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Autophagy in cancer

Ranjan Agrawal

The 2016 Nobel Prize in Physiology and Medicine has been awarded to Yoshinori Ohsumi for the discovery of mechanism for autophagy. His work published in 1993 greatly transformed the understanding of autophagy and now its role in human physiology and disease is well appreciated.

Autophagy is a general term for the degradation of cytoplasmic components within the lysosomes. There are three types of autophagy — macroautophagy, microautophagy, and chaperone-mediated autophagy. The term usually indicates macroautophagy unless otherwise specified. Autophagy is mediated by a unique organelle called the autophagosome.

The most important trigger of autophagy is nutrient starvation. Lack of any type of essential nutrient can induce autophagy. Autophagy plays a housekeeping role in removing misfolded or aggregated proteins, clearing damaged organelles, such as mitochondria, endoplasmic reticulum and peroxisomes, as well as in eliminating intracellular pathogens. Thus, it is actually a survival mechanism. Autophagy can be either non-selective or selective in the removal of specific organelles, ribosomes and protein aggregates. It promotes cellular senescence and cell surface antigen presentation, limits necrosis making its role important in preventing diseases such as cancer, neurodegeneration, cardiomyopathy, diabetes mellitus, autoimmune diseases, liver diseases and certain infections. Thus, it can easily be said that disruption of autophagy leads to diseases. Autophagy can be considered as a cellular ‘recycling factory’ that promotes energy efficiency through ATP generation and mediates damage control by removing non-functional proteins and organelles [1].

Autophagy has a vital role in cancer; both in protecting against cancer and potentially contributing to the cancer growth. Autophagy promotes survival of tumor cells that have been starved, or that degrade apoptotic mediators such as the use of inhibitors of the late stages of autophagy (such as chloroquine). Role of autophagy is both as a tumor suppressor and as a factor in the tumor cell survival. It is more likely to be used as a tumor suppressor. Necrosis and chronic inflammation also have been limited through autophagy which helps protect against the formation of tumor cells. In neoplastic cells, autophagy is used as a way to deal with stress on the cell. Once this autophagy related genes are inhibited, cell death is potentiated. Metabolic stresses include hypoxia, nutrient deprivation, and an increase in proliferation. Autophagy is activated to recycle ATP and maintain survival of the cancerous cells. Autophagy enable continued proliferation of tumor cells by maintaining cellular energy production. By inhibiting autophagy genes in these neoplastic cells, regression of the tumor and extended survival of the organs affected by the tumors has been reported. Inhibition of autophagy has also a role in enhancing the effectiveness of anticancer therapies [2].

**********

Keywords: Autophagy, Autophagosomes, Cancer

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Richter’s hernia: Two observations in the Baka pygmies of Eastern Cameroon

Tsopmene Dongmo Marvin, Nkeck Jan René, Eloundou Ngah Joseph

ABSTRACT

Introduction: Richter’s hernia has a misleading clinical presentation at the onset of the disease due to common lack of obstructive signs whereas there is visceral strangulation. The delay in diagnosis is therefore long leading to high morbidity and mortality in a rural context where access to essential surgical care is limited.

Case Series: We report two cases of Richter’s hernia, discovered intraoperatively in a woman and a man of respectively 24 and 29 years of age. They had direct inguinal hernia with partial incarceration of the distal bowel loop without obstruction. The treatment consisted in both cases in a segmental resection and end to end anastomosis followed by the inguinal ring closure and a parietal repair. The postoperative follow-ups in both cases were unremarkable.

Conclusion: Clinical signs of Richter’s hernia are generally misleading at the onset of pathology and imaging is inconclusive. Therefore, awareness during the clinical examination remains the key for proper diagnosis and timely management, for a good postoperative outcome.

The procedure depends on the peroperative findings.

Keywords: Cameroon, Diagnostic delay, Richter’s hernia, Strangulation

INTRODUCTION

The first scientific description of the Richter’s hernia was made in 1778 by the German surgeon August Gottlob Richter. It accounts for about 5–15% of strangulated hernias, in which only part of the circumference of the antimesenteric border of the bowel wall is incarcerated within the hernia sac leading to strangulation [1]. The lack of bowel obstruction is the known cause of diagnosis and management delay. A delay in management is associated with a risk of bowel ischemia, gangrene and perforation, leading to high morbidity and mortality.

CASE SERIES

Case 1

A 24-year-old female presented with a three-day history of a painful, non-reducible right inguinal swelling without any sign of bowel obstruction. The diagnosis of
a right inguinal hernia was made. The surgical findings were those of a direct inguinal hernia with 5 cm of the antimesenteric side of the distal ileum incarcerated and gangrenous (Figure 1). A short sub-umbilical median laparotomy allowed mobilization of the incarcerated loop. Resection of the segment of ileum involved was done with ileoileal hand sewn anastomosis followed by a parietal repair. The total duration of hospitalization was 10 days and the postoperative outcome good with a four month follow-up.

Case 2

A 29-year-old male presented with a week history of a right inguinal pain. The patient was ill-looking with a body temperature 38.9°C, heart rate 102 bpm, and blood pressure 99/52 mmHg. In addition, he had an abdominal tenderness in the hypogastric and right iliac fossa regions with a non-reducible inflammatory groin mass. Surgery revealed a strangulated direct inguinal hernia with a gangrenous sac. Following a kelotomy, we performed a resection of the necrotic part of the ileum loop and performed an end-to-end anastomosis. The parietal cure was done according to Shouldice technique. The total duration of hospitalization was seven days and a good outcome following three months postoperative follow-up.

DISCUSSION

Richter’s hernia is the incarceration of part of the circumference of the wall of the antimesenteric side of the loop through a small hernia ring. It was named after its first scientific description by the German surgeon August Gottlieb Richter in 1778 [1]. In the Caucasian subjects, the most frequent localization is the femoral canal (36–88%) followed by the deep inguinal ring (12–36%) [2, 3]. The two cases presented were direct inguinal hernias. This result is similar to that obtained by Wolfgang et al., and other authors who reported a clear predominance of direct inguinal hernias ranging from 78–94% [4–6]. The common use of laparoscopic surgery has led to increase abdominal wall incisional hernias [7]. Richter’s hernia represents in the general population 5–15% of the strangulated hernias [1]. This prevalence is higher in Africa where Hancock et al. reported in a study in Uganda a prevalence of 25% and Wolfgang et al. a prevalence of 81% in Sudan [1, 4]. This discrepancy could be explained on one hand by anatomical particularities, namely the small hernia rings with firm margins and on the other hand the malnutrition responsible for the increased elasticity of the intestinal wall. Kadirov et al. reported a female predominance with 57% of women, while Tomaszewski et al. and Wolfgang et al. reported a female predominance with sex ratio 6.3 and 1.4 respectively [1, 8, 9]. The clinical diagnosis of partial enterocele is not easy, the clinical presentation being misleading at the beginning, marked by an irreducible painful inguinal swelling without bowel obstruction. This bowel strangulation without obstruction would be one of the causes of diagnostic delay hence, a high morbidity and mortality. Medical imaging has a prime role in the diagnosis of Richter’s hernias, particularly with ultrasonography and computed tomography. However, they are generally inconclusive at the onset of the disease [1, 10, 11]. Surgical treatment should be carried out as soon as possible, either by segmental resection with an end to end anastomosis or by extra-mucosal suture invaginating the gangrenous segment (Figure 2). This operative technique proposed by Horbach et al., which avoids bowel resection, is performed only under certain conditions; the gangrenous segment does not extend across more than half the circumference of the gut and its margins are clearly healthy [4].

CONCLUSION

The two cases presented illustrate the picture of the Richter’s hernia whose clinical signs are misleading. In presentation of any groin swelling, the need for an early and accurate diagnosis followed by prompt treatment cannot be overemphasized. The surgical treatment remains in relation with local and parietal conditions.

Figure 1: (A–D) Lateral incarceration of part of the circumference of the distal ileum.

Figure 2: (A–B) Invaginating a Richter’s hernia.
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REFERENCES
Childbearing at very advanced maternal age, the challenges and complications: A report of two cases

Felix Mwembi Oindi, Evan Sequeira, Steve Kyende Mutiso

ABSTRACT

Introduction: The desire for childbearing at very advanced maternal age (maternal age ≥45 years at the time of delivery) is becoming increasingly common. This has resulted in increased demand and utilization of assisted reproductive technologies to counteract the declining fecundity associated with the advanced maternal age. However, the older gravidas are at increased risk of having various medical conditions with potential adverse impact on their pregnancies making such pregnancies be considered as high risk pregnancies. Case Report: We present two cases of very advanced maternal age pregnancies (54-year-old and 49-year-old), both conceived through assisted reproductive technology, we successfully managed through their pregnancies outlining the conception challenges, pregnancy complications and the subsequent management and outcomes. We further undertake a literature review to assess the challenges and outcomes of pregnancies at very advanced maternal age. Conclusion: Childbearing at very advanced maternal age is challenging from conception to delivery. Owing to the age related decline in fertility, most of these women require assisted reproductive technology (ART). Moreover, they are more likely to have adverse pregnancy outcomes principally stemming from preterm births with a resultant greater maternal and perinatal mortality and morbidity. With the increasing trend towards delayed child bearing, and owing to the anticipated adverse pregnancy outcomes, the hospitals and physicians needs to equip themselves for the increased demand for ART and the need for sophisticated prenatal, perinatal and postpartum care.

Keywords: Anti-mullerian hormone (AMH), Assisted reproductive technology (ART), In vitro fertilization (IVF), Very advanced maternal age (VAMA)

INTRODUCTION

The desire for childbearing at very advanced maternal age (defined as maternal age ≥45 years at the time of delivery) is becoming increasingly common [1–3]. This is partly due to better access to safe, effective and reversible contraception, longer education, higher career goals, later marriage, desire for financial stability and the advances in reproductive technology [3–5]. The
increased maternal age is associated with a non-linear decline in fecundity increasing the need for reproductive assistance. Indeed, advances in reproductive technology such as oocyte donation and fertility preservation through oocyte cryopreservation, to counteract the age related decline in fertility has made pregnancy possible even at very advanced maternal age [6].

The older gravidas are more likely to have medical conditions such as obesity, hypertension and diabetes mellitus which could complicate their pregnancies [7, 8]. In addition, they are at increased risk of adverse pregnancy outcomes with resultant increase in maternal and perinatal mortality and morbidity [7]. These include ectopic pregnancy, spontaneous miscarriage, fetal chromosomal abnormalities, placenta praevia, gestational diabetes, preeclampsia, multiple births, preterm delivery and cesarean section [2, 6]. Moreover, unlike previous trends where mothers at VAMA were of high gravidity, current mothers are more likely to be of lower gravidity but with a higher socio-economic status and as such can access any possible health care of their choice [2]. Health care providers need to equip themselves for this upcoming challenge of VAMA to help meet the maternal need of having a baby.

CASE SERIES

Case 1

A 49-year-old female, para 0+1 lady had presented with a five-year history of subfertility. Her past history was significant for multiple uterine fibroids for which she had undergone two open myomectomies with the most recent being two years prior to her presentation. She had also been successfully medically managed for hyperprolactinemia three years prior. There was a remote history of an elective pregnancy termination during her early twenties with no associated complications. She had no history of hypertension and her initial blood pressure was 110/65 mmHg. Her fertility assessment was as follows. A hysterosalpingogram showed an irregular endometrial cavity with an endometrial polyp for which she underwent a hysteroscopic polypectomy. The fallopian tubes were patent bilaterally. The rest of her fertility workup was as follows: a normal seminalysis (volume: 3 ml, concentration: 70 million/ml, total motility: 35%, vitality: 65% and >14% normal forms) and an anti-Mullerian hormone (AMH) level of 0.6 ng/ml (low fertility range).

The patient had a natural conception which ended in 23 weeks gestation and severe pre-eclampsia at 28th week with an admitting blood pressure of 160/110 mmHg. The investigations done revealed an elevated urine albumin creatinine ratio of 64.5 mg/mmol (macroalbuminuria range) with elevated liver transaminases (aspartate aminotransferase (AST) 122 units/liter; Alanine aminotransferase (ALT) 99 units/liter). The full blood count revealed normal parameters with hemoglobin of 11.6 g/dl and a platelet count of 157,000/ml.

The patient received magnesium sulfate for seizure prophylaxis (4 g slow intravenous infusion followed by 1 g per hour maintenance dose until 24 hours post-delivery) and oral labetol 200 mg thrice daily for blood pressure control. Two doses of betamethasone 12 mg were administered intramuscularly 24 hours apart for fetal lung maturation after which delivery was performed via cesarean section. The babies weighed 1.60 kg, 1.07 kg and 1.22 kg at birth and were admitted to the neonatal high dependency unit.

The liver function tests gradually improved post-delivery normalizing by the fifth day (AST: 34 units/liter, ALT: 28 units/liter) with normalization of blood pressure. Labetalol was stopped two weeks post-delivery and patient encouraged to continue having daily blood pressure monitoring at home. The painful vulval swelling and the elevated blood pressure resolved by two weeks post-partum. The blood pressure at the six week post-natal review was 110/70 mmHg. The babies did well in the nursery and were allowed home at 2 kg body weight by 1 g per hour maintenance dose until 24 hours post-delivery normalizing by the fifth day (AST: 34 units/liter, ALT: 28 units/liter) with normalization of blood pressure. Labetalol was stopped two weeks post-delivery and patient encouraged to continue having daily blood pressure monitoring at home. The painful vulval swelling and the elevated blood pressure resolved by two weeks post-partum. The blood pressure at the six week post-natal review was 110/70 mmHg. The babies did well in the nursery and were allowed home at 2 kg body weight as per the hospital guidelines for preterm babies and are currently doing well. The mother remained normotensive during her post-natal follow-up to the last review six months post-delivery.

Figure 1: Ultrasound showing the three fetuses (fetus A, B and C) in different amniotic sacs as shown by the arrows.
Case 2

A 54-year-old female, para 0+1 presented with a four-year history of subfertility. She previously had a missed miscarriage for which she underwent a manual vacuum aspiration about 20 years prior to her presentation. There were no previous hospitalizations or other surgical procedures. The blood pressure on initial assessment was 110/70 mmHg.

Fertility assessment of the patient was as follows; the HSG showed bilateral tubal blockage, the hormonal profile was in the post-menopausal range (AMH level of below 0.3 ng/ml and FSH level of 60.14 μIU/ml) while the seminal analysis was essentially normal. She achieved a successful singleton pregnancy following an IVF cycle with donor eggs. Her antenatal profile was unremarkable (Hb 12.5 g/dl, HBsAg Negative, HIV Negative, VDRL Negative and blood group O and Rhesus factor positive). The trisomy (18 and 21) and neural tube defect screening classified her as low risk with a normal anomaly ultrasound scan at 19 weeks gestation.

At 34th week, the patient developed severe gestational hypertension (blood pressure 180/110 mmHg), and the growth scan showed severe fetal growth restriction (FGR) necessitating her delivery. She received a 20 mg intravenous bolus of labetalol after which her blood pressure dropped to 160/90 mmHg after which it was controlled on oral labetalol 200 mg thrice daily. The laboratory tests revealed no proteinuria (urine albumin creatinine ratio of 1.5 mg/mmol), with normal full blood count and liver function tests. In addition, she received antenatal steroids for fetal lung maturation (2 doses of betamethasone 12 mg administered 24 hours apart).

Induction of labor was commenced with dinoprostone 3 mg administered per-vaginally every six hours after the second dose of betamethasone. The patient subsequently underwent an urgent cesarean section after a failed induction having had no cervical changes after the second dose of dinoprostone. The outcome was a live infant with a birth weight of 1.46 kg. The baby was transferred to the neonatal high dependency unit for further management till discharge. The baby did well and weighed 4.8 kg at the 10th week postnatal review. The mother’s blood pressure settled by sixth week post-delivery (measured value of 110/60 mmHg). The patient has remained normotensive during her follow-up post-delivery and is currently on lifestyle measures to maintain her normotensive state.

DISCUSSION

The number of women seeking to achieve a pregnancy at very advanced maternal age (VAMA) is progressively increasing [1, 2]. This is due to various factors [4, 5, 9] making delayed childbearing possible and desirable at such ages. Some of these factors include better access to contraception, higher career goals and the advances in reproductive technology. In Tanzania for instance, the number of women giving birth after the age of 35 increased from 10.3% to 14.5% over a seven-year period (2005–2011) [10]. This has similarly been observed in the India, USA, Norway and South Africa where there is a general increase in the number of women giving birth at an advanced age (maternal age >35) [2, 4, 11, 12].

Very advanced maternal age is uncommon in communities where earlier marriages is the norm. In these communities, pregnancies at VAMA most often represent the final or one of the final births for women who continue to child-bear until menopause for social or cultural reasons. Such women are more likely to be of a lower socioeconomic status and of a high parity. This is in contrast to the picture in many countries where advanced and even very advanced maternal age is notably on the rise. Women presenting at VAMA from these communities are more likely to be of a lower parity and of a higher socio-economic status and hence have access to high standards of healthcare and interventions. As such, they are more likely to use ART and generally have more favorable pregnancy outcomes with less still births [7]. Both our clients fall in this latter category. They were of a lower parity and of a high socio-economic status and were willing to incur whatever costs necessary to enable them get a baby.

The very advanced maternal age is associated with a decline in fecundity [6]. This is due to in part the declining ovarian reserve as evidenced by low levels of anti-Mullerian hormone (AMH) and antral follicular counts (AFC) which are proxy indicators of the ovarian reserve [13]. Both of our patients had AMH levels in the low fertility range. Moreover, there is a greater occurrence of uterine and tubal anomalies with greater maternal ages. The uterine pathologies include endometrial polyps and uterine fibroids whose occurrence is greater with advancing maternal age [14]. Fibroids, especially those with a sub mucosal or intramural component are associated with low fertility. Myomectomy would result in uterine scarring and synechiae further worsening the subfertility. This can be ruled out through hysteroscopy which is useful for diagnosis and treatment of the intrauterine adhesions [15]. One of our clients had undergone two previous myomectomies and was found to have an endometrial polyp which was hysteroscopically resected after which she was able to conceive through ART. This was possibly contributing to her inability to conceive.

Tubal factors are another major cause of subfertility accounting for up to 50% of the subfertility causes [16]. Their occurrence has been shown to be equally high in the VAMA group [7] possibly due to a greater lifetime chance for genital infections to occur, a major cause of tubal blockage [17]. One of our clients had bilateral tubal blockage on HSG potentially making natural conception difficult.

Other contributory factors for low fecundability at VAMA is the reduced coital frequency owed to declining sexual desire, lubrication difficulties and the
Our patients were hospitalized in the third trimester and reported to be high due to these pregnancy complications. The hospitalization rates during pregnancy have been positive for gestational diabetes [4, 11]. Moreover, gestational diabetes though none of our patients screened as aspirin) which our patients were on [21].

to reduce severe pre-eclampsia such as Ascard-75 (junior aspirin) [2]. This necessitates close follow-up and interventions for the increased demand for ART and the need for sophisticated prenatal, perinatal and postpartum care with the goal of meeting the maternal need of having a baby.

CONCLUSION
Childbearing at very advanced maternal age is challenging from conception to delivery. Most of these women are subfertile requiring assisted reproductive technology (ART) and are more likely to have adverse pregnancy outcomes principally resulting from preterm births with a resultant greater maternal and perinatal mortality and morbidity. With increasing trend towards delayed child bearing, the society needs to equip itself for the increased demand for ART and the need for sophisticated prenatal, perinatal and postpartum care with the goal of meeting the maternal need of having a baby.

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Author Contributions
Felix Mwembi Oindi – 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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The corresponding author is the guarantor of submission.

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Authors declare no conflict of interest.

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REFERENCES

A case of tuberculosis in the appearance of a metastatic tumor

Yılmaz Baş, Ali Bulgan, İkram Abdikarim İbrahim

ABSTRACT

Introduction: We report a case of chronic osteolysis of the ribs, iliac bones and vertebrae, Pott's abscess due to tuberculosis that was previously misdiagnosed as cancer metastasis on computed tomography imaging. Case Report: We report a rare case of multiple vertebrae, ribs and iliac bones lesions associated with Pott's disease and psoas abscess caused by tuberculosis in a 28-year-old female. This case illustrates an exceptional location of osteoarticular tuberculosis with Pott's disease and supports the diagnostic difficulties encountered: (i) bone imaging is not specific, in view of if cancer metastasis is suspected, (ii) sampled from the rib lesion with fine needle aspiration biopsy, (iii) there is psoas abscess drained. Conclusion: Tuberculosis should come to mind in patient with multiple bony lesions, especially in endemic countries such as Somalia. Additionally, we recommend cytohistomorphological testing for any tuberculosis cases.

Keywords: Bone and joint infections, Pathology, Respiratory infections, Tuberculosis

INTRODUCTION

Tuberculosis remains a world-wide problem for more than a century ago, and it affects all age groups [1]. In Somali Regional State, Eastern Ethiopia, a total of 31,198 all types of new tuberculosis cases were registered and reported from 2003–2012 had a total population of 4,445,219 [2], and the estimated incidence rate in Sub-Saharan Africa is also nearly 700 cases per 100,000 populations [3]. In 2014, the World Health Organization (WHO) identified nine million new cases of the disease with 1.5 million deaths [1].

Vertebra-localized tuberculosis is known as Pott's disease and psoas abscesses are suppurative collections within the fascia surrounding the psoas and iliacus muscles [3, 5]. Clinically and radiologically, it can be confused with cancer metastatic lesions as a result of multifocal bone involvement [6]. The diagnosis of such cases is made by evaluating the clinical, microbiological, radiological and histopathological findings collectively...
Histopathological findings are very important in diagnosis [5]. A case of multiple bone lesions and with psoas abscess, presented in the literature because of its clinico-radiological presentation as a cancer metastasis.

CASE REPORT

A 28-year-old Somali female was admitted to the Somali Mogadishu-Turkey Training and Research Hospital general surgery service for abdominal pain, subcutaneous left anterior chest wall swelling and sensitivity, lumbovertebral sensitivity, backache, severely pale, sweating, fever, and weight loss.

Historically, she had a negative history of contact with active tuberculous and not received the Bacillus Calmette-Guérin (BCG) vaccine. Patient’s family had never been diagnosed with tuberculosis. She was not diabetic or immunosuppressed and she denied any trauma to the site. Physical examination revealed swelling of the tissue overlying the ribs. The vertebral-spines, ribs area and left thigh were firm and painful to touch. And she had moderate kyphosis. There was no BCG scar.

Computed tomography (CT) scan of the chest, abdomen and iliac bone confirmed multiple expansive, destructive hypodense mass or abscess lesion on the right 1st, 2nd, 4th ribs, the left 1st, 2nd, 6th ribs and 1st, 2nd, 6th, 10th, 12th thoracic vertebrae, 1st, 2nd, 3th, 4th lumbar vertebrae, the left and the right iliac bones (Figure 1A–D). Additionally, there was a 8x7 cm hypodense area compatible with abscess or hematoma on the left psoas muscle and along the left iliopsoas muscle.

Laboratory tests of the patient showed leukocytosis at 15,000/mL, erythrocyte sedimentation rate (ESR) 65 mm, CRP 38 mg/l and CEA 1.5 ng/ml. Other laboratory tests were within normal limits. No tuberculin skin test was performed. HIV and Brucella serology was negative.

A fine needle aspiration biopsy (FNAB) of the 6th rib lesion was performed. Cytopathological examination revealed an epithelioid and giant cell granulomatous inflammatory process consistent with tuberculosis (Figure 2). An ultrasonography (US) guided percutaneous drainage of the abscess was performed. Purulent cream colored liquid was drained (Figure 3A). Slides of cell block were prepared from the drainage material. The Ziehl–Neelsen staining of aspiration was positive (Figure 3B).

DISCUSSION

Tuberculosis is a disease that remains endemic in many parts of the world particularly in developing countries [1]. Immune deficiency, low socio-economic level, dialysis, transplantation, malignancy pathology, infants, and malnutrition are essential risk factors [1, 5]. In addition, there are populations who do not have access to BCG vaccine in regions such as Somalia. In our case, low socio-economic status factor was present and also did not have BCG scar.

In countries such as Somali, where tuberculosis is endemic, and the diagnosis is traditionally made on clinical and radiological grounds. Histopathological confirmation is not usually undertaken, because it is expensive and the cytopathology department has not yet settled in many centers in spite of there are the large case load. Therefore, we performed FNAB at the lesion in the rib because of our pathology department in Somali Magodishu-Türk Hospital has enough facility.
In the study of S Bhojraj et al. [7] biopsy for pathologic examination was not attempted in patients with osteitis. Most of their patients (93%) were diagnosed and treated satisfactorily on the basis of clinical and radiological evidence, and without histopathological diagnosis. According to them study, a therapeutic trial of antituberculous treatment is a practical alternative to taking a biopsy [7]. But as noted in Urbanczik's study [5] in bone samples from areas other than the spine, histopathology remains the gold standard technique disclosing the classical caseating tubercle granulomas. Our case was diagnosed according to radiological and cytomorphological findings.

Total 14,097 tuberculosis cases reported to Centers for Disease Control (CDC) in 2005 [4]. There were 329 (11.1%) patients with bone and/or joint involvement, and ranks third after the pleural and lymphatic tuberculosis [4]. In study of Lemnouer et. al. [1], osteoarticular tuberculosis has found the fourth most prevalent tuberculosis localization after pulmonary, urogenital and lymphatic.

In our case, the patient had multiple expansive, destructive lytic, mass or abscess lesion vertebræ, ribs, iliac bones with psoas abscess. The lung and other organ involvement were not observed.

Tuberculosis is second only to metastatic neoplasms as a cause of destructive multifocal bone lesions such as ribs lesions [6]. Typhoid fever or paratyphoid fever, actinomycosis, syphilis, infections due to streptococci or staphylococci, coccidioidomycosis, blastomycosis, and brucellosis are other infectious etiologies [6]. Malignancies include metastases from primary carcinoma of the liver, breast, thyroid, and kidney; Ewing’s sarcoma; fibrosarcoma; multiple myeloma; or histiocytosis [6, 8]. Cytomorphological examination revealed no malignant cells in our case, and Brucella serology was negative.

Vertebral tuberculosis, which has two different patterns, is a chronic, slow-progressive disease [8]. The first is spondylitis without disc involvement, which is exceedingly more common, and multilevel vertebral body involvement could observe in this form as skip lesions [8]. The second pattern is destruction of two or more contiguous vertebræ associated with late-onset disc infection, which results in intervertebral space narrowing due to disc herniation into the collapsed vertebral body [8].

Spinal spondylitis is the most common manifestation of osteoarticular tuberculosis, and 1–3% of patients with tuberculosis have skeletal involvement [9]. The lumbar spine is the most common site of the disease followed by the thoracic region [3]. In metastatic disease, thoracic region is most commonly involved, while posterior wall of the vertebral body (60%), pedicles and lamina (50%) are involved. However, intervertebral disc heights are preserved [3].

In multiple vertebral body involvement, the characteristic gibbous deformity and paraspinous mass or collection are common. Posterior element involvement is a characteristic of vertebral tuberculosis [8]. In this case, the patient has lesions at T2nd, T10th, T12th, L1st, L2nd, L3th corpuses, T5th right pedincule, L1st left pedincule and spinous process, L4th left pedincule expansive, destructive lytic, mass or abscess lesion. When severe, distinct loss of vertebral height can lead to moderate kyphosis with facet joint subluxation or dislocation. Preservation of disc space as well as posterior element abnormalities and multiplicity of vertebral body involvement in tuberculosis may lead to difficulty in distinguishing tuberculosis from tumoral processes. Early symptoms of vertebral tuberculosis are relatively nonspecific, and include backache, vertebral sensitivity, fever, and weight loss [8].

In the patient, there was backache, sweating, weight loss, tenderness in ribs and vertebrae. She had moderate kyphosis.

Tuberculosis of the ilium constitutes less than 1% of all skeletal tuberculosis [8]. A typical radiological feature of tuberculosis in cancellous bones is feathera sequestra or lytic lesions with irregular margins associated with mild surrounding sclerosis [8]. In our patient has multiple expansive, destructive lesions on the left and the right iliac bones.

Tuberculosis of the rib occurs in males about twice than females [6]. This infection occurs most often in patients between 15 and 30 years of age which occurs most often in children between the ages of 2 and 10 years [6]. Our patient was a 28-year-old female.

Rib involvement within skeleton tuberculosis is at 1.7%, however, it is the second widespread disease next to metastatic lesions among the destructive lesions of the ribs [10]. Hence, physical and radiological examination findings can often be confused with primary and metastatic chest wall tumors [6]. Rib tuberculosis frequently emerges until 18 months after infection, and <50% of patients have active pulmonary disease [6, 10]. Therapy is based on 9–12 months of antituberculous chemotherapy [1]. The disease may be drug-resistant, however, and therefore a poor response to standard hemotherapy at the end of 6–12 weeks does not imply that surgery is indicated [7]. In our case, the patient has multiple expansive, destructive mass or abscess lesion on the right 1st, 2nd, 4th ribs and on the left 1st, 2nd, 6th ribs. Likewise, tuberculosis cases with rib involvement with Pott’s disease are rare too [7].

Iliopsoas abscess may be classified as primary or secondary, and primary iliopsoas abscess occurs probably as a result of hematogenous spread of an infectious process from an occult source in the body [11]. Crohn’s disease, urinary tract infections, vertebral osteomyelitis, infected aortic aneurysm, endocarditis, intrauterine contraceptive device, suppurative lymphadenitis etc. are also associated with secondary to iliopsoas abscess [11]. Treatment involves the use of appropriate antibiotics along with drainage of the abscess [11]. Drainage of an abscess in iliopsoas compartments may be indicated if it causes symptoms which are simply due to its size.
and location, such as a flexion deformity of the hip [7]. A neurological deficit is not an absolute indication for surgery, and responds well to conservative treatment [7]. However, the mortality rate increases with delayed and incorrect diagnoses and reaches 100% in cases without drainage [12]. Our patient denied any trauma and had an abscess formation, starting from the T12 vertebra and extending up to thigh. The case in light of these findings has been investigated as a secondary Pott’s disease. Abscess drainage was performed and tuberculosis treatment was started. There was no neurological deficit on physical examination. But, as noted in the study of Woldeyohannes et al. [2] treatment completion rate in Somalia was relatively lower than the value in the world, Europe, Turkey and report from Arsi Zone, Central Ethiopia.

CONCLUSION

To conclude, there are population that do not have access to BCG vaccine in these regions such as Somali. We have shown the clinicoradiological findings of a case with multifocal osseous, initially misdiagnosed as cancer metastases. Familiarity with the imaging features of musculoskeletal tuberculosis and a high index of clinical suspicion are necessary for the correct diagnosis and proper treatment. Histopathological findings are also very important in diagnosis.

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Author Contributions

Yılmaz Baş – 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

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.
Paraplegia as initial presentation of acute myeloid leukemia: A case report

Adama Isah Ladu, Aisha Abba Mohammed, Halima Talba, Yakaru Kundili, Yakura Abba Kawu, Ridhwan Aliyu Tukur

ABSTRACT

Introduction: Acute myeloid leukemia (AML) represents a state of dysregulated clonal expansion of immature myeloid progenitor cells with arrest of differentiation at a particular stage. Involvement of the central nervous system (CNS) may result acutely from the disease at the initial diagnosis, and the symptoms range from muscle weakness, paresthesias, and hypoesthesia to fecal and urinary incontinence. Paraplegia is an extremely rare initial presentation of undiagnosed leukemia. Case Report: A 15-year-old girl presented with acute urinary and fecal incontinence, and a day later developed inability to move both lower limbs. There was two weeks preceding history of fever, malaise and lower back pain, no associated history of paresthesias or numbness. Examination of both lower limbs revealed decreased tone and reflexes with bilateral extensor reflexes and power of 0/5. There was a loss of sensation around the perianal region with absent anal tone. Cranial nerve and sensory examination were normal. Both peripheral blood film and bone marrow aspiration cytology stained with Leishman stain were consistent with acute myeloid leukemia (AML) FAB M-4 subtype. Cerebrospinal fluid analysis was negative for malignant cells. Magnetic resonance imaging of the lumbosacral region was obtained; both T1- and T2-weighted sequences showed no signal intensity in the spinal cord, theca and cauda, with normal signal intensity involving the intervertebral disc. Patient was commenced on supportive therapy and induction chemotherapy using daunorubicin 45 mg/m² days 1–3 and cytosine arabinoside 200 mg daily days 1–7. However, patient succumbed to her disease and passed away 10 days into treatment as a result of respiratory complication. Conclusion: Central nervous system involvement is a rare initial presentation of AML. Absence of abnormality on MRI scan does not rule out spinal cord involvement; consequently late recognition may delay treatment and lead to irreversible neurological complication.

Keywords: Acute myeloid leukemia, Central nervous system, Paraplegia

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INTRODUCTION

Acute myeloid leukemia (AML) represents a state of dysregulated clonal expansion of immature myeloid progenitor cells with arrest of differentiation at a particular stage, it accounts for 15–30% of all cases of newly diagnosed cases of leukemia [1]. The condition is more frequently encountered in infants and adolescents. Clinical features include pallor, easy fatigability, lethargy, bleeding and fever. Organ infiltration is less frequent compared to acute lymphoblastic leukemia (ALL). However, AML is more likely to produce aggregates of leukemic cells known as chloroma; these are frequently encountered in the head and neck region, the central nervous system (CNS), where they may produce neurological symptoms [2]. Symptoms range from muscle weakness, paresthesias, hypoesthesia to fecal and urinary incontinence. Paraplegia due to epidural mass is an extremely rare presentation of undiagnosed leukemia [3]. The thoracic region is the most frequently involved region, followed by the lumbar region. The sacral cord is the rarest region implicated [4, 5]. Here, we present a case of AML presenting with paraplegia at the initial diagnosis.

CASE REPORT

A 15-year-old girl was seen by the medical team with history of fever, malaise and lower back pain, she had two units of blood transfused along with antibiotics and antimalarials for severe malaria and presumed septicemia, however, two weeks later, she developed acute urinary and fecal incontinence, and a day later developed inability to move both lower limbs. There was no preceding history of trauma; no associated history of paresthesias, numbness, visual blurring. On physical examination, she was febrile at 38.2°C, pale, anicteric. No lymphadenopathy was noted. Her chest was clear to auscultation. Cardiovascular and abdominal examinations were unremarkable. There was vertebral tenderness involving the lower lumber to sacral region. The examination of both lower limbs revealed decreased tone and reflexes with bilateral extensor reflexes and power of 0/5. There was loss of sensation around the perianal region with absent anal tone. Cranial nerve and sensory examination were unremarkable. There was vertebral tenderness involving the lower lumber to sacral region. The examination of both lower limbs revealed decreased tone and reflexes with bilateral extensor reflexes and power of 0/5. There was loss of sensation around the perianal region with absent anal tone. Cranial nerve and sensory examination were normal.

Complete blood count revealed hematocrit of 21%, total white cell count of 29.3x10⁹/L (myeloblast 47%, neutrophil 29%, eosinophil 1%, lymphocyte 20%, monocyte 3%) and platelet 90x10⁹/L. Peripheral blood film (PBF) showed leukocytosis with variable number of myeloblast (Figure 1A). Bone marrow aspiration (BMA) cytology showed a hypercellular marrow with maturation arrest along the myeloid cell line. The myelogram showed a myeloblast count of >20% with maturation arrest at the myelocytic stage, some of the blast cells contained Auer rods (Figure 1B), megakaryocytes were reduced. Both PBF and BMA were consistent with acute myeloid leukemia (AML) FAB M-4 subtype. Cerebrospinal fluid analysis (CSF) analysis was negative for malignant cells. A lumbosacral X-ray was normal. Magnetic resonance imaging of the lumbosacral region was obtained in the axial, sagittal and coronal view. Both T1- and T2-weighted sequences showed no signal intensity in the spinal cord, theca and cauda, with normal signal intensity involving the intervertebral disc (Figure 2).

An assessment of cauda equina syndrome secondary to leukemic infiltration was made. The patient was commenced on supportive therapy and induction chemotherapy using daunorubicin 45 mg/m² days 1–3 and cytosine arabinoside 200 mg daily days 1–7. However, patient succumbed to her disease and passed away 10 days into treatment as a result of respiratory complication.

DISCUSSION

The exact incidence of central nervous system (CNS) involvement in patients with AML is unknown, but is lower compared with the incidence in children and adults with ALL; for that reason routine CSF examination is not performed as part of the diagnostic workup in asymptomatic patients [6, 7]. Involvement of the CNS may result acutely from the disease at the diagnosis, as in the index case; from relapse of the disease or from complications of agents used in treatment. It may be...
classified into three as CNS 1 (when there is no blast in the CSF), CNS 2 (< 5 WBC/μl of CSF with blast cells) and CNS 3 (> 5 WBC/μl of CSF with blast cells or signs of cranial nerve palsy, meningeal involvement) [8]. The presence of certain factors has been associated with increased risk of neurological complication. These include hyperleukocytosis, high level of lactate dehydrogenase and high expression of CD56 [7]. In a single institution study of 290 pediatric patients with AML, the authors found significant relation between CNS involvement in patients with AML having the cytogenetic abnormality Inv 16, t [9], or AML M4 and M5 subtype. The authors concluded that CNS involvement is common in those with favorable cytogenetics [8]. As mentioned above, the PBF and BMA cytology of the index patient was in keeping with AML M4 subtype, and therefore belonged to a high risk group for CNS involvement.

Several mechanism have been put forward to explain the pathogenesis of CNS involvement in leukemia, this include contamination of the CSF through the choroid plexus or infiltration of the cerebral parenchyma through brain capillaries; direct extension of aggregates of leukemic cells called chloroma, from the bone marrow through the cortical bones into the spinal cord, may cause symptoms as a result of mass effect on neural tissue [9]. Hyperleukocytosis with thrombosis may result in leukostasis and consequently poor CNS perfusion. Intracranial hemorrhage with focal neurological deficit may also arise. Hemorrhage into the spinal canal can occur following diagnostic procedures in cases presenting with thrombocytopenia [9]. In the index case, the CSF was negative for malignant cells and the MRI scan showed no obvious pathology. The presence of urinary and fecal incontinence indicates pathology involving the sacral canal. Relatively small tumor in this region may manifest with profound symptoms, unfortunately such tumors can be particularly difficult to image [9]. Consequently, the absence of findings on MRI scan does not completely exclude cord compression as seen in the index case report.

The involvement of CNS may be without symptoms. In symptomatic patients, the clinical picture will depend on which part of the CNS is involved. Features of advanced disease include irritability, headaches, seizures symptoms of raised intracranial pressure and cord compression [7, 9]. Amongst the leukemia, AML is the most common leukemia to produce cord compression; however, it is a rare initial presentation of AML [4]. The patient may presents with pain in the back, abdomen, lower limbs and perianal region. Muscle weakness to paraplegia may also be present. Fecal and urinary incontinence often represents dangerous signs as in all cases of cord compression [10]. Therefore, the acute physician needs to have a high index of suspicion of AML in young patients presenting with neurological symptoms, and make every effort to diagnose it. Early identification and prompt intervention can prevent the development of irreversible neurological complications. This is reflected in this case report, where the patient had developed irreversible neurological deficit prior to referral to the hematology team. Therefore, the presence of laboratory evidence of AML can be used to commence active treatment and prevent further delay.

**CONCLUSION**

In conclusion, central nervous system involvement is a rare initial presentation of acute myeloid leukemia. Several pathogenic mechanisms have been implicated including direct extension of leukemic aggregate resulting in compressive effect on the neural tissue. The absence of abnormality on magnetic resonance imaging (scan) does not rule out spinal cord involvement. Consequently, late recognition may delay treatment and lead to irreversible neurological complication.

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**Author Contributions**

Adama Isah Ladu – 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**
The corresponding author is the guarantor of submission.

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Severe hypernatremia in an adolescent male with anorexia nervosa

Kene Ebuka Maduemem, Comfort O. Adedokun

ABSTRACT

Introduction: Anorexia nervosa is a commonly encountered cause of severe underweight in pediatric settings. It is an eating disorder characterized by the inability to maintain a minimally normal weight, a devastating fear of weight gain, relentless dietary habits that prevent weight gain, and a disturbance in the way in which the body image is perceived. Hypernatremia is an unusual electrolyte imbalance in anorexia nervosa. Case Report: This is a case of a 12-year-old male admitted with progressive weight loss following restrictive food intake for the preceding one year. He presented with severe hypernatremia. Correction of severe hypernatremia was successful after four to five days. This correction was slow but gradually achieved. There was no altered sensorium throughout admission which remains the worst nightmare of any managing team. Feeding protocol as per Junior MARSIPAN guidelines was adhered to. He made an excellent recovery and was discharged after 24 days of admission with a weight gain of 6.1 kg. Conclusion: Anorexia nervosa is commonly encountered in pediatric settings and can cause potentially life-threatening physical and psychological complications. Hypernatremia is an uncommon metabolic abnormality; slow correction averts untoward side effects.

Keywords: Anorexia nervosa, Hypernatremia, MARSIPAN guidelines

INTRODUCTION

Diagnostic and Statistical manual of Mental Disorders, Fifth edition (DSM-V), published in May 2013 revised the definition of anorexia nervosa from DSM-IV by removing amenorrhea as one of the criteria for diagnosis while focusing more on behaviors [1]. It is a commonly encountered cause of severe underweight in pediatric settings that can cause potentially life-threatening physical and psychological complications [2]. Hypernatremia is an unusual manifestation in anorexia nervosa. It is caused by net water loss (increased loss or decreased intake) or, rarely, sodium gain.

Herein, we describe a case of a 12-year-old boy admitted with progressive weight loss following restrictive food intake for the preceding one year. He presented with severe hypernatremia.
CASE REPORT

A 12-year-old boy was referred by his general physician on account of a week history of progressively worsening dizziness and lethargy. He had been refusing feeds for the last three months with dramatic weight loss. This is on a background of the desire to be ‘skinny’ for the preceding two years. There was a history of hiding food. Parents were suspicious of self-induced vomiting but never witnessed. No known use of laxatives or purgatives. Past medical history was insignificant. He has been a very well adolescent who gets on very well with siblings and friends. He looked up to Mo Farah (a British athlete) as his mentor.

He lived with parents and has two older siblings (16-year-old sister and 14-year-old brother). He was in 6th class and academically sound. He was very athletic: engages in at least one sporting activity per day during school year. No history suggestive of bullying in school. However, noted to have been called ‘fat’ by a classmate in 2nd class.

Examination revealed a cachectic, lethargic, pale looking and dehydrated adolescent male. Vital signs were: temperature 36.4 °C, heart rate 40/min, respiratory rate 15/min, supine blood pressure 108/66 mmHg, standing blood pressure 98/58 mmHg. Other significant findings were prominent ribs, scaphoid abdomen, dry, scaly skin, lanugo hair on arms, yellow palms. Weight was 32.4 kg, height 156.6 cm, body mass index (BMI) 13 kg/m²

A diagnosis of restrictive type anorexia nervosa was made. He was admitted for fluid resuscitation. Vital signs were continuously monitored with ECG monitor. Referral was sent to CAMHS, medical social work and dietician. He was strictly managed following the Junior MARSIPAN guidelines. There was a steady improvement in demeanor and weight.

Urgent electrocardiogram confirmed sinus bradycardia; heart rate of 38 beats per minute, normal QTc 420 ms (Figure 1). Admitting blood glucose was 8.2 mmol/L. Serum electrolytes and urea levels were deranged (sodium 182 mmol/L, potassium 2.7 mmol/L, chloride 143 mmol/L, urea 17.2 mmol/L). Creatinine: 82 umol/L. Calcium 2.42 mmol/L, Magnesium 1.28 mmol/L, phosphate 0.95 mmol/L. Hemoglobin 14.2 g/dl, White cell count 8.2 per μL (neutrophil 2.79, lymphocyte 4.92). C-reactive protein <0.2 mg/L. Venous blood gas: pH 7.38, pCO2 7.6, bicarbonate 33.7, base excess 8.6. Serum osmolality 369 mosm/kg. Serum ferritin 223 ng/ml, vitamin B12 650 ng/L, serum folate 5.8 mg/ml. Spot urine sodium 305 mmol/L. Thyroid function test was normal. Celiac serology was negative.

Initial rehydrating fluid and regime was 5% dextrose in 0.9% saline. Aim of correction of hypernatremia was 0.5 mmol/l/h or 12 mmol/l/day. Potassium chloride was added in the fluids (20 mmol/500 ml). Serum electrolytes were checked every 4–6 hours. Serum calcium, phosphate and magnesium were checked daily to prevent refeeding syndrome.

Serum sodium level decreased from 182 mmol/L to 174 mmol/L at the end of the first 24 hours of admission (drop of 8 mmol/L). This gradually and steadily decreased to baseline on day-5 of admission to 140 mmol/L (Figure 2).

Oral feeding was gradually initiated by dietician. Sodium level returned to normal on day-5 of admission.

Figure 1: Electrocardiogram on admission showing sinus bradycardia with heart rate of 38 beats per minute.

Figure 2: Trend of serum sodium and chlorine during the first five days of admission.
This correction was gradual and painstaking. Intravenous fluid was completely weaned off by the end of the first week. Twice weekly weighing was adhered to.

Daily ECG showed gradual improvement of bradycardia. Dietician plan was based on refeeding meal with initial daily calorie intake of 400 kcal which was gradually increased to 900 kcal within four days. Oral fluid intake comprising milk only was successively increased from 500 ml to over 800 ml within three days. The caloric requirement for his age was worked out to be 2250 kcal/day. On discharge, his daily caloric intake was 2000–2200 kcal.

The patient made a remarkable recovery. He was discharged after 24 days of admission. Weight on discharge was 39.5 kg (weight gain of 6.1 kg), BMI of 16.1 kg/m². He has been reviewed severally on the assessment unit and making good progress. He has been subsequently discharged from CAMHS services.

**DISCUSSION**

Electrolyte imbalances associated with anorexia nervosa increase the morbidity level significantly. Hypokalemia is the most common electrolyte abnormality. Hyponatremia is often due to excessive water ingestion, but may also occur in chronic energy deprivation or diuretic misuse [3]. Hypernatremia can also be classified based on the intravascular volume status-euvolemia, hypervolemia and hypovolemia. Hypernatremia initially causes fluid movement out of the brain that leads to cerebral contraction and consequently manifests as altered sensorium.

Manifestations of hypernatremia vary from non-specific central nervous system symptoms such as nausea, vomiting, irritability and lethargy to confusion, seizures, myoclonic jerks, coma and even death. Severe symptoms are likely to occur with acute increase in plasma sodium levels or concentration >160 mmol/L [4]. Hypernatremia often is the result of several concurrent factors. The most prominent is fluid intake.

Our patient was restricting food intake including water. There was no history of polyuria or polydipsia to suggest a possible diabetes insipidus given the level of hypernatremia. There was no strong evidence of any diuretic abuse or any form of gastrointestinal fluid loss. In this case, severe hypernatremia was thought to be chronic than acute. Correction was done slowly over 4–5 days. The choice of fluid was normal saline with 5% dextrose as the patient was not drinking. The fluid was subsequently changed to 0.45% saline. The volume of fluid was reduced from full to 50% maintenance. There was a gradual drop in the serum sodium level (Figure 2).

In patients with hypernatremia that developed slowly, the sodium level should be corrected at a rate of 0.5 mEq/L/h, with no more than 10–12 mEq/L in 24 hours [5, 6]. The target sodium level should be 145 mEq/L. However, no prospective studies completely validate such recommendations [7].

Liam et al. published a teaching case highlighting management guidelines for chronic hypernatremia with emphasis on slower correction rate [7]. Conversely, Alshayeb et al. published a clinical investigation highlighting the association of persistent hypernatremia with increased mortality [8]. Fang et al. concluded that sodium correction rate of >0.5 mEq/L/h is associated with the development of cerebral edema [9]. By contrast, Robertson and team did not find any association between the rate of decline in serum sodium and clinical outcome [10]. He was not particularly volume depleted evidenced by his stable vital signs. Appropriately high urine osmolality will point towards a non-renale cause of euvoletic hypernatremia such as isolated hypodipsia or increased insensible losses. The former would fit into this case. The rate of correction of the hypernatremia was relatively slow but less dangerous. Feeding protocol was as per MARSIPAN guidelines to avoid refeeding or underfeeding syndromes [11].

**CONCLUSION**

Anorexia nervosa is commonly encountered in pediatric settings and can cause potentially life-threatening physical and psychological complications.

The management of anorexia nervosa is based on guidelines published by the Royal College of Psychiatrists with an emphasis on multidisciplinary approach. The goal in managing hypernatremia is to correct the water deficit in a reasonable time frame while avoiding untoward side effects.

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**Author Contributions**

Kene Ebu Maduemem – 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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The corresponding author is the guarantor of submission.

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REFERENCES

Aneurysmal changes in silent patent ductus arteriosus in a child (year time follow-up)

Aso F. Salih, Rozhin O. Qadir

ABSTRACT

Ductus arteriosus aneurysm is a saccular or fusiform dilation and elongation of the patent ductus arteriosus (PDA). It can be either congenital or acquired. Their time of presentation can be before or after treatment. In our case, aneurysmal found during her follow-up with cardiologist, it shows itself after a cardiac catheterization for trail of closure after 15 months from first catheterization. So decision was made for surgical closure rather than trial of occlusion with occluder devices. Surgery was performed after nearly one month from catheterization, under general anesthesia and median sternotomy, cardiopulmonary by-pass without complications. In conclusion, duct aneurysm may happen in any type of PDA even very restrictive one and should be treated affectively. We conclude that although PDA relatively is common and safe heart defect but they may complicate further with age, even restrictive PDA may complicate by aneurysmal PDA if left untreated and it should be consciously treated.

Keywords: Aneurysm, Catheterization, Patent ductus arteriosus

INTRODUCTION

Patent ductus arteriosus (PDA) is a vascular communication between descending thoracic aorta distal to the origin of the left subclavian artery and main pulmonary artery near the origin of the left pulmonary artery, it is an abnormal persistency of the fetal ductus arteriosus, which is an essential fetal structure, that should close spontaneously in about 90% of full-term infants during the first 48 hours after birth. It is persistency beyond a few weeks is considered abnormal [1]. It may be an isolated or occur with other congenital cardiac anomalies. Ductus arteriosus aneurysm (DAA) is a saccular or fusiform dilation and elongation of the PDA [2]. It can be either congenital or acquired as a complication of surgical closure of PDA or after a ductal infection [3, 4]. Their time of presentation can be divided in two three groups according to the age at diagnosis: infantile < 2 months, childhood (2 months to 15 years) and adult [5]. DAA has been considered as a rare congenital anomaly with potential fatal complications [5–8], although the exact incidence of DAA is unclear, but it can be ranged between 0.8–8% among fetuses and neonates [6, 9], and approximately 5.2% of all adult thoracic
aneurysm [10], with some reported childhood cases [3, 11, 12]. Some authors believe that neonatal DAA is resulted from transient widening of the ductus arteriosus prior to spontaneous obliteration and are not true aneurysm [5, 13, 14]. So the incidence of true DAA may be lower than previously believed [15]. Here we report a five-year-old child with PDA that was diagnosed incidentally at age four years, during second angiographic trial of closure found to have a DAA (pulmonary end aneurysmal), successful closure done with surgery.

CASE REPORT

A five-year-old girl with history of premature birth and normal vaginal delivery, since then she was completely normal and had regular follow-up with her pediatrician. During routine checkup at age four year, found to have abnormal heart sound on physical examination of the chest by her pediatrician without aving any signs and symptoms. Then she was referred to pediatric cardiologist, found to have a non-restrictive medium sized patent ductus arteriosus (3.3 mm) with mild left ventricular enlargement, trivial MR and mildly increased pulmonary pressure by transthoracic echocardiography without evidence of any other congenital heart anomaly.

On physical examination, she was normal for her height and weight, no any signs of respiratory distress, no cyanosis and no clubbing. Vital signs were normal. On chest examination no any deformity found, no thrill, with grade three continuous machinery murmurs in the left upper sternal boarder. Both electrocardiography and chest radiography were normal.

The patient was arranged to have a cardiac catheterization with trial of closure of the PDA if possible, her PDA was type D subtype (complex duct with multiple constrictions) with aortic side is 3.1 mm *midpoint 4.7 *pulmonary end 1.0 mm (Figure 1) but because of unavailability of occluder devices, closure was postponed.

During her follow-up with cardiologist, no any additional echocardiographic change was noticed. Only during physical examination found to have grade 4 thrill with radiation to the neck. She was arranged for another cardiac catheterization for trail of closure once occluder devices became available in our center after 15 months from first catheterization. During the second catheterization aneurysmal change seen at the pulmonary end rather than aortic end of the PDA with aortic end 3.4 mm* midpoint 7.5 mm* pulmonary end 15.9 mm (Figure 2), so decision was made for surgical closure rather than trial of occlusion with a occluder devices (Figure 3).

Surgery was performed after nearly one month from catheterization, under general anesthesia and median sternotomy, cardiopulmonary by-pass without cardioplegia or circulatory arrest. Aneurysm found at the pulmonary end, simple ligation with non-absorbable suture material then aneurysmorrhaphy done at both pulmonary and aortic end with a pericardial pledged, smooth postoperative period without any complication. Immediate postoperative echocardiography showed trivial shunt across the PDA but with a week later no shunt was seen across the PDA anymore (Figure 4).

DISCUSSION

The incidence of isolated PDA accounts for 9–12% of all CHD with higher proportion for female, with a female to male ratio 2:1 [16, 17]. Postnatal closure of PDA occurs in two stages. First stage with the contraction of the mediastinal smooth muscle in the wall of the ductus that leads to shortening and increased wall thickness. There

![Figure 1: Initial angiographic appearance of the patent ductus arteriosus.](image1)

![Figure 2: Second angiography with lateral aortogram showing dilated pulmonary end of patent ductus arteriosus.](image2)
is also protrusion of intimal cushions into the lumen [18, 19]. These lead to functional closure of the ductus that take place within 12–15 hours after birth in full term infant. Second stage within 2–3 weeks after birth there is infolding of endothelium and proliferation of sub intimal layers which leads to fibrosis and permanent sealing of the lumen to produce ligamentum arteriosum [20]. The size of the PDA (length and the size of the ampulla) can vary from few millimeters to several centimeters, it may enlarge with age [21]. The PDA can be classified according to their size of the internal ductal diameter and shapes on lateral angiographic views. PDA is silent if it is less than 1 mm, very small if less than or equal to 1.5 mm, small if it is 1.5 to 3 mm, moderate if it is 3–5 mm and large if it is more than 5 mm [22]. The angiographic appearance of isolated PDA can be classified into five types that has been described by Kirchino in 1989 [23]: Type A or conical type of PDA has a narrow end at the pulmonary insertion with a well-defined ampulla at the aortic end. Type B or window type of PDA is short and wide it may be narrow at aortic insertion. Type C or tubular type of PDA is tubular without any constriction. Type D or complex type of PDA has multiple constrictions (like our patient). Type E elongated type of PDA has a bizarre shape with an elongated conical shape and the constriction been remote from anterior border of trachea. Type A and B are further classified into three subgroups according to their site of insertion at the pulmonary end to the tracheal shadow [23]. Usually, closure of ductus begins at pulmonary end [5, 24], and may remain incomplete at aortic end leaving small diverticulum adjacent to aorta, which is regarded as a transient and physiological part of normal spontaneous closure of ductus arteriosus called ductus bump [25, 26]. Delayed closure of aortic orifice of the ductus exposing ductal tissue to systemic pressure is the most likely pathogenesis of aneurysm formation [5, 24].

In our patient aneurysmal change was noticed at the pulmonary end than aortic end that represent about 30% of neonatal cases of DAA [5], mostly in neonates with pulmonary hypertension [6], but in our patient (childhood type), because she had a mild pulmonary hypertension, we think the same pathophysiology can applied here too. However, this does not explain the fetal or early neonatal development of DDA, when pressure in pulmonary artery is equal or higher to that in the aorta. In fact, there are several other theories about pathogenesis of DDA. First, congenital wall weakness may result from necrosis and mucoid degeneration of the media in the ductal wall [5]. Second increased flow throws the ductus arteriosus in the uterus a concomitant exposure to an increased arterial pressure which occurs in the third trimester lead to aneurysmal formation [7, 27]. Third although there is no evidence of ductal stenosis on the prenatal echocardiogram [6, 7, 15, 28, 29], but intrauterine ductal constriction may cause post-stenotic dilatation of ductus arteriosus [30]. Finally, abnormal elastic fiber and extracellular matrix deposition within the wall of ductus arteriosus may occur in the setting of connective tissue disorder like Marfan, Ehlars–Danlos and Larsen syndromes has been observed in some cases of DAA which may result in weakening of the wall of the ductus arteriosus [2, 6]. None of these theories can explain the pathogenesis of DAA in all age groups. Even
some papers consider DAA as normal variant of elongated ductal bump [9].

Previously, DAA was considered as rare cardiovascular lesions, mostly was described in isolated case report [2]. The most common age at diagnoses is within the first two months after birth [6], but the pathology can also be diagnosed prenatally [6, 7, 15, 28]. Sign and symptom of ductus arteriosus aneurysm are usually scarce and discrete. Majority of affected neonate with DAA usually clinically asymptomatic [6, 9], and complication may occur but very rare in newborn with DAA, with more symptomatic patient having larger DAA diameter and more commonly associated with complications [9]. However even smaller DAA might be associated with fatal complications in patient with connective tissue disease [5]. There are report suggesting that congenital DAA that developed postnatally may be associated with a higher rate of complications especially those who are symptomatic [5, 8, 14, 31, 32]. They may present with wide spectrum of symptoms with or without complications like spontaneous rupture, thromboembolism, erosions and compression of the adjacent structures including airway and the recurrent laryngeal nerve and even death [5–8, 33–35].

Our patient was asymptomatic until age fourth year when she was examined by her pediatrician as a part of regular neonatal follow-up because of her prematurity which we think that the PDA was present initially and of silent type and have gone undetected, even though she was clinically asymptomatic but once the diagnosis was made the parents start to notice that their daughter having symptoms like easy fatigability and shortness of breath. The diagnostic modality for DAA after initial physical examination and chest X-ray is echocardiography which can provide a reliable and valuable method for both diagnosis and demonstration of DAA anatomy [9, 12], there are three important signs on echocardiography including unusual ductal shunt jet, triple star sign and rabbit ear sign [9], and other imaging modality like CT angiography and MRI scan have also been used for diagnosis and the nature of DAA but they are rarely necessary unless there are clinical evidence of compression of extravascular structure [9]. But in this patient, although there were some echocardiographic changes in the nature of the PDA unfortunately was not diagnosed through serial echocardiographic follow-up until after the second trial for PDA closure through catheterization then we found that the PDA have gone through aneurysmal dilatation at the pulmonary end, probably such aneurysmal change was unexpected over this 14 months, rapidly checked and missed. In adult patient, a limited acoustic window may impair detail evaluation of this region by echocardiography and color Doppler may be unable to detect a DAA when there is no left-to-right shunting [36].

When we found that PDA was aneurysmal during the second catheterization decision was made to close the PDA surgically rather than trans-catheter closure because our patient had no high risk for surgery, apparently the application of these devices may be associated with later complications of the aneurysm that mandate re-operative surgery, which carries a highest mortality rate 26%, and without intervention mortality reaches 91% due to rupture and infection [4]. Our patient had a clear indication for operation rather than watchful waiting despite that she was completely asymptomatic. From our review of literature we found that surgical resection for DAA should be proposed if any of the following condition existed: first, patent ductus arteriosus with DAA persist beyond the neonatal period (our patient); second, DAA associated with connective tissue diseases; third: there is evidence of thrombus extension into adjacent vessels or thromboembolism and finally, significant compression of adjacent structures [6, 9].

From our review of literature, we found that most of surgical resection of DAA with or without complications was carried out through median sternotomy with cardiopulmonary bypass machine [7], separate perfusion of the arch and the lower extremities [3], and circulatory arrest [33]. and method of closure were hand sewing aneurysmorrhaphy [37] that has been performed in our patient or stapling in case of uncomplicated ductus aneurysm having a narrow neck [9] or with the help of a Gott shunt bypassing the aneurysm and patching the neck of aneurysm from inside the aorta [8]. In our case, both aortic and pulmonary end aneurysmorrhaphy with pledged from pericardium done without resection of the PDA after putting the patient on cardiopulmonary bypass as a stand by for any undesired complication intraoperatively. Simple ligation of PDA in case of DAA is contraindicated because of the likelihood of incomplete discontinuation of the flow and risk of rupture both intraoperatively and long-term [6]. Giving the likelihood of long-term complication that might happen with DAA even after complete surgical resection even without association of connective tissue diseases, these children need continued follow-up.

CONCLUSION

We conclude that although PDA relatively is common and safe heart defect but they may complicate further with age, even restrictive PDA may complicate by aneurysmal PDA if left untreated and it should be couisously treated.

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Author Contributions
Aso F. Salih – 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
Rozhin O. Qadir – Analysis and interpretation of data, Drafting the article, Revising it critically for important
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REFERENCES

The combined use of surgical rehearsal platform and BrainLab navigation for mandibular osteotomy in Nager syndrome

Tian Ran Zhu, Justine C. Lee

ABSTRACT

Introduction: Nager syndrome is a rare genetic condition characterized by defects primarily of the face, arms, and hands. Children with Nager syndrome are frequently born with maxillary hypoplasia in conjunction with micrognathia and associated cleft lip/palate anomalies. These abnormalities severely restrict proper feeding, impair normal speech and language development, and contribute to life-threatening breathing problems. Surgical management options include mandibular osteotomy and distraction osteogenesis to correct the maxillofacial defects. Operative complications include damage to surrounding nerves and vessels, entry to the skull base, and recurrent temporomandibular joint (TMJ) ankylosis. Case Report: Nine-year-old girl with Nager syndrome presents for recurrent bilateral TMJ ankylosis. SuRgical Planner™ surgical rehearsal platform (SRP) was used in conjunction with Brainlab intraoperative computed tomography (CT) navigation system to decrease operative time, enhance visualization of key anatomical landmarks and extent of dissection, and minimize intraoperative risks and complications. Conclusion: SRP and Brainlab intraoperative CT navigation system first developed for neurosurgery have been successfully applied to craniofacial procedures. This case highlighted the synergistic benefit of SRP and Brainlab image guidance software to enhance a surgeon’s ability to increase operative efficiency, minimize surgical complications, and improve overall patient outcome.

Keywords: Brainlab, Craniofacial, Distraction osteogenesis, Mandibular osteotomy, Nager syndrome, Surgical planner

INTRODUCTION

Nager syndrome is a congenital disorder of the first and second branchial arches and appendicular system that result in underdeveloped face, arms, and legs [1]. Nager and Reynier first coined the term acrofacial dysostosis in 1948 to distinguish Nager syndrome as a craniofacial malformation from mandibular dysostosis [2]. While the exact cause is unknown, most cases are sporadic with case reports of autosomal dominant and recessive pattern of inheritance that is associated with deletion in SF3B4 gene in the long arm of chromosome.
The resulting malformations of the branchial arches manifest as mandibular hypoplasia, malocclusion, micrognathia, cleft palate, and microtia [1, 3, 4]. These abnormalities frequently cause feeding problems in infants with Nager syndrome secondary to mandibular hypoplasia and palatal defects. In addition, micrognathia with subsequent glossoptosis can lead to life-threatening apnea and asphyxiation, necessitating distraction osteogenesis concomitant with mandibular osteotomy to advance the jaw both anteriorly and inferiorly to alleviate the soft-tissue obstruction of the airway [5–8].

Similar patterns of craniofacial anomalies have been observed in other genetic syndromes affecting children including Treacher Collins syndrome, Pierre Robin syndrome, and Cleidocranial dysplasia. All of these syndromes share commonality in that specific genetic alterations affect key craniofacial developmental pathways leading to micrognathia, glossoptosis, and subsequent air obstruction [9, 10]. Therefore, the goals of treatment for these children focuses on breathing and feeding and optimizing growth and nutrition.

What differentiates Nager syndrome from these other aforementioned syndromes is involvement of distal limb buds that can result in deformed or absent thumbs, shortened or absent forearms, hammer toes, and leg and feet bone abnormalities [3, 5, 11].

Herein, we report a case of recurrent temporomandibular joint (TMJ) ankylosis in a child with Nager syndrome and demonstrate the efficacy of SuRgical Planner™ surgical rehearsal platform (SRP) in conjunction with Brainlab intraoperative computed tomography navigation system to augment intraoperative visualization, enhance surgical proficiency and safety, and improve overall patient outcome.

CASE REPORT

We present a case of a nine-year-old girl with Nager syndrome who was born with severe micrognathia, auricular atresia, high arched palate, bilateral radial deficiencies, and left club foot. At birth, she presented with significant respiratory distress secondary to mandibular hypoplasia and subsequent glossoptosis. Because of her difficult airway, intubation was attempted but failed, requiring emergent tracheostomy tube placement and mechanical ventilation. The patient was weaned off the ventilator prior to discharge. Since then she has not required mechanical ventilation. The relevant surgical history includes tracheostomy at birth, right index finger pollicization at age two, implantation bone anchored hearing aid at age five, and previous bilateral mandibular osteotomy at five years of age.

Prior to her last surgery, the patient had reankylosis of her bilateral TMJ resulting in severe limited jaw opening that required repeat mandibular osteotomies (Figure 1). For the previous mandibular osteotomy, Brainlab intraoperative CT-guided navigation system was used to aid in the preoperative planning of localizing and assessing the extent of the TMJ fusion. The surgery was completed in nine hours with no complications. For most recent mandibular osteotomy, SuRgical Planner™ was used in conjunction with Brainlab intraoperative CT-guided navigation system (Figure 2). This additional surgical guidance tool allowed us to rehearse the operation for TMJ resection and also assess the extent of our dissection, thereby accelerating operative efficiency (Figure 3). Additionally, based on principle of CT scanning Hounsfield unit, the SRP simulator can display or hide slices of tissue in real time, thereby allowing us to visualize surrounding skull base, vessel, and soft tissue anatomy as well as the location of our surgical probe to minimize skull base complications [12]. Overall, the operative time was four hours, a notable decrease from nine hours previously. There were similar scars noted from prior surgeries and no complications. The exact same sequence of surgery and placement of Matthews device was performed for these two operations to justify the comparison.
DISCUSSION

Severe airway obstruction in Nager syndrome remains the major cause of morbidity and mortality. Respiratory instability secondary to micrognathia and glossoptosis frequently necessitates a tracheostomy, while a gastrostomy tube may be needed for adequate nutrition [6, 13]. Later, mandibular osteotomy and distraction osteogenesis are necessary to address any mandibular and TMJ anomalies. Significant intraoperative risks surround the use of instrumentation to resect fused TMJ, in which the close vicinity to facial nerve branches, adjacent soft tissues, and the undersurface of skull base presents a notable challenge for craniofacial surgeons. Brainlab image guidance, a technology first developed for neurosurgery, has been applied to craniofacial surgery to address these concerns. Thus, by using Brainlab imaging in our operative planning we were able to design osteotomy lines, track the extent of bilateral TMJ and condylar resections, and identify the stylomastoid foramen to minimize damage to local tissues, nerves, and vessels and avoid entry into the skull base.

CONCLUSION

With the increased risk associated with surgical approaches to craniofacial reconstructions that closely border skull base and orbital floors, it is critical to advance training to minimize complications and improve surgical outcome. The trend to using 3D image guidance has advanced neurosurgery with improved operative time and patient safety. In addition, visual simulation technology further enhances resident training and provides an additional modality to guide operative decision making. We present this case to highlight the applicability of these innovations for mandibular osteotomy in Nager syndrome and other craniofacial operations. In particular, we demonstrate that the combined use of surgical rehearsal platform and Brainlab image guidance resulted in additional improvement in operative efficiency, enhanced real-time visualization of important anatomy, and minimized surgical complications.

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Author Contributions
Tian Ran Zhu – 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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Authors declare no conflict of interest.

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REFERENCES

Papillary carcinoma arising in thyroglossal duct cyst: A rare case presentation
Roopa Arora, Fatima Al-Hashimi

ABSTRACT

Introduction: Thyroglossal duct cyst (TGDC) is a common congenital anomaly but carcinoma arising in thyroglossal duct cyst is rare, occurring in 1% of cases. Case Report: We report a case of a 22-year-old female who was reported to have TGDC on radiology and fine needle aspiration cytology (FNAC). However, histopathological examination of TGDC post-Sistrunk procedure revealed an incidental presence of papillary carcinoma arising within a thyroglossal duct cyst which invaded its wall as well as involved a resection margin. Conclusion: The diagnosis of thyroglossal duct cyst carcinoma can be missed due to its rarity.

Keywords: Diagnostic procedures, Management, Papillary carcinoma, Thyroglossal duct cyst

INTRODUCTION

Thyroid gland develops as an endodermal diverticulum in floor of pharynx, at the level of foramen cecum and descends down as thyroglossal duct in front of neck, anterior to hyoid bone and laryngeal cartilages, between third and eighth weeks of gestation. Its distal end is enlarged to form thyroid gland. It obliterates completely by ninth week of gestation. Up to 60% cases of thyroglossal duct cyst (TGDC) show islands of functioning thyroid follicles in its wall [1]. However, parafollicular cells are absent. Incomplete involution of thyroglossal duct causes formation of thyroglossal cyst. Thyroglossal duct cysts are frequently seen in clinically practice in pediatric age group and 7% in adults. They present as slowly growing palpable anterior neck masses which move with swallowing [2]. Occurrence of thyroid carcinomas arising in a TGDC is rare [3]. Brentano in 1911 and Uchermann in 1915 were the first to describe a neoplasm in a remnant of thyroglossal duct [4]. Most common malignancy in TGDC is papillary carcinoma followed by others. However, medullary carcinoma is not seen owing to absence of parafollicular cells.

Sistrunk operation is the treatment of choice but other treatment modalities like partial/total thyroidectomy are considered depending on status of thyroid gland (any cold nodules), cervical lymph node enlargement and neck irradiation [5].

CASE REPORT

A 22-year-old female was referred to our hospital with a midline neck swelling for one year. There was no history of pain while swallowing. She did not give any history of shortness of breath, fever, weight loss or discharge from the lump. There was no change in bowel habits.
Clinically, the thyroid was not enlarged and no cervical lymph node enlargement was palpable. Thyroid function tests were within normal range and other baseline blood investigations were also normal.

Ultrasound of neck revealed a well-defined multiloculated midline cystic lesion measuring 4.5x2 cm, with thin wall which was embedded in muscle and located in infrahyoid region with extension to both paramedian areas. The thyroid gland appeared normal. Also, there was no cervical lymphadenopathy. Based on clinical and ultrasound findings, a diagnosis of thyroglossal duct cyst was made.

Fine needle aspiration cytology (FNAC) of the cyst revealed it to be a benign cystic lesion composed of lymphocytes, macrophages, few neutrophils and groups of bland follicular cells (Figure 1).

A Sistrunk procedure was performed for removal of cyst and the surgical specimen was sent for histopathology. Grossly, the collapsed cystic mass measured 3.5x2.2x2 cm. A separate piece of hyoid bone was also received.

Microscopic examination revealed a cyst lined by respiratory and squamous epithelium. Its lumen revealed a papillary carcinoma, composed of complex, branching randomly oriented papillae with fibrovascular cores and many psammoma bodies. The wall of cyst revealed islands of normal colloid-filled thyroid tissue as well as foci of papillary carcinoma (Figures 2 and 3). The papillae were lined by cuboidal cells with optically clear and overlapping nuclei, some showing intra-nuclear inclusions and nuclear grooves (Figure 4). The tumor measured 1.1 cm. Connective tissue surrounding the cyst showed many lymphoid aggregates and fibrosis. The tumor was very close to one resection margin. The hyoid bone did not show any tumor.

Based on histopathological report, a post-surgery thyroid scan was performed which showed a normal sized thyroid gland with homogeneous radiotracer uptake. No cold or hot nodules were seen.

Figure 1: Fine needle aspiration cytology showing clusters of benign follicular cells surrounded by histiocytes and inflammatory cells (May Grunwald Giemsa, x400).

Figure 2: Thyroglossal cyst lined by respiratory epithelium with papillary carcinoma inside its lumen. Wall shows normal thyroid follicles (upper left corner). Small focus of papillary carcinoma with psammoma bodies also seen (upper right corner), (H&E stain, x100).

Figure 3: Thyroglossal cyst with papillary carcinoma in its wall (H&E stain, x100).

Figure 4: Papillary carcinoma with characteristic nuclear features and psammoma bodies (H&E stain, x400).
Since the tumor invaded the wall of cyst and was close to one resection margin, a need for total thyroidectomy was discussed with the patient but the patient refused further surgical intervention.

DISCUSSION

Thyroglossal duct cyst (TGDC) is a developmental abnormality of the thyroid gland. Due to embryological remnants of thyroid tissue located in the TGDC, the same malignant tumors that develop in the thyroid gland can also develop in the TGDC [6]. Most common are papillary carcinoma (75–80%), but other thyroid tumors such as follicular, Hürthle cell, and mixed papillary–follicular carcinomas also have been reported [7]. Anaplastic and squamous cell carcinoma can also occur but are very rare [8, 9].

Thyroglossal duct carcinoma may be clinically indistinguishable from benign TGDC. It is an incidental finding on histopathology as was seen in our case. In such cases, diagnosis is usually made after histopathologic examination of a specimen obtained after a Sistrunk resection [6]. Ultrasound, computed tomography (CT), magnetic resonance imaging (MRI), and fine-needle aspiration biopsy (FNAB) can be useful techniques to differentiate TGDC carcinomas from simple TGDC before a surgical operation [5].

Fine needle aspiration cytology and intra-operative frozen section may help in a preoperative diagnosis of malignancy in TGDC. However, incidence of diagnosis of malignancy on histopathology after a benign diagnosis on FNAC is very rare [10, 11]. Inadequate yield of aspirated material due to cystic degeneration in the tumor could have led to this error. However, in presence of adequate cellular material, presence of tridimensional fragments, anisonucleosis, nuclear grooves, pseudo-inclusions and psammoma bodies are identified as the best indicators for a diagnostic resection of thyroid carcinoma.

Strict criteria by Widstrom et al. are followed for diagnosis of de novo TGDC carcinoma as opposed to metastasis from an occult primary in thyroid [12]. They are: histological identification of TGDC lined by columnar/squamous epithelium and normal thyroid tissue in its wall along with absence of carcinoma in thyroid gland [12, 13]. The same were adhered to in this case. A cystic lymph node metastasis is also differentiated from TGDC carcinoma based on histological demonstration of epithelial lining in the latter.

Similarly, cystic variant of papillary thyroid carcinoma would not have an epithelial lining of the cyst wall as opposed to TGDC carcinoma.

As regards management of such a case, an algorithm has been proposed for treatment of papillary carcinoma in TGDC which involves a simple Sistrunk procedure in patients less than 45 years of age with tumors less than 1.0 cm that are confined to the cyst and which show an ultrasonographically normal thyroid gland and no suspicious lymph nodes [5, 14]. There is a need of a total thyroidectomy followed by I\(^{131}\) ablation and thyroid-stimulating hormone suppression is considered [14]. If a tumor is more than 1 cm in size, multifocal or invades cyst wall. A similar treatment plan was recommended to the patient in the present case since the tumor extended outside cyst wall and was close to one resection margin but the patient refused another surgery.

CONCLUSION

The diagnosis of thyroglossal duct cyst carcinoma can be missed due to its rarity. For rapidly growing midline neck masses, relevant investigations involving imaging of the neck and fine needle aspiration cytology are required before surgical operation for an accurate diagnosis. The Sistrunk operation alone is sufficient for small tumors confined to the cyst, but total thyroidectomy with neck dissection is recommended for larger tumors which have invaded the wall of cyst, associated thyroid nodules and enlarged cervical lymph nodes.

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Author Contributions

Roopa Arora – 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

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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SUGGESTED READING

Recurrent posterior reversible encephalopathy syndrome in systemic lupus erythematosus

Melissa Ng, Sadia Saber, Richard Stratton

ABSTRACT

Posterior reversible encephalopathy syndrome (PRES) is an acute encephalopathy that manifests as headache, visual disturbance, altered mental state, and seizures. There are striking characteristic findings on neuroimaging. The PRES is associated with a number of conditions, including autoimmune disease. We describe the case of a 37-year-old female with a history of systemic lupus erythematosus presenting with headache and visual changes. Prompt diagnosis in PRES is important because if it is not recognized and treated early, it may progress to irreversible neurological damage. This patient made a good initial recovery, but suffered a relapse secondary to severe resistant lupus nephritis and refractory hypertension.

Keywords: Encephalopathy, Headache, Lupus, Posterior reversible encephalopathy syndrome (PRES), Systemic lupus erythematosus (SLE)

INTRODUCTION

Headache with visual changes is a common presentation with a broad differential and it is important not to miss serious causes. Neuroimaging is becoming more readily available, leading to quicker diagnosis of intracranial pathology. Posterior reversible encephalopathy syndrome (PRES) should be suspected in cases presenting with headache, visual disturbance, altered mental state, and seizures. Its associations include toxic agents, hypertension, sepsis, and autoimmune diseases. The PRES, as the name implies, is potentially reversible provided it is recognized and treated early through control of blood pressure and of the underlying cause [1]. However, if it is not adequately treated, it can progress to irreversible neurological damage, hemorrhage, and infarction [2]. Early diagnosis may also be important because treatment of other causes can differ, for example, hypertension as a cause of PRES is controlled as a mainstay of treatment, whereas hypertension in the context of ischemic strokes is more cautiously treated [2].

CASE REPORT

A 37-year-old female presented to the emergency department with a six-day history of worsening headache, severe photophobia, generalized body aches and weakness, and episodes of pyrexia, rigors, and vomiting. She had a background of systemic lupus erythematosus (SLE) complicated by lupus nephritis, pancytopenia, and antiphospholipid syndrome, migraine, and previous complex partial seizures. Previously, she had a left preretinal hemorrhage while on low molecular weight
heparin therapy. She was taking prednisolone 10 mg per day and mycophenolate mofetil 1 g twice a day, and was having monthly plasmapheresis. She was a non-smoker and non-drinker. There was no relevant family history.

She had a heart rate 115 beats/min, respiratory rate 19 mmHg, temperature 37.5°C, blood pressure 124/88 breaths/min, and oxygen saturations 99% on room air. On examination, heart sounds were normal, chest was clear, and abdomen was soft and non-tender. She had peripheral pitting edema. Neurologically, she was very photophobic and had loss of vision in both eyes. She was unable to finger count. No ocular pathology was found by ophthalmology.

Computed tomography scan of head with contrast showed extensive bilateral low attenuation changes along the white matter tracts, particularly in the parietal and occipital lobes. There was no evidence of hemorrhage and no filling defects identified along the venous sinuses. Magnetic resonance imaging (MRI) scan showed bilateral T2/FLAIR hyperintense change involving the parieto-occipital lobes – features most consistent with PRES. There was also mild generalized cerebral volume loss.

The patient’s blood pressure increased during her admission to 170 systolic. She was started on nifedipine and perindopril to control her blood pressure and limit progression of PRES. The dose of prednisolone was increased to 40 mg daily. She was discharged after five days with rheumatology follow-up.

The patient was re-admitted several weeks later with a progression of lupus nephritis and refractory hypertension (blood pressure 190/110 mmHg). During her admission, she experienced seizures. Repeat neuro-imaging showed that the previously abnormal areas had almost completely resolved. There were new areas of signal change in the frontal, parietal, and occipital lobes, as well as the cerebellar hemispheres, midbrain and pons. She then had another episode of severe headache and visual loss. Imaging once again showed significant resolution but revealed new lesions in the occipital and frontal lobes, right corpus callosum, and left caudate. The development and resolution of lesions are demonstrated in Figures 1–3. The blood results at the time of each flare are given in Table 1. Her condition improved after treatment with intravenous labetalol and nitrate, steroids, plasma exchange, intravenous immunoglobulin, and rituximab. She has now made a good recovery and remains clinically stable.

**DISCUSSION**

Posterior reversible encephalopathy syndrome is an under-recognized clinical and radiological syndrome which usually presents with headache, visual changes including cortical blindness, nausea and vomiting, altered mental state, and seizure activity [3]. Acute hypertension is strongly associated with PRES, though it does not correlate with severity [2]. Posterior reversible encephalopathy syndrome has been associated with toxic agents such as immunosuppressive therapies, sepsis, pre-eclampsia and eclampsia, and autoimmune conditions such as systemic lupus erythematosus, systemic sclerosis, and polyarteritis nodosa and other vasculitides.

Magnetic resonance imaging scan remains the gold standard of diagnosis in PRES. Previously, the radiological findings were classically reported as cerebral edema along the white matter tracts in the posterior parietal and occipital lobes [4], but other patterns on neuroimaging are increasingly recognized [2]. The pathophysiology is not well understood but the current theory postulates that impaired cerebral autoregulation results in vasogenic edema secondary to capillary leakage and endothelial disruption [4].

![Figure 1: Magnetic resonance imaging scan of head, axial section, T2-weighted images, (A) (On admission): hyperintense changes in parieto-occipital lobes, (B) (Five weeks): resolution of changes. New patchy areas of high signal change in frontal and parietal regions, and (C) (Seven weeks): significant resolution with new area at right posterior temporal-occipital junction.](image)

![Figure 2: Magnetic resonance imaging scan head, axial section, T2-weighted images, (A) (On admission): bilateral hyperintense changes in parieto-occipital lobes, (B) (Five weeks): resolution of changes, (C) (Seven weeks): new lesions in occipital lobes, posterior temporal-occipital junction, splenium of corpus callosum.](image)

![Figure 3: Magnetic resonance imaging scan head, axial section, T2-weighted images, (A) (On admission): no significant areas of hyperintense change, (B) (Five weeks): extensive signal change of pons, right middle cerebellar peduncle, cerebellar hemispheres, and inferior vermis, and (C) (Seven weeks): resolution of previously abnormal areas.](image)
A number of cases of PRES have been described in the context of SLE [1, 5]. Given the multisystem nature of SLE, it is likely that there are several contributing factors to the development of PRES, such as hypertension, renal disease, and use of immunosuppressive agents. Furthermore, given the wide range of neuropsychiatric manifestations in SLE, the diagnosis of PRES can be difficult if neuroimaging is not readily available. The fact that our patient was not hypertensive at the time of presentation clouded the picture given the strong association between hypertension and PRES.

The mainstay of treatment of PRES is early blood pressure control and removal of the causative agent to prevent progression to permanent neurological damage. Prompt treatment has a good prognosis, and patients often make full neurological recovery, but the risk of neurological impairment and up to 15% risk of mortality need to be noted [2]. Recurrent PRES is uncommon. This patient had acute flare-ups of severe resistant lupus nephritis and refractory hypertension which contributed to recurrent PRES.

### CONCLUSION

In conclusion, posterior reversible encephalopathy syndrome presents as headache, visual changes, altered mental state, and seizures. It is associated with toxins, hypertension, sepsis, and autoimmune conditions. Typical magnetic resonance imaging scan show cerebral edema in the white matter tracts with predominance towards the posterior parietal and occipital lobes. This case brings to light a few key take-home lessons: firstly, early diagnosis of posterior reversible encephalopathy syndrome (PRES) is crucial so blood pressure and any underlying cause can be aggressively treated. Secondly, flares of associated conditions can lead to PRES recurrence. Finally, it is important to bear in mind that PRES if often, but not always, reversible and can result in permanent neurological damage.

### REFERENCES


An unconventional ameloblastic fibro-odontoma with compound odontoma like features

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ABSTRACT

Introduction: Odontogenic tumors cover a wide and diverse spectrum of tumors. Some odontogenic tumors have a varied etiopathogenesis and share a presentation very similar to hybrid odontogenic tumors. Ameloblastic fibro-odontoma is one such tumor. Though the WHO 2017 Classification of Head and Neck tumors continues to support this hypothesis, it is clear, with more than 215 case reports, that it is a tumor that does not feature in the continuum of the spectrum of ameloblastic fibroma ending with ameloblastic fibro-odontoma and odontoma. It, therefore, deems recognition as an independent entity. The ameloblastic fibro-odontoma irrespective of central or peripheral location will show an ameloblastic fibroma like presentation along with a compound or complex composite odontome histologically. It has also been conjectured whether it is a true neoplasm or hamartoma. Case Report: We report a case of an ameloblastic fibro-odontoma in an eight-year-old boy with a diffuse swelling extraorally in the infraorbitalzygomatic area. Intraoral examination revealed a large 4.5x3 cm soft tissue mass on the right maxillary alveolar area involving primary molars and permanent 1st and 2nd molar region. Radiologic examination revealed a lesion with 32 denticles and destruction of alveolar bone, which showed a histopathologic picture of ameloblastic fibro-odontoma with a composite compound odontoma like pattern. Conclusion: Our case describes an ameloblastic fibro-odontoma, which presented as a painful mass of the maxilla with compound composite odontome like features supporting the tumor concept and as an independent entity not belonging to the continuum spectrum.

Keywords: Ameloblastic fibro-odontoma, Central, Composite compound odontoma, Dentin, Enamel, Induction, Maxillary posterior quadrant

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INTRODUCTION

Odontogenic tumors cover a wide spectrum and comprise an array of tumors. These have been classified into epithelial, mesenchymal, and mixed tumors by the 2017 WHO classification of odontogenic tumors [1]. Ameloblastic fibro-odontoma (AFO) previously was listed under the category of mixed odontogenic tumors formerly described under names like immature ameloblastic odontoma (Slootweg) and ameloblastic odontoma (Hooker). This group of tumors shows the evidence of odontogenic induction along with mesenchymal tissue proliferation. The present tumor morphologically has resemblance of ameloblastic fibroma along with odontome like areas. It strides ahead of ameloblastic fibro dentinoma by showing the presence of induction in formation of enamel or enamel matrix [2].

Ameloblastic fibro-odontoma is a rare tumor with approximately 215 well documented reports (211 – central, 3 peripheral lesions) [3]. Irrespective of its location, the tumor presentation is similar histologically. It is a biologically non-aggressive tumor, responding well to conservative treatment. It is unfortunate that this distinct lesion has not been recognized by the recent classification of odontogenic tumors and has been grouped under the umbrella term developing odontomas or as a variant of ameloblastic fibroma [1, 4].

We present a case of ameloblastic fibro-odontoma with a very classic histopathologic presentation supporting the tumor concept and as an independent entity not belonging to the continuum spectrum.

CASE REPORT

An eight-year-old boy reported to the clinical department with a complaint of painful mass in the upper jaw since a month. The child presented with a diffuse swelling of the right cheek, extraorally, measuring approximately 5x4.5 cm and extending anterior-posteriorly from the nasolabial fold to the pre-auricular area and superior-inferiorly from the infraorbital region to 2 cm below the ala-tragal line. Intraoral examination revealed a large 4.5x3 cm soft tissue mass on the right maxillary alveolar area involving primary molars and permanent 1st and 2nd molar region. Mucosa covering the lesion appeared erythematous on the superior surface and the mass was obstructing the occlusion (Figure 1A–B).

On palpation, a soft tissue mass with well-defined borders was noted, with expansion of both buccal and palatal plates. A provisional diagnosis of odontome was given. On radiologic examination, the orthopantomogram revealed a mixed dentition state of the child. The upper right maxillary quadrant showed an ill-defined radiolucent lesion with multiple small pebble size radio-opaque masses. Approximately, 32 such denticle like structures were noted in the radiograph. Destruction of the alveolar bone in the right maxillary molar region and retromolar area was observed. Developing tooth follicles of permanent molar 16 and 17 were pushed superiorly into the infraorbital area. The incompletely formed teeth 14 and 15 were mesially displaced (Figure 1C).

The lesion was provisionally diagnosed as compound odontome based on radiographic features and a conservative excision was planned. The differential diagnoses included complex composite odontome and odontoameloblastoma.

Surgical note described a well-delineated mass, soft in nature, which shelled out of the bone. It was irregular in shape with multiple hard tooth crown like structures within the tumor mass.

On grossing, a 4.5x4 cm tumor mass, irregular in shape was seen. Cut section revealed off white, pale myxoid area and chalk white area intervened with numerous small tooth like structures. Some of the tooth like structures separated and shelled out from the tumor mass. A radiograph of the gross specimen reconfirmed multiple such tooth like structures, approximately 32 in number (Figure 2).

On histopathologic examination with hematoxylin and eosin staining, the lesional tissue showed a predominance of immature cellular ectomesenchyme – made up of a myxoid stroma with plump spindle cells, which are monotonous. The odontogenic epithelium is seen in the form of strands and small nests. The strands were similar to the dental lamina (Figure 3).

At multiple areas the dental lamina like strands gave rise to dental follicles very similar to advanced bell stage, showing the presence of ameloblasts, odontoblasts along with enamel matrix and dentin formation. Enamel was partially mineralized or was seen as an un-mineralized matrix. Mineralized enamel showed the presence of well-formed enamel prisms (Figure 4). Dental follicles showed the presence of thick tubular predentin like areas lined by odontoblasts. The area of dentin next to the enamel is well mineralized with globular dentin. Different areas showed dental follicles in various stages of tooth development. The enamel organs bore resemblance to cap, bell and advanced bell stages very frequently. No evidence of cementum like tissue was seen, but primitive pulp in the form of dental papilla was observed (Figure 5). These areas appeared similar to a composite compound odontome. Based on these findings, a final diagnosis of ameloblastic fibro-odontoma was given.

DISCUSSION

Ameloblastic fibro-odontoma is defined as a lesion similar to ameloblastic fibroma but also shows inductive changes that lead to the formation of enamel and dentin [5]. It is a rare tumor accounting for 1–3% of all odontogenic tumors [3].

Ameloblastic fibro-odontoma bears resemblance to ameloblastic fibroma, ameloblastic fibrodentinoma...
and odontomes because of which investigators have hypothesized that ameloblastic fibro-odontoma could be one entity in different stages of development, in which: ameloblastic fibroma evolves to ameloblastic fibrodentinoma (AFD); ameloblastic fibrodentinoma evolves to ameloblastic fibro-odontoma and ameloblastic fibro-odontoma matures to an odontoma (Figure 6).

Figure 1: (A) Extra-oral view of patient reveals an enlarged diffuse facial asymmetry of the right side, (B) Intra-orally a 4x5 cm lobulated mass is seen on the right maxillary alveolar ridge posterior to 55, involving the tuberosity with interspersed erythematous areas. 16 is clinically missing, and (C) Orthopantomogram reveals a mixed radiopaque radiolucent lesional mass in the right posterior maxillary posterior region involving tuberosity with numerous globule like radiopaque masses along with impacted 16.

Figure 2: (A, B) Irregular soft tissue mass with numerous tooth crown like structures obtained on excisional biopsy, (C) Radiograph of the gross specimen showing numerous radiopaque densities within the soft tissue lesion, and (D) Cut section of 4.5x4 cm of soft tissue specimen showing off-white myxoid areas along with whitish-brown areas with the presence of numerous hard tissues resembling denticles.

Figure 3: (A) Photomicrograph showing proliferation of dental lamina in the form of cords and strands in an ectomesenchymal background along with engorged capillaries (H&E stain x100). Inset: Higher magnification of the odontogenic epithelial strands (H&E stain, x200), (B) Dental follicles resembling bell stage surrounded by primitive ectomesenchymal tissue (H&E stain, x40), (C) Dental follicle showing predominantly hard tissue formation with strands and cords of dental lamina in the stroma, and (D) Dental follicle in advanced bell stage with formation of enamel, dentin and pre-dentin (H&E stain, x100).

Figure 4: (A) Histologic section showing presence of enamel organ comprised mainly of enamel and dental papilla on a primitive ectomesenchymal background with strands and cords of odontogenic epithelium of about 2–3 cells in thickness. (H&E stain, x100) (B) Higher magnification showing arrangement of enamel rods. (H&E stain, x400) (C) Higher magnification of enamel showing ‘fish scale’ pattern of arrangement of the enamel rods. (H&E stain, x400).
Another hypothesis proposed is on the two lines of development of ameloblastic fibro-odontoma. A neoplastic line comprised ameloblastic fibroma and ameloblastic fibrodentinoma and a hamartomatous line comprising of ameloblastic fibro-odontoma as a stage of odontoma [3, 5] (Figure 7). Some authors suggest that AFO should be considered as an immature complex odontoma [7]. Trodahl suggested that AFO exists in a stage in between the two schools of thought [8]. However, ameloblastic fibro-odontoma cannot be unanimously considered as a hamartoma, as it can show neoplastic biological behavior, cause bone destruction, deformity and instances of malignant transformation and variants have also been noted [9–11].

The present case definitely shows neoplastic biological behavior as it presented as a large tumor mass with bone destruction and cortical plate expansion. It also emerges as an independent entity and not as a part of continuum spectrum based on the clinical data.

Ameloblastic fibro-odontoma arises in patients between 8–12 years of age, [4] with a mean age of 9 years [5]. A slight male predilection is seen with more than half of the cases arising in the posterior mandible region intraosseously. It presents as a well-delineated slow growing painless mass; as a central lesion, generally associated with unerupted teeth, which is a common hallmark presentation of ameloblastic fibro-odontoma. On radiography, it presents as a unilocular or multilocular radiolucency with radio-opacities of different shapes and density. Displaced unerupted teeth show the presence of the tumor coronally [6, 12, 13].

The present case sits in the classical age group with eight years at the time of presentation in a boy, which was a typical feature. But the tumor presented in the maxillary posterior quadrant as a large tumor with cortical plate expansion and displaced unerupted teeth in a unilocular radiolucency. Multiple, as many as 32–40, tooth like structures were also seen on the radiograph. Approximately 80% of the lesions are associated with a tooth and 87% show radio-opacities as seen in our case, which showed the displacement of incompletely formed 14, 15, 16 and 17 [3, 5, 6].

Histopathological findings speak of immature complex odontome with enamel, dentin, cementum and pulp-like ectomesenchyme with odontogenic epithelium. Dentin can be in the form of tubular dentin or dentinoid [2].

Some authors describe it as a biphasic tumor with odontogenic epithelium proliferating in a highly cellular ectomesenchyme with primitive apparatus. It also contains tooth like structures – enamel and dentin with
varying degrees of maturation throughout the tumor [12, 13].

This case is in unison with the reports of De Lopes et al, De Riu, Nelson BL [3, 12, 13] as it displayed ameloblastic fibroma like areas with multiple areas of developing tooth like follicles with expression of enamel and dentin. No evidence of cementum was seen. Hence, it shows an ameloblastic fibroma like picture with composite compound odontome like areas in a very primitive cellular ectomesenchyme which is contrary to reports of Reichart and Philipsen which described presence of a complex odontome like tissues with ameloblastic fibroma like areas [5, 14].

A differential diagnosis of ameloblastic fibroma, ameloblastic fibrodentinoma and odontome is very important to consider. Ameloblastic fibro-odontoma and ameloblastic fibrodentinoma have a stand out difference of enamel induction in ameloblastic fibro-odontoma, which is not seen in ameloblastic fibrodentinoma. Also, studies have shown that visible radio-opacities have a higher tendency to occur in AFOs [3, 15].

Ameloblastic fibroma can be differentiated from ameloblastic fibrodentinoma and ameloblastic fibro-odontoma as induction is not seen in ameloblastic fibromas. Sometimes it is difficult to differentiate ameloblastic fibro-odontoma from odontomas; but generally odontomes show presence of enamel, dentin, cementum and pulp like areas in a complex composite or compound pattern whereas ameloblastic fibro-odontoma does not always induct cementum as seen in this case [15]. The above considered differential diagnosis can also be ruled out with the help of clinical data of age, site, gender distribution and roentgenographic details. Hence, taking all the above features into consideration, the case was confirmed as an ameloblastic fibro-odontoma.

A conservative therapy is adequate for treatment of ameloblastic fibro-odontoma [5, 6, 16]. The lesion in the present case was surgically excised. A periodic recall for two years did not show any evidence of recurrence.

CONCLUSION

In conclusion, this a rare case of ameloblastic fibro-odontoma present in the maxillary posterior quadrant with ameloblastic fibroma pattern along with compound composite odontoma like features, treated adequately by conservative surgery. It is most definitely an independent lesion and cannot be clubbed with odontome or ameloblastic fibroma. It should be given its due and its position as an individual entity by the World Health Organization (WHO).

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Author Contributions
Radhika Manoj Bavle – 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
The corresponding author is the guarantor of submission.

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Authors declare no conflict of interest.

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SUGGESTED READING

A rare case of a plastic foreign body (suction tip) within the soft tissues in the lingual aspect of the mandible

Eirini Boutiou, Ioannis A. Ziogas, Georgios Koloutsos, Margarita Vafiadou, Konstantinos Antoniades

ABSTRACT

Introduction: A rare occasion for the oral and maxillofacial surgeon as well as the dentist is to find a foreign object in a patient's oral cavity. The underlying etiology may be a trauma, therapeutic interventions (dentoalveolar or implant surgery), or even a tooth dislocation. It may cause symptoms and thus be a reason for the patient to visit the dentist or the oral and maxillofacial surgeon or it can be an incidental finding on radiographic evaluation. Case Report: A 54-year-old edentulous patient presented with pain upon wearing his lower denture. An orthopantomographic evaluation showed nothing unusual and the patient was scheduled for the removal of a clinically diagnosed lingual exostosis under local anesthesia. Intraoperatively, we found and removed a suction tip, which caused the discomfort. Apparently, it was an iatrogenic leftover from a previous dental therapeutic procedure (an inferior 3rd molar extraction), right after which it was noticed.

Conclusion: The dentist in general should be very careful, when treating his patients, so as not to leave a foreign object in the oral cavity.

Keywords: Adverse event, Foreign bodies, Maxillofacial surgery

INTRODUCTION

Coming across a foreign body in the oral cavity is an uncommon event. Foreign bodies can be ingested, aspirated, inserted by certain patient's habits or even accidentally placed by traumatic or iatrogenic injury. They can be fractured burr tips, dislocated teeth, dental implants or restorative material. Most of the times, the foreign bodies are made of plastic, metal, glass and they can be identified on X-ray, computed-tomography (CT) scan and magnetic resonance imaging (MRI) scan [1]. When it comes to the occurrence of an adverse event in relation with dental practice, oral surgery comes third [2]. At the same time cases with residual foreign bodies related to oral and maxillofacial surgery are extremely rare [3]. The majority of them can induce tissue reactions, such an abscess formation, or even a
septicemic or a hemorrhagic event. They may also cause pain, discomfort, functional disorders and infection, but they do not usually threat patient’s life [1, 4]. Retrieval of foreign objects can be a challenging and difficult aspect of the therapeutic approach, mostly due to the restricted access of the oral cavity and the close anatomic relations of the foreign body with vital structures, while normal anatomy may also come up with variations [1, 5]. The aim of this case report is to present a case of a patient with an uncommon foreign object in the oral cavity that was identified during surgery.

CASE REPORT

A 54-year-old edentulous man was referred to our outpatient department, complaining of a pain in the deep lingual aspect of the mandible on his left side, when he was wearing his lower denture. His past medical history was free and his surgical history only comprised of an appendicectomy. A bulge that was thought to be arising lingually of the left inferior 3rd molar post-extraction socket, right under the internal oblique line, was palpated upon clinical examination and the patient was scheduled for surgical removal of the diagnosed exostosis in order to relieve his relentless pain. Before proceeding in the operation, the patient was radiographically assessed, but nothing unusual was noticed (Figure 1). In the operating room, we performed left inferior alveolar nerve block along with several lingual and parietal local infusions. Then we went on with a longitudinal incision on the edentulous alveolar ridge of the left mandible and we raised the mucosalveolar flap. This revealed an absolutely normal bone surface and at the same time an abnormal mass within the corresponding soft tissues. Initially, it was thought to be a residual root of the 3rd molar (Figure 2), which was extracted several years before. This was a two step procedure for an initial unsuccessful attempt from a dentist was followed by an oral surgeon’s intervention. Then as we tried to pull out the foreign object, we surprisingly discovered that it was a suction tip (Figures 3). After removing it, the wound was closed with continuous locked suture (Figure 4). The patient was prescribed antibiotics and analgesics and was discharged.

DISCUSSION

In this case, an adverse event occurred by the previous two step dentist and oral surgeon’s intervention, during the extraction of the lower 3rd molar. Unfortunately, we cannot be aware of what exactly happened during this procedure. Thus, a meticulous history of the patient, which can provide more accurate information about the etiology, as well as radiographs are the cornerstone of delivering high quality medical therapy. Radiographs, in particular, can be found very useful, especially in case of a radiopaque foreign body [5]. Regarding radiolucent objects, such as those made of plastic or wood, CT scan may be the most reliable method. CT scan can also provide information about their relationship with the surrounding anatomical structures [6]. We suggest taking CT preoperatively in case we are aware of a
foreign body existence. The fact is that the patient was totally unaware of this and the panoramic X-ray was not indicative of such a case. He was referred to us in order to remove surgically a lingual exostosis. So, no CT scan was prescribed. Clinical presentation can also vary according to the type of the foreign object; steel and glass may not cause severe inflammation, but organic materials can lead to secondary infection with formation of an abscess, or a fistula [3, 7].

Gui et al. [4] reported an increase in the incidence of identifying foreign bodies in the deep maxillofacial area during the last years. Also, it is suggested that all foreign bodies presenting with clinical manifestations or found near vital structures should be removed in order to prevent further complications. A problem that an oral and maxillofacial surgeon may face during the removal procedure is the tendency of the foreign objects to move within the soft tissues, thus implying difficulties due to its mobility. Sometimes fibrous connective tissue can develop around them, just as in this case, which may prevent further movements, hence providing an obvious advantage to the surgeon.

As far as removal of the foreign bodies is concerned, a computer-based image guided navigation system can be beneficial for the surgeon. After acquiring intraoperative three dimensional imaging data of the anatomy, injury to anatomical areas can be prevented and preoperative planning can be improved. Additionally, operating time may be decreased with the simultaneous precision and accuracy of a minimally invasive procedure. However, its implementation depends on the location of the foreign object and as such it cannot be universal [1].

Generally, in order to achieve high-level patient safety, the dentist should be careful enough to prevent adverse events from happening.

**CONCLUSION**

The dentists and the surgeons in general should be very careful, when treating their patients, so as not to leave a foreign object in the oral cavity. However, if an adverse event occurs, the patient should be informed accordingly.

*********

**Author Contributions**

Eirini Boutiou – Substantial contributions to conception and design, Acquisition of data, 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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Cerebral hemorrhagic infarction as the initial manifestation of deep venous thrombosis in a child with patent foramen ovale: A case report

Dimitrios Panagopoulos, Sofia Loukopoulou, Georgios Markogiannakis, Nikos Eleftherakis

ABSTRACT

Introduction: Arterial ischemic stroke (AIS), with an estimated incidence of 1.1–4.3 per 100,000, is an important cause of morbidity and mortality in children and the recurrence risk is high. Case Report: We present the case of an 11-year-old child who presented with a symptomatology of acute ischemic stroke of unknown etiology. The radiological investigation did not reveal any underlying brain abnormality that could cause the event. The diagnostic work up included an echocardiogram, which revealed a thrombus in the right atrium, in conjunction with a patent foramen ovale. The patient was initiated immediately on anticoagulation therapy with low molecular weight heparin and warfarin, but two days later she suffered pulmonary emboli, diagnosed with spiral thorax computed tomography scan. An ultrasound study of the vessels of the lower extremities revealed deep venous thrombosis, which was considered to be the underlying causative mechanism. Conclusion: To the best of our knowledge, this is the first documented case of right atrial thrombus resulting from deep venous thrombosis in a pediatric patient with patent foramen ovale and associated ischemic stroke event. A discussion regarding the definition of cryptogenic stroke, its etiology and relationship with deep venous thrombosis and the currently proposed therapy, follows.

Keywords: Cryptogenic stroke, Foramen ovale

INTRODUCTION

Ischemic stroke in children is a relatively rare entity relative to the adult population. The definition includes ischemic and hemorrhagic infarction in children, 55% are believed to be ischemic, and the remainder hemorrhagic. The wide range of pathophysiological processes associated with pediatric strokes mandates a careful diagnostic evaluation to maximize the chances for optimal patient outcome.
Pediatric arterial ischemic stroke (AIS) is an important cause of neurologic morbidity in children. Consequences can include sensorimotor deficits, language impairment, and intellectual disability, behavioral problems, and epilepsy [1]. Unfortunately, the diagnosis of stroke in children is often delayed [2, 3].

Children with cardiac disease represent one of the most significant subsets of pediatric AIS patients. Across most series, cardiac risk factors are present in 2–31% of children with AIS [4–9].

A point of uncertainty persists regarding the role of an isolated patent foramen ovale (PFO), in part because there is significant variability in how this has been considered across studies, with some lumping it in with other structural heart diseases while others have separated it as a distinct diagnosis. While there is some evidence suggesting an important role of right-to-left shunting across an atrial defect, particularly among patients with prothrombotic conditions or cryptogenic stroke [10], the role of device closure remains undefined due to lack of sufficient evidence [11].

We present a rare clinical case of a young girl harboring a latent deep venous thrombosis (DVT), a thrombus in the right atrium with subsequent arterial ischemic stroke (AIS) of the brain, possibly due to a patent foramen ovale (PFO). This was the first clinical manifestation of an otherwise unrecognized clinical condition.

CASE REPORT

An 11-year-old girl presented with headache, vomiting, dizziness, dysphasia and gaze dedication for a few seconds. The initial computed tomography (CT) scan revealed intracerebral hematoma (ICH) in the right parieto-occipital region with perilesional edema (Figure 1).

Patient was admitted in the neurosurgical clinic and was initiated on anticonvulsant medication. Neurological and ophthalmological examination did not reveal any focal deficits. Magnetic resonance imaging performed the same day revealed edematous configuration of the nearby gyri with concurrent presentation of hemorrhagic elements (Figure 2).

An electroencephalography (EEG) study detected focal cerebral disturbances. A repeat MRI scan (with contrast) and magnetic resonance angiography (MRA), indicated a hemorrhagic infarct in the territory in a subacute phase with related edema (hemorrhagic stroke). Magnetic resonance angiography further recognized stenosis of the right middle cerebral artery (MCA) with obstruction of the posterior peripheral branches (Figure 3).

Consequently, a digital subtraction angiography (DSA) from the femoral artery was performed which did not reveal underlying vascular abnormalities (Figure 4).

A thorough investigation for hypercoagulable states (deficiencies of protein C and anti III, protein S, antithrombin and plasminogen, molecular studies for factor V Leiden, prothrombin 20210A, homocysteine, MTHFR gene mutations), and immunological – rheumatological conditions (HLA-51, c-ANCA, p-ANCA, anti – GBM, LA1 and LA2, β2GPI, ACA IgM and IgG antibodies) did not reveal any abnormalities.

An ultrasound study of the vessels of the lower extremities revealed an intraluminal thrombus of the left superficial femoral and popliteal vein. A thoroughly detailed investigation of patient’s history revealed a minor sports related blunt injury of the left lower extremity a day before the initial symptoms, which was associated with lower extremities ultrasound findings. Additionally, an echocardiogram visualized a thrombus (2x1.5 cm) attached to the right atrium in conjunction with patent foramen ovale (Figure 5).

Figure 1: The initial computed tomography scan, after the ictus. Arrow depicts the area of ischemic stroke (hypodense), with a surrounding area of hemorrhagic transformation.

Figure 2: Initial magnetic resonance imaging scan, within the first day after the event. Arrows depict the pathologic changes at T1W, T2W, FLAIR and T2 GRE sequences (images corresponding from left to right).
Due to the relative contraindication for thrombolysis, patient was initiated immediately on anticoagulation therapy with low molecular weight heparin and warfarin. Two days later, she developed acute symptoms of dyspnea and chest pain and a subsequent spiral thorax CT scan revealed pulmonary emboli at the left pulmonary artery, as long as the persistence of the atrial thrombus. Anticoagulation therapy was continued and a foramen ovale umbrella placement was later performed (Figure 6).

Patient remained symptom free in the follow-up period and serial cardiac ultrasound examinations revealed gradual resolution of the right atrial thrombus (Figure 7).

A few months later, a repeat MRI scan was performed, while the patient being neurologically normal. The examination verified the known lesion at the right temporal-occipital lobe region, which revealed characteristics, compatible with a chronic lesion. More specifically, the imaging of the lesion identified a territory with intermediate to hypo-intensity signal at FLAIR sequences and hemosiderin ring at T2-GRE sequences (Figure 8).

Figure 3: Magnetic resonance imaging and magnetic resonance angiography scan after 10 days of the event (from above to below, T1W, T2W, FLAIR, MRA reconstruction and T2 GRE images). Arrows depict the evolution of signal changes.

Figure 4: Digital subtraction angiography imaging, revealing no pathological findings.

Figure 5 (A, B): Arrows indicate the location of the thrombus with its approximate dimensions.

Figure 6: The arrow indicates the site of placement of the umbrella.

Figure 7: Arrow indicates the position of the umbrella.
DISCUSSION

Epidemiology: Definition of cryptogenic stroke

Arterial ischemic stroke (AIS), with an estimated incidence of 1.1–4.3 per 100,000, is an important cause of morbidity and mortality in children and the recurrence risk is high [12]. It is defined as an acute clinical syndrome with a neurological deficit referable to a cerebral arterial territory and a brain MRI scan showing a corresponding area of acute infarct.

A stroke is termed cryptogenic when its etiology cannot be attributed to any specific cause after an extensive search for the most common causes, such as atherosclerosis of the intracranial vessels, lacunar damage from hypertension, or embolus derived from a thrombus located in the left atrium, the left ventricular apex, or at the level of an ulcerated plaque of the aortic arch.

Etiology

The etiology of AIS remains undetermined in a high proportion of children. Predisposing conditions for ischemic cerebrovascular accidents in children include congenital heart malformations (congenital cyanotic complex heart malformations or acquired heart disease), sickle cell disease, infections, and collagen tissue abnormalities [13], but around half occur in children who were previously well (cryptogenic stroke) [14]. It is well known from literature, that one of the most common conditions associated with AIS is congenital heart malformations (like patent foramen ovale) [15–25].

Paradoxical embolism and stroke

Recently, paradoxical embolism across the PFO was suggested as a possible etiology in some of these children [26]. Patent foramen ovale is reported, as an autopsy finding, to remain patent in about 25% of adults, thus presenting a potential passageway for paradoxical embolization. On some particular circumstances such as during Valsalva maneuver, which is reproduced by the act of defecating or coughing, the reversal of the physiologic inter-atrial pressure gradient results in right-to-left shunting across the PFO and contributes to the passage of embolic material. The prevalence of PFO was significantly higher in patients with cryptogenic stroke versus those with known causes of stroke (42% vs 7%), indicating that PFO is associated with cryptogenic stroke [27, 28]. The association is documented in case reports [13, 14, 29–33]. However, the direct role of a PFO in stroke remains unclear [34].

In our case, the dual (and simultaneous) detection of thrombi in the deep venous system and the right atrium along with the rapid sequence of embolic events in the absence of other underlying pathological conditions, point out the paradoxical embolism through a patent PFO as the most plausible scenario for the ischemic stroke.

Prothrombotic disorders

Prothrombotic disorders are frequently identified in pediatric patients with stroke [18] and case control studies demonstrate an association of arterial ischemic stroke in children with hereditary prothrombotic risk factors [15]. Another study reports prothrombotic abnormalities present in 20–50% of children with arterial ischemic stroke [22, 35].

Reasoning the coexistence of deep venous thrombosis and right atrial cavity thrombus in our patient with the absence of positive laboratory results for hypercoagulable disorders, we speculate either a transient hypercoagulable state, possibly associated with patient’s minor sport related injury or an unidentified mechanism by our thrombophilia screening.

Deep venous thrombosis and cryptogenic stroke

Young adults with cryptogenic ischemic stroke are more likely to have both patent foramen ovale and pelvic deep vein thrombosis (DVT) than young adults with ischemic stroke of known cause. Young patients with cryptogenic transient ischemic attack (TIA) or stroke and patent foramen ovale (PFO) should be evaluated for lower-extremity or pelvic venous thrombosis, which would be an indication for anticoagulation. In this case, screening for underlying causes of cryptic stroke with ultrasound of the lower extremities revealed venous thrombosis. The most probable releasing factor (and causative) of this event was a few days previously reported, sport’s related, minor lower extremity injury, a relationship supported by literature.
Right atrial thrombus and stroke

- Right-sided mobile thrombi in-transit from the deep venous system are found in adult case reports or case series in which clots were detected incidentally or during acute pulmonary thromboembolism.
- A recent pediatric literature review article reports in a sum of 122 cases, 91% of cases to be associated with central venous catheters, 40.8% in premature neonates, 27.2% in post cardiac surgery patients, and 19.2% to have underlying malignancies [36].
- In our case, the only causative mechanism for the formation of the right-sided thrombus, which was detected upon admission with esophageal ultrasound, was lower extremity deep venous thrombosis, in an otherwise healthy child. A Medline search of PubMed database using the keywords ‘right atrial thrombus’ and ‘children or pediatric or pediatric’ and ‘patent foramen ovale’ and ‘stroke’ did not reveal any relevant case so, to the best of our knowledge, this is the first documented case of right atrial thrombus resulting from deep venous thrombosis in a pediatric patient with patent foramen ovale and associated ischemic stroke event.

Treatment guidelines

Young patients with cryptogenic TIA or stroke and PFO should be evaluated for lower-extremity or pelvic venous thrombosis, which would be an indication for anticoagulation. In the setting of a large acute stroke, however, full-dose anticoagulation is not recommended, and an inferior vena cava filter may be the safest alternative. In patients with cryptogenic TIA or stroke, a PFO, and DVT, guidelines from the ACCP currently recommend VKA therapy for three months and consideration of PFO closure rather than no VKA therapy or aspirin therapy.

For patients with an ischemic stroke or TIA and both a PFO and a venous source of embolism, anticoagulation is indicated, depending on stroke characteristics (Class I; Level of Evidence A). When anticoagulation is contraindicated, an inferior vena cava filter is reasonable (Class IIa; Level of Evidence C). (New recommendation). In cases of concomitant venous and arterial embolism that paradoxical embolism is strongly considered, chronic anticoagulant therapy and an inferior vena cava filter can be justified to prevent further recurrences of both pulmonary and paradoxical embolism [37].

In the incident of right atrial thrombus, different treatment modalities are reported such as surgical thrombectomy, thrombolysis, anticoagulation therapy or observation only, the choice of which depended mainly on underlying etiology [36].

In this case, because of the hemorrhagic transformation of the cerebral stroke was an absolute contraindication for the initiation of fibrinolytic therapy, anticoagulation with subcutaneous low molecular weight warfarin along with foramen ovale umbrella placement constituted the selected treatment strategy. This strategy proved to be efficacious during the follow-up period.

Prognosis

Regarding outcome data, it is referred that permanent moderate-to-severe motor or cognitive disabilities occur in 75–87% of children with stroke, and death occurs in 5–28% [25].

In this case, patient presented with indirect symptoms, such as headache and epileptic fit, these symptoms appearing late from stroke ictus, as seen from the initial MRI presentation of the stroke which was in the hemorrhagic transformation phase and the patient was not on anticoagulation therapy for any reason or did not report aspirin uptake. Furthermore, she did not suffer any major clinical and neurologic sequelae from the event. These findings, possibly due to the clinically silent anatomical area of the stroke, are contrary to the majority of the cases described in literature that have unfavorable neurological prognosis.

CONCLUSION

Cryptogenic arterial ischemic stroke is a diagnosis of exclusion. The emergence of cases reporting patients with cryptogenic AIS harboring a patent foramen ovale, tends to reveal the presence of an associated causative factor. For all the aforementioned reasons, and because many of the aspects of the issue of AIS remain unresolved, we consider that it would be meaningful to present a case that mismatches a lot of aspects of the reported clinical cases and promotes a non-well elucidated pathophysiologic mechanism, supported widely by a lot of clinical and laboratory data.

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Author Contributions

Dimitrios Panagopoulos – 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

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

Georgios Markogiannakis – Acquisition of data, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published

Nikos Eleftherakis – Substantial contributions to conception and design, Analysis and interpretation of
data. Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

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REFERENCES

Esophageal dysmotility and abscess formation post Roux-en-Y bariatric surgery: A case report

Ryan D. Nicklas, Daniel S. Swink, Alyssa E. Heinrich, Christopher Boyd, Christina Li

ABSTRACT

Introduction: Abscess formation in the postoperative bariatric patient can be the result of anastomotic leak, a dreaded complication of Roux-en-Y gastric bypass. However, there is little data on bowel obstruction disrupting the anastomotic site resulting in leak and esophageal dysmotility. Case Report: A 50-year-old female status post laparoscopic Roux-en-Y presents with nausea, vomiting, and epigastric pain. Her clinical course was complicated with small-bowel obstruction, esophageal dysmotility, and an anastomotic leak with anaerobic abscess formation. The patient required a second operation to resolve the obstruction and a third operation to drain the resulting abdominal abscess. Conclusion: This case highlights a unique presentation of a post obstructive abscess following Roux-en-Y gastric bypass. A high index of suspicion should be maintained in postoperative bariatric patients presenting with symptoms of dysphagia and bowel obstruction without overt signs of infection to optimize prognosis and quality of care.

Keywords: Abscess, Esophageal dysmotility, Roux-en-Y

INTRODUCTION

Roux-en-Y is a safe and effective means of weight control in patient’s population who understand the potential risks, as well as the behavioral modifications required postoperatively. The procedure involves dividing the stomach and the jejunum and the rejoining the segments in a manner that bypasses a segment of the small bowel. By this process the absorptive length of the small bowel decreases and the volume of the remaining gastric pouch is restricted (Figure 1). There is convincing evidence that bariatric surgery can mitigate or even cure obesity related disease such as gastroesophageal reflux, sleep apnea, and type-2 diabetes. Furthermore, long-term mortality rates are lower for morbidly-obese patients who have undergone bariatric surgery than those who have not [1, 2]. However, it is important to be aware of the potential postoperative complications and management in these patient populations. The most frequently described complications include leak, obstruction, and anastomotic ulcer [2]. Another complication is abscess formation, which can result from seeding of intestinal contents into the abdomen during surgery, or spillage
from the anastomosis. Uncommonly, a bowel obstruction can lead to transient increase in intraluminal pressure leading to disruption of the anastomotic site. Both abscess and anastomotic leak require surgical intervention for complete resolution [2].

In this case, we aim to bring attention to the treatment and management of complex postoperative bariatric patients with obstruction, leak, dysmotility and consequential abscess formation.

CASE REPORT

A 50-year-old female presented with a past medical history of gastroesophageal reflux, hypertension, hypothyroidism, inflammatory polyarthropathy, and obesity with a BMI of 44 kg/m². Her surgical history included tonsillectomy, hysterectomy, cholecystectomy, and left knee replacement. Her medications included omeprazole, plaquenil, levothyroxine, and liothyronine. The patient underwent laparoscopic Roux-en-Y gastric bypass and hiatal hernia repair. She tolerated the procedure well, was functioning within normal limits, and was subsequently discharged on postoperative day-1. The patient was doing well on her follow-up appointment on postoperative day-5 where she was tolerating 80 oz of fluid a day; well above the minimum requirement. However, a few hours later the patient developed severe cramping epigastric pain, nausea, vomiting, and presented to the emergency department. The patient had a CT scan of her abdomen and pelvis, which showed obstruction at the level of jejunojejunostomy. There was no evidence of a leak or abscess (Figure 2). After intravenous fluid resuscitation, an X-ray of the abdomen and pelvis was performed eight hours later and showed transit of contrast into the ascending and transverse colon (Figure 3). However, given the lack of resolution of her symptoms, she was taken to the operating room urgently.

A diagnostic laparoscopy was performed revealing a single adhesion of her jejunojejunostomy down to the base of the mesentery, effectively torsing the distal Roux limb. An adhesiolysis was performed. The patient recovered well, tolerated fluids, and was discharged one day later. However, at her follow-up appointment 12 days post Roux-en-Y and six days post adhesiolysis, the patient again developed nausea, vomiting, epigastric pain and dysphagia. She denied fevers, was not tachycardic, and had no signs or symptoms suggestive of infection. An outpatient Gastrografin swallow test was performed. The films were interpreted as showing no evidence of anastomotic obstruction or leak. However, it was suggestive of significant narrowing of the gastrojejunal anastomosis site and marked distal esophageal dysmotility as there was a five-minute delay of transit from the distal esophagus into the gastric pouch (Figure 4). Due to her persistent symptoms and impaired oral intake, the patient was directly admitted to the hospital.

On admission, the patient received fluid resuscitation for dehydration and pharmacologic management of her nausea and vomiting with rectal Phenergan. The patient was afebrile, had a CBC and CMP within normal limits, and showed no other signs of infection on examination. Upper endoscopy was performed to rule out recurrence of hiatal hernia, stenosis from the repair, or anastomotic problems. It revealed no evidence of stricture or obstruction. Despite these findings, the patient’s persistent nausea, epigastric pain and clinical examination conflicted with these results. A diagnostic laparoscopy for possible revision of gastrojejunal anastomosis, evaluation of hiatal hernia repair, and possible endoscopy were planned for the next day.

During the laparoscopy, the jejunojejunostomy did not appear obstructed, although a large amount of edema
was appreciated around the distal gastric pouch and by the liver. While defining the borders of the gastrojejunal anastomosis and gastric pouch, there was a sudden rush of purulent fluid. A cavity of purulent fluid was revealed, extending below the left lobe of the liver and medial to the gastric pouch beneath the gastrohepatic ligament. The location of the abscess suggested possible impingement on the gastric pouch and pressure on the gastroesophageal junction. The purulence was completely aspirated, a Blake drain was placed in the abscess cavity, and a gastrostomy tube was placed in the gastric remnant for nutritional support. Intraoperative anastomotic leak test revealed no evidence of current leakage of contents into the abdomen. The remainder of the surgery was uneventful and the patient was placed on empiric piperacillin/tazobactam and infectious disease was consulted.

Culture of the abscess revealed *Hemophilus parainfluenza* and *Prevotella* species, normal oral flora. Infectious disease recommended modifying antibiotic therapy to a two-week course of ampicillin/sulbactam and a repeat abdominal CT scan to evaluate for remaining abscesses. Computed tomography scan of abdomen and pelvis with intravenous contrast taken on day-1 post laparoscopy showed an edematous, postoperative appearance of the stomach, with gastric tube and drain in good positioning, and no evidence of remaining abscess (Figure 5). Clinically, the patient reported improvement in her abdominal pain and nausea. The patient was advised to continue to meet fluid and protein requirements, with a combination of PO and G-tube feeds, and to advance diet to soft foods as tolerated. One week after discharge, the

Figure 3: Abdominal and pelvic X-ray showing contrast from recent computed tomography scan has progressed through the small bowel and is now present in the right colon and transverse colon. Taken on postoperative day-6.

Figure 4: Gastrograffin swallow study showing significant narrowing of the gastrojejunal anastomosis from edema and marked distal esophageal dysmotility. No evidence of the anastomotic stricture/obstruction or leak can be visualized, taken 6 days post adhesiolysis. Time elapse images A,B,C,D in series.

Figure 5: Computed tomographic scan of abdomen and pelvic with intravenous contrast showing postoperative appearance of the stomach. Percutaneous gastric tube and Jackson-Pratt drain are in place. No abscess identified, but small amount of free fluid is in the pelvis. Taken on day-1 post laparoscopy.
patient was seen in the office and was doing reasonably well, she reported relatively controlled gastroesophageal reflux and nausea with zantac and zofran, respectively. By this visit the patient had lost 30% of her excess body weight.

DISCUSSION

The American Society for Metabolic and Bariatric Surgery estimates that 23% of bariatric surgeries performed in 2015 were Roux-en-Y gastric bypass, making it the second most common bariatric procedure performed in the United States following gastric sleeve which comprised 54% of bariatric procedures [3]. Of Roux-en-Y procedures, less than 7% of laparoscopic cases are associated with major 30-day complications [4]. Exceptional in our patient, is the rapid onset of bowel obstruction, leading to progressive increase in intraluminal pressure, causing presumed disruption of the gastrojejun anastomosis and leak, ultimately resulting in abscess formation and esophageal dysmotility by mass effect (Table 1).

The patient’s initial postoperative hospitalization was for small bowel obstruction. Small bowel obstruction is a potential post Roux-en-Y gastric bypass complication. Small bowel obstruction occurs in 1–11% of patients postoperatively [5, 6]. The timing of obstruction is highly variable, but one study suggests an average onset of 7–19 days postoperatively for jejunojejunostomy obstructions [7]. Another study suggests a mean time to presentation of 313 days after Roux-en-Y bypass [8]. The most common cause of obstruction, occurring in 42–54% of cases is due to internal hernias [2, 7, 8]. These are caused by defects in the mesentery created during the operation. Other sources of obstruction include mesocolon scarring as well as adhesions. In our patient, the obstruction was the result of an adhesion located at the jejunojejunostomy occurring only five days postoperatively. Laparoscopic surgeries generally carry a lower risk of intra-abdominal adhesions compared to open surgeries due to less tissue damage, and less organ manipulation [2, 7]. While not specific; nausea, vomiting, and abdominal pain out of proportion to exam in the postoperative picture should lead to increased clinical suspicion for obstruction. Patients may be diagnosed with a CT scan and the treatment is early reoperation to prevent further complications [2, 7]. Our patient’s obstruction was relieved by performing adhesiolysis shortly after her presentation to the emergency department, with rapid resolution of her symptoms. However, it would later be discovered that the obstruction contributed to additional complications later in her clinical course.

The patient's return with nausea, vomiting, and impaired oral intake, prompted evaluation with a Gastrografin swallow study, which identified evidence of alterations in normal esophageal function. Esophageal dysmotility has multiple etiologies and is traditionally not associated with Roux-en-Y gastric bypass surgery. In fact, Roux-en-Y is the preferred bariatric surgery in patients with known esophageal disorders as there is evidence that it prevents and improves esophageal disorders [9, 10]. Etiologies include scleroderma, achalasia, and malignancy. Symptoms suggestive of esophageal dysmotility are variable and range from regurgitation, epigastric pain, and sensation of food being retained in the esophagus. The dysmotility our patient experienced was likely related to mass effect from her abscess on the gastroesophageal junction and gastric pouch. Management involved treatment of the underlying disease process, as evidenced by the resolution of the patient’s symptoms after surgical drainage of her abscess.

Although not supported radiographically, the patient’s clinical course and microbiologic evidence was highly suspicious for occult anastomotic leak. Anastomotic leak is a feared complication of gastric bypass, carrying a mortality rate of nearly 10%, but occurs in less than 0.8% of cases within the first 30 postoperative days [11]. Leakage of gastric contents from an anastomotic site can lead to contained infection/abscess or even lead to diffuse peritonitis. Signs of leak traditionally manifest within the first postoperative week, and vary in presentation from tachycardia, tachypnea, and abdominal pain. Our patient’s leak demonstrated an uncommon presentation, given that occurred 12 days post Roux-en-Y and was accompanied with evidence of esophageal dysmotility. Instead of overt signs of peritoneal irritation or infection, the patient had a normal white count and was afebrile without tachycardia or tachypnea. Given the timing of these events and absence of radiographic and intraoperative evidence of anastomotic leak throughout

<table>
<thead>
<tr>
<th>Postoperative Day (POD)</th>
<th>Clinical Events</th>
</tr>
</thead>
<tbody>
<tr>
<td>Surgery</td>
<td>Roux-en-Y gastric bypass and hiatal hernia repair</td>
</tr>
<tr>
<td>POD 1</td>
<td>Discharge home from hospital</td>
</tr>
<tr>
<td>POD 5</td>
<td>Normal office visit, return to ER that night with acute abdominal pain and admitted for small bowel obstruction</td>
</tr>
<tr>
<td>POD 6</td>
<td>Exploratory laparoscopy and adhesiolysis</td>
</tr>
<tr>
<td>POD 7</td>
<td>Discharge home from hospital</td>
</tr>
<tr>
<td>POD 12</td>
<td>Clinic follow up with nausea, vomiting, dysphagia, upper GI series with 5 min delay, admitted to hospital, EGD to rule out acute anastomotic stricture and obstruction from hernia repair</td>
</tr>
<tr>
<td>POD 13</td>
<td>Exploratory laparoscopy, abscess drainage, G-tube placement</td>
</tr>
<tr>
<td>POD 14</td>
<td>CT no evidence of remaining abscess</td>
</tr>
<tr>
<td>POD 21</td>
<td>Discharge home from hospital</td>
</tr>
<tr>
<td>POD 28</td>
<td>Clinic follow up doing well</td>
</tr>
</tbody>
</table>
the case, it is reasonable to believe that an increase in intraluminal pressure during complete obstruction caused tension on the anastomosis leading to transient failure of the junction and subsequent seeding of gastric contents into the lesser sac [12]. Due to the limited length of her obstruction and surgical intervention, the path of least resistance again became the Roux limb and the leak sealed spontaneously. Management of leak includes aggressive evaluation for signs and symptoms of early sepsis, imaging with CT or upper gastrointestinal study, and diagnostic laparoscopy if tests are inconclusive [2]. In our patient, operative abscess evacuation with drain placement, anaerobic antibiotic coverage, and G-tube placement for early feeding were important components of care. Ironically, treatment of this patient's leak began with her adhesiolysis performed seven days prior to her current presentation.

CONCLUSION

In conclusion, this patient initially presented with a transient small-bowel obstruction resulting from an adhesion, leading to a transient increase in intraluminal pressure. We theorize that this resulted in a brief disruption of the anastomotic site, which then progressed to an abscess. Interestingly, instead of appearing with an acute infectious process, the patient presented with esophageal dysmotility. In discussing this case, we illustrate a unique constellation of events following Roux-en-Y gastric bypass surgery.

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REFERENCES


Jejunal angiodysplasia: A rare cause of obscure gastrointestinal bleeding

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ABSTRACT

Introduction: Obscure gastrointestinal bleeding (OGIB) represents about 5% of all gastrointestinal bleeds. This can pose a huge diagnostic challenge for clinicians. Small bowel angiodysplasia is a rare but important cause of OGIB that is difficult to diagnose and treat.

Case Report: We present a case of a 53-year-old female with a one-month history of melena and symptomatic anemia who presented to general hospitals in Yangon, Myanmar. After multiple investigations, she was diagnosed with jejunal angiodysplasia. We highlight the challenges that come with diagnosing and managing a rare but important cause of upper gastrointestinal bleeding, made even more difficult in a resource-limited setting where healthcare is not always affordable and accessible. Conclusion: Small bowel angiodysplasia should be on the differential list for all patients who present with obscure gastrointestinal bleeding or any gastrointestinal bleeding. Early diagnosis is important so that appropriate treatment can be administered for this potentially life-threatening condition.

Keywords: Jejunal angiodysplasia, Obstructive gastrointestinal bleeding

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INTRODUCTION

Angiodysplasia is a type of vascular malformation with fragile and leaky vessels that can affect anywhere along the gastrointestinal tract. It is responsible for about 6% of lower gastrointestinal bleeding and between 1.2–8% of upper gastrointestinal bleeding [1]. Obscure gastrointestinal bleeding (OGIB) represents about 5% of all gastrointestinal bleeds [2], with small bowel angiodysplasia estimated to account for around 30–40% of cases [3]. Thus, it is an important differential that must be considered. Clinically, its presentation can vary from being an incidental finding to OGIB or chronic anemia. These patients should be evaluated with oesophagogastrroduodenoscopy (OGD), colonoscopy, video capsule endoscopy (VCE) and angiography. Additionally, a red blood cell scan can increase the sensitivity of diagnosis when used in combination with angiography [4, 5]. In acute presentations, endoscopic therapy should be used to treat active bleeding from angiodysplasia. Options include argon plasma coagulation...
(APC), sclerosant injections and gel foam embolization. Double balloon enteroscopy (DBE) can be used for both diagnostic and therapeutic purposes. For long-term management, somatostatin analogues, hormonal therapy or anti-angiogenics can be used [6]. In refractory bleeding or if lesions are at inaccessible sites, surgical localization and resection of the diseased segment can be performed.

**CASE REPORT**

A 53-year-old female presented to Thingangyun Sanpya General Hospital, Yangon, Myanmar in February 2017 with a one-month history of increasing dyspnea, palpitations and pallor. She recalled a few episodes of melena. There were no other cardiac, respiratory or gastrointestinal symptoms. Apart from severe conjunctival pallor, physical examination and digital rectal examination were unremarkable. Blood tests revealed a hypochromic microcytic anemia (Hb 5.3, MCV 53.4) and she was found to have HbE trait on hemoglobin electrophoresis. She was transfused 5 units of blood. An OGD performed showed a duodenal ulcer and gastric vascular ectasia which was subsequently treated with argon plasma coagulation (APC). However, these findings could not explain her severe anemia and a colonoscopy was performed in March 2017. The colonoscopy revealed small polyps in the descending and sigmoid colon which were tubular adenomas on histology. These findings still could not explain her severe anemia. A month later, video capsule endoscopy was performed to locate potential sources of bleeding. This showed active small bowel bleeding in the last part of the duodenum and jejunum (Figure 1). An ultra-slim colonoscope was then used to better visualize the small bowel, showing multiple angiodysplastic spots with active bleeding in the proximal jejunum. Argon plasma coagulation was applied and a diagnosis of jejunal angiodysplasia was made (Figure 2).

The patient presented again, this time to Yangon General Hospital, Yangon, Myanmar towards the end of April 2017 with similar worsening anemic symptoms (Hb 3.5). She was transfused 4 units of blood, stabilized and discharged. A computed tomography angiography (mesenteric angiogram) was performed in May 2017. The result showed contrast-enhancing wall thickening at the second part of the duodenum, but no serpiginous vascular channels or contrast extravasation was observed. Due to the equivocal results, a slim colonoscope was used to visualize the jejunum again, and this was normal up to the duodenal-jejunal junction. The patient was transfused 2 units of blood, discharged and followed-up in clinic, with a plan for a DBE if symptomatic anemia recurred.

**DISCUSSION**

Small bowel angiodysplasia is a rare but important cause of OGIB or upper gastrointestinal bleeding. It should be considered in the differential list for gastrointestinal bleeding and chronic anemia, especially in patients with negative OGD and colonoscopies [3]. In this case, multiple gastrointestinal pathologies were initially discovered in this patient on OGD and colonoscopy. Sound clinical judgment was required to deduce that the findings could not explain the patient’s clinical presentation, hence further investigations were undertaken. In all, there was a four-month time period between symptom onset and final diagnosis. Small bowel angiodysplasia is a challenging condition to formally diagnose; video capsule endoscopy and computed tomography angiography play a key role, and these should be requested in patients with negative gastrointestinal scope procedures, or with results that do not correlate with clinical presentation.

In this case, however, computed tomography angiography results were equivocal for angiodysplasia. This could be because the patient was not actively bleeding at the time of investigation. Additionally, radiologists recommended that the gastrointestinal hemorrhage computed tomography angiogram protocol should have been specifically requested according to local hospital guidelines. This should be stated on the request form to prompt radiologists to focus particularly on gastrointestinal mucosal vessels. A multidisciplinary team meeting was later conducted, with the team concluding...
that there was enough evidence for a formal diagnosis of jejunal angiodysplasia to be made despite the equivocal angiogram. This is a rare condition not commonly encountered by healthcare professionals, hence many were unfamiliar with diagnostic methods. A multidisciplinary team approach involving gastroenterologists, general surgeons and emergency physicians must be adopted early to ensure quick diagnosis and management of this potentially life-threatening condition.

In terms of treatment, our patient was mainly managed supportively with blood transfusions when indicated. Endoscopic intervention via APC was only performed once. However, the patient’s symptoms recurred over four months. Long-term therapies such as octreotide could be considered in this case, and further surgical intervention should symptoms continue to persist.

Many problems exist in the field of healthcare in Myanmar, including poor access to hospitals and a shortage of resources and health professionals [7]. There are a total of only five public endoscopy centers in the entire country and a shortage of gastroenterologists, which is insufficient for a population of over 51 million people. Private facilities are available, but many people live in poverty, and they are often unable to afford these services. In this case, the delay from presentation to diagnosis could be explained by the challenging nature of the illness, and also the difficulty in accessing relevant investigations more quickly. Doctors in resource-limited settings like these are forced to rely more on their clinical judgment in making diagnoses. They must also be more familiar with local hospital protocols and stay up to date with their knowledge given the lack of national guidelines and rare presentation of this case.

CONCLUSION

In conclusion, small bowel angiodysplasia is a rare but important cause of obscure gastrointestinal bleeding (OGIB). It should be on the differential list for patients presenting with any gastrointestinal bleeding. There should be a higher suspicion for small bowel angiodysplasia if oesophagogastroduodenoscopy (OGD) and colonoscopy yield inconclusive results. Video capsule endoscopy and computed tomography angiography are useful investigations for its diagnosis. In the acute setting, management options include supportive blood transfusions and interventional endoscopy. Double balloon enteroscopy is a useful tool that can be considered for both diagnostic and therapeutic purposes. In resource-limited settings, it is important for doctors to be familiar with this condition and the diagnostic and management options available.

Author Contributions
Gareth Zigui Lim – 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
Than Than Aye – 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
The corresponding author is the guarantor of submission.

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Authors declare no conflict of interest.

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REFERENCES
Complete spontaneous resolution of vanishing lung syndrome: A rare case

Roy Cho, Felix D. Zamora, Heidi H. Gibson, Erhan Dincer

CASE REPORT

A 59-year-old male with chronic obstructive pulmonary disease (COPD) returned to the pulmonary clinic three-years after initial evaluation for bronchoscopic lung volume reduction for a giant left upper lobe bullae. He had moderately severe COPD (FEV1 1.6L, 37%) controlled on inhaler therapy and a greater than 35-pack/year smoking history. Previously, he was deemed a candidate for lung volume reduction. However, declined and was lost to follow-up (Figure 1). On presentation, he denied any decline in his physical capacity for the past three-years. His repeat chest CT scan demonstrated complete resolution of the large left upper lobe bullae. As a result of these findings, we did not recommend any further intervention with regard to the previous bullae. However, recommended annual lung cancer surveillance with low-dose chest CT scan.

DISCUSSION

Giant pulmonary bullae or vanishing lung syndrome (VLS) is defined by radiographic criteria including presence of giant bullae in one or both upper lobes, occupying at least one-third of the hemithorax and compressing surrounding normal lung parenchyma [1].

The natural history of VLS is unpredictable and is based on case reports and experience forming expert opinion. The leading theory for bullae expansion is air trapping that impedes expiratory air-flow leading to tension and gradual enlargement of the air spaces [2]. Although most bullae enlarge, only three-case reports since 1990 have shown regression of the disease [3–5]. These cases report partial regression following instances of airway obstruction (infection, inflammation, etc.); in which, the leading hypothesis is complete isolation of the space leading to shrinkage via air and fluid resorption over time. Notably, this is the first case to demonstrate complete resolution of VLS radiographically.

Figure 1: Chest computed tomography scan at the level of the upper lobe, carina, and lower lobes. A very large left upper lobe bulla is observed (left) with spontaneous resolution after three-years follow-up (right).
CONCLUSION

Usual course for vanishing lung syndrome (VLS) is progression of disease and worsening pulmonary function as measured by FEV1. However, we report a very rare case of VLS that has completely resolved without any intervention.

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Keywords: Emphysematous bullae, Interventional pulmonary, Vanishing lung syndrome

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