) scan can be done [7]. The current case was successfully diagnosed preoperatively by ultrasound. When the inguinal hernia sac contents formed by ovary and uterine tube, they are usually associated with developmental anomalies of the genital tract like bicornuate uterus, vaginal atresia and renal anomalies. The current case was free from other anomalies [8]. Inguinal hernia is treated with reduction of its content provided that there is no abnormality of the ovary or the fallopian tube, no impaired blood supply and there are no features of salpingitis. Reduction of the contents should be followed with ligation at high level of the sac then closure of deep inguinal ring and finally re-supporting of the posterior wall of the inguinal canal by a non-absorbable mesh in females aged more than 20 years, the procedure which is performed in this case [8]. Associated injury to the hernia content is a common problem especially in females with sliding inguinal herniation of vital reproductive organs like ovaries, fallopian tubes and uterus. Therefore, ligation of the hernia sac without opening should be avoided. However, in all cases with inguinal hernia, the sac should be opened during repair to exclude the presence of sliding ovaries or fallopian tubes, and if present, should be dissected and reduced carefully to abdominal cavity before high ligation of the sac to avoid their associated injuries [1]. In current case, the herniated ovary and uterine tube was safely reduced.

CONCLUSION

Inguinal hernia with ovarian and uterine tube herniation is a very rare disease, when it present, it can be diagnosed by ultrasonography, and management is by surgery with careful reduction of the content.

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Author Contributions
Diyaree N. Ismael – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
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Guarantor of Submission
The corresponding author is the guarantor of submission.

Source of Support
None

Conflict of Interest
Authors declare no conflict of interest.

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Kyphoscoliosis correction with recurrent tethered cord syndrome due to myelomeningocele and neurofibromatosis type 1

Rachana Tyagi, Smit Shah

ABSTRACT

Introduction: Recurrent tethered cord syndrome (TCS) is a neurosurgical condition that is associated with spinal dermoid cysts, myelomeningoceles and lipomyelomeningoceles most commonly. However, recurrent TCS due to myelomeningocele and neurofibromatosis type 1 (NF1) has not been reported. Case Report: We describe a case of a 13-year-old boy with recurrent TCS and progressive spinal deformity with a severe kyphoscoliosis due to myelomeningocele and NF1. We also review the anterior and posterior surgical approaches to vertebrectomies along with their indications and contraindications. Finally, we also discuss specific risks of scoliosis repair in NF1 patients due to high probability of pseudoarthrosis. Conclusion: We conclude that in patients with recurrent TCS, posterior fusion with shortening and without detethering was a successful procedure which should be considered as a primary surgical option. Anterior fusion may be delayed in such patients and used only if necessary given the additional risks in such medically fragile patients.

Keywords: Neurofibromatosis type 1, Pseudoarthrosis syringohydromyelia, Recurrent tethered cord syndrome, Vertebrectomy

How to cite this article


Article ID: Z01201712CI10022RT

doi: 10.5348/ijcri-201704-CI-10022

INTRODUCTION

Recurrent tethered cord syndrome (TCS) is a progressive neurological and orthopedic condition that occurs due to recurrent fixation or tethering of the distal spinal cord after an initial detethering procedure in pediatric and adult patients [1]. It develops due to scar tissue adhering to the spinal cord after an initial detethering procedure for spinal cord anomalies including: dorsal lipomas, congenital spinal dermoid cyst or sinus tract and myelomeningoceles and lipomyelomeningoceles [2]. As the child grows, there is a progressive stretching of the spinal cord which can lead to worsening neurological and orthopedic conditions along with deterioration of blood supply to the spinal cord.

Based on animal studies, Yamada et al. have demonstrated TCS with a tight filum to be a stretch induced functional disorder at the lumbosacral spinal cord that occurs due to failure of oxidative metabolism at the level of the mitochondria due to marked reduction in cytochrome a and a3 which possibly results in abnormally inelastic filum [3]. As a result, ‘...abnormal inelastic filum
interferes with normal cord ascension and results in low lying conus medullaris...’ which lies below the level of L1 and L2 [4]. Thus symptoms are increased with activity, particularly flexion and extension which increases tension on the distal cord. It is possible that similar mechanisms of spinal cord dysfunction are likely in play with recurrent tethered cord syndrome due to more extensive tethering lesions.

Clinical manifestations of recurrent TCS vary between pediatric and adult patients. Unlike adult patients, pediatric patients usually complain of poorly localized diffuse pain in the lower extremities and perineum, or back mostly confined to the lower back with infrequent radiation to legs [5]. Neurological symptoms include bowel/bladder dysfunction, weakness and lethargy with activity, exacerbating gait and balance problems, numbness or parasthesia along with frequent bruising. Spinal signs/symptoms include muscle spasms, tenderness on palpation, scoliosis and kyphosis with possible truncal decompensation. The lower extremities may show deformities or atrophy, commonly worse on one side, with weakness and sensory loss on examination. Urodynamics may be the most sensitive test showing minor changes consistent with neurogenic bladder even without obvious physical findings [6]. Magnetic resonance imaging scan will show recurrent tethering at the site of the previous surgery. A proximal syrinx may also be noted, or arachnoiditis of the cauda equina.

Neurofibromatosis 1 (NF1) is an autosomal dominant syndrome including café-au-lait lesions, Lisch nodules, intracranial tumors, peripheral neurofibromas, capillary hemangiomas, dural ectasia, scoliosis and other bony abnormalities. Despite its dominant genetic inheritance, spontaneous mutations of NF1 gene commonly occur. Due to variable genetic expressivity, severity of symptoms varies. Anterior sacral neural tube defects and neurofibromatosis type 1 are closely associated [7]. Even though, neurofibromatosis type 1 (NF1) is rarely associated with tethered cord, it is highly associated with dural meningocoele [8, 9]. Scoliosis is relatively common in these patients, and surgery is often indicated. The poor bone quality associated with this disease increases the risks of pseudoarthrosis and often requires more aggressive fusion techniques or repeated procedures [10].

Surgery for repeated untethering has lower success rates with increased risk of complications, particularly new neurologic deficits. For pediatric patients with meningomyelocele and a Gibbus, possible treatment options include kyphectomy of the vertebrae responsible for kyphosis. This results in improved seating balance as well as restoration of an intact skin envelope by decreasing the incidence of pressure sores at the apex. For patients with severe kyphoscoliosis, a combined anterior and posterior spinal reconstruction can be used. This approach allows for maximal exposure, enhanced mobilization of the spinal elements, safety, and efficiency when operating around the spinal cord.

Such an approach facilitates posterior compression and anterior distraction, thereby allowing for greater correction and control [11].

Shortening the spine also allows for significant deformity correction without performing an initial detethering procedure, but still reducing the risk of a new neurologic deficit due to manipulation of the neural elements [11].

**CASE REPORT**

The patient was a 13-year-old boy with a history of de novo NF1 and thoracolumbar myelomeningocele. He had a progressive kyphoscoliosis with a large Gibbus complicated by pressure ulcers over the Gibbus as well as the sacrum. The patient also had a past medical history of Chiari Type II malformation with tethered cord, syringohydromyelia and hydrocephalus shunted at birth. He had significant difficulty with positioning in his chair and chronic back pain. He did have 4/5 iliopsoas function, but no significant function distally, with contractures of the knees and ankles. He also had worsening neurogenic bladder symptoms associated with left sided hydronephrosis.

We can see severe progression of scoliosis deformity as the age progresses (Figures 1–3). In Figure 1, when patient was nine year and eleven months old, there was severe right convex 20 degree curve from L1 to L3 which...
progressed to severe 85 degree curve from T9 to L3 with pelvic tilt at age 12 years and 5 months (Figure 3). In addition, preoperative imaging at age 12 demonstrated significant kyphosis in the thoracolumbar area with holocord syrinx and thinned neural tissue with minimal soft tissue covering at closure site (Figure 2).

Patient’s mother expressed significant reservations regarding detethering prior to deformity correction due to the risk of loss of hip flexion. He was currently able to assist with diaper changes by flexing his hips. Therefore, a detethering procedure was not planned, but instead a vertebrectomy to shorten the spine and prevent stretch on the tethered cord after deformity correction.

The initial procedure was placement of a halo head frame, with upright traction for one week. Minor improvement in the scoliosis at age 12 years 11 months was noted from 85–78 degrees, with no change in the neurologic examination (Figure 4). Therefore, posterior releases were performed with Ponte osteotomies from T6 to L2 and pedicle screw placement from T5 to L4, and the patient again placed in traction for two weeks, with further decrease in the deformity to 45 degrees (Figure 5). As adequate correction was not obtained with these measures, a hemivertebrectomy from the posterior approach was then performed at the vertex of the curve at T11, and the posterior fusion was performed with successful correction of the deformity and no loss of hip function, extending the fusion to the pelvis with iliac bolts with a final coronal curve of 16 degrees along with good sagittal balance (Figure 6 and Figure 7). He was maintained in a rigid TLSO clamshell brace for three months postoperatively to decrease the risk of pseudoarthrosis, with a plan for possible further anterior surgery if needed.

Follow-up studies at 41 months show excellent fusion with no loss of correction, and the patient had developed no further pressure ulcers, back pain, or change in lower extremity function. As we can see in Figures 8–11, there was no loss of correction two years postoperative with 15 degree curve (Figure 8), 41 months postoperative at age 16 years 5 months showing successful fusion of facets bilaterally (Figure 9) and no loss of correction along with mild kyphosis at age 18 years and 9 months (approximately six years status post initial surgery) (Figure 10).

Figure 2: Preoperative magnetic resonance imaging scan at age 12 depicts significant kyphosis in the thoracolumbar area; holocord syrinx and thinned neural tissues with minimal soft tissue covering at closure site seen.

Figure 3: Anteroposterior and lateral images showing (A) Right convex 85 degree curve from T9 to L3, pelvic tilt age 12 years and 5 months, (B) Forward sagittal balance along with a forward tilt before traction.
DISCUSSION

The thoracolumbar spine can be accessed anteriorly, posteriorly or in combination [12, 13].

Anterior access [7] is indicated in spinal conditions like kyphosis, scoliosis, lordosis, compression fracture or dislocation of the vertebral body, degenerative disc disease, thoracolumbar disc herniation, pyogenic or parasitic infection of spine and malignancy involving primary tumor of the vertebral body. The retroperitoneal

Figure 4: (A) Initial upright traction with minimal improvement in curve to 78 degrees age 12 years and 11 months, (B) After 48 hours, curve decreased to 60 degrees.

Figure 5: Further improvement in curve after posterior release with Ponte osteotomies to 45 degrees with continued traction.

Figure 6: (A) Corrected pelvic tilt and Gibbus reduction after final hemivertebrae resection and fusion with posterior instrumentation eight days after final surgery, and (B) Final coronal curve 16 degrees, good sagittal balance on lateral film.

Figure 7: (A) Postoperative lateral view showing correction in sagittal imbalance; no residual Gibbus and (B) Postoperative image showing pedicle screws placement from T5 to L4; significant reduction in scoliosis; correction of coronal balance; leveled pelvis seen in seated position post hemivertebrectomy.
area is accessed after the abdominal organs are retracted anteriorly. The discectomy above and below and the vertebrectomy/hemivertebrectomy is performed with placement of graft and possible intervertebral instrumentation with lateral plating to provide stability to fuse the vertebral column rostrally and caudally.

Thus, an anterior approach vertebrectomy reduces the risk of spinal cord or dural injury during the vertebrectomy, by eliminating the need for retraction and allows for greater sagittal deformity correction by allowing a larger lordotic cage to be placed without risk of neurologic injury. Anterior stability of the construct with a true 360 degree fusion construct also reduces the risk of pseudoarthrosis and crankshafting in the young patient. However, as compared to posterior approach, morbidity due to respiratory failure is more common with anterior approach [14]. A shortening procedure also cannot be performed without resection of the posterior elements, and thus would actually require a posterior laminectomies/facetectomies prior to the anterior fusion.

The posterior approach [15, 16] is more commonly used for fusion and stabilization of the vertebral column as compared to the anterior approach that is reserved for anterior column reconstruction and vertebrectomy. Since, the hemivertebrae in the thoracolumbar region deviates dorsolaterally with a kyphotic deformity, it can be identified posteriorly without significantly increased risk of neurologic injury. After the vertebral disc and endplate in both the cranial and caudal adjacent segments are curetted, bone chips obtained during the vertebrectomy can be inserted into the defect rather than a cage to shorten the spinal column, but with less correction of the kyphosis. A long construct with extensive deformity correction is possible with the posterior approach using osteotomies at the facets to further mobilize the segments [17]. Posterior only approach does have higher recurrence

Figure 8: No loss of correction at second year postoperative with 15 degree curve.

Figure 9: Coronal computed tomography scan showing successful fusion of facets bilaterally at 41st months postoperative; age 16 years and 5 months.
rates of vertebral fractures along with contraindications like fragmented fractures of thoracolumbar spine [6, 7] for example: burst fractures which occurs due to vertical compression of the spine.

Scoliosis correction in NF1 patients carries increased risks relative to other causes of deformity. Crawford et al. demonstrated increased risk of pseudoarthrosis, paraplegia and intraoperative hemorrhage with complex spinal surgeries specifically in patients with NF1. Furthermore, dystrophic spinal deformities like scoliosis, kyphosis or kyphoscoliosis are challenging to manage postoperatively because they are rapidly progressive in nature and result in pseudoarthrosis very quickly. Decortication, abundant autogenous bone grafting, and segmental instrumentation are necessary to minimize the occurrence of pseudoarthrosis [11]. In addition, during the anterior approach, it is extremely important to resect all the pathologic disc soft tissue in order to prevent the resorption in the mid portion of the graft material due to incomplete resection. Finally, even if rigid fixation has been achieved using modern instrumentation, orthotic immobilization is recommended until a fusion mass with trabecular pattern is seen in CT scan performed six months after surgery [18]. In these patients, an anterior and posterior construct for increased construct rigidity is recommended.

For the current patient, the risk of new neurologic deficit with loss of hip function was a significant concern, and would have adversely affected his mother’s ability to provide care for the patient, likely necessitating two care givers for diaper changes as he grew, and further limiting his ability to reposition himself with increased risk of pressure ulcers. Correction of the deformity without detethering placed him at risk for increased stretch on the placode with likely new neurologic deficit. However, due to the holocord syrinx and severely thinned placode, the repeat detethering procedure itself would likely have led to permanent neurologic deficits from manipulation of the neural elements. Thus, a shortening procedure was planned if traction alone did not provide adequate deformity correction.

This case was discussed with a group of experienced spine surgeons with a consensus of proceeding with staged surgeries with initial posterior fusion only, and a delayed anterior fusion at sixth month if necessary. A step-wise escalation of deformity correction was followed with initial awake vertical traction in his wheelchair to allow for continuous monitoring of his neurologic function and to limit the risk of surgery. The initial surgical procedure of only osteotomies and placement of screws was then performed. Inadequate correction determined the hemivertebrectomy was required, with excellent coronal and sagittal balance and no loss of function. Subsequent imaging showed excellent fusion of the posterior columns with no loss of correction, and no need for anterior stabilization.

**CONCLUSION**

In this patient with recurrent tethered cord and a severe kyphoscoliosis due to myelomeningocele and NF1, posterior fusion with shortening and without detethering was a successful procedure. This technique may be considered as a primary surgery in similar patients. Although scoliosis correction in NF1 is known to have an increased risk of pseudoarthrosis, and therefore an anterior fusion was considered, our patient did have a successful posterior-only procedure. Anterior fusion may be delayed in such patients and used only if necessary given the additional risks in these medically fragile patients.

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**Author Contributions**

Rachana Tyagi – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Smit Shah – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor of Submission
The corresponding author is the guarantor of submission.

Source of Support
None

Conflict of Interest
Authors declare no conflict of interest.

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SUGGESTED READING

• https://rarediseases.org/rare-diseases/tethered-cord-syndrome/
Obturator with favorable intaglio extension

Abdul Habeeb Bin Mohsin

CASE REPORT

A 47-year-old female reported to the department of prosthodontics with a chief complaint of broken prosthesis causing difficulty in speech and nasal discharge of water and exudates into the mouth. Past dental history revealed that patient underwent surgical treatment for cleft lip and cleft palate 10 years back, with missing incisors and canine in right quadrant of maxilla. Post-surgery patient was using labial obturator to cover the maxillary defect. Clinical examination revealed maxillary defect extending from alveolar ridge of maxillae in relation to 11, 12 and 13 missing teeth to hard palate involving part of mid palatal raphae. The patient was rehabilitated with obturator closing the defect and replacing the missing teeth along with it to restore esthetics. The obturator was relined with soft liner with Intaglio Extension utilizing favorable undercuts for retention atraumatically (Figures 1–3).

DISCUSSION

Obturator prosthesis is commonly used in cases of maxillary defects to separate oral cavity and nasal cavity. It helps to restore speech and normal deglutition [1]. Designing of prosthesis is critical in maxillary defects [2]. In this case, retentive design of prosthesis was planned in a way to extend the intaglio soft liner extension into the favorable undercut into the defect area approximating the soft tissue [3, 4]. Favorable soft intaglio extension will impart proprioception and retention. Overall treatment provided a positive impact on physical health and psychological well-being of the patient.

Figure 1: Maxillary defect (A) Occlusal view, and (B) Labial view.
CONCLUSION

Obturator prosthesis improves oral function. Appropriate planning of obturator improves deglutition, speech, esthetics, retention and enhances psychological comfort. It benefits the morale of the patients.

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Keywords: Intaglio extension, Obturator, Prosthesis, Soft liner

How to cite this article


Article ID: Z01201712CL10138AM

doi:10.5348/ijcri-201728-CL-10138

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Author Contributions

Abdul Habeeb Bin Mohsin – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor of Submission

The corresponding author is the guarantor of submission.

Source of Support

None

Conflict of Interest

Author declares no conflict of interest.

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Figure 2: Obturator prosthesis with intaglio extension (A) Saggital view, and (B) Lateral view.

Figure 3: Postoperative view.
REFERENCES


