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Laparoscopic treatment of ovarian vein syndrome: A case series

Wissem Hmida, Mouna Ben Othmen, Faouzi Mallat, Sidiya Oueld Chavey, Mehdi Jaidane, Faouzi Mosbah

ABSTRACT

Introduction: Ovarian vein syndrome is a rare entity of ureteral obstruction. Its pathogeny and clinical expression are highly polymorphic. The treatment gains great advances because of the development of laparoscopy. Case Series: We reported two cases of ovarian vein syndrome occurring in two multiparous women, with unremarkable past medical history. The patients were presented with isolated right flank pain. The diagnosis was confirmed by computed tomography angiography in the two cases. Ovarian vein ligation was successfully performed throughout a retroperitoneoscopic approach with excellent outcomes. Conclusion: The ovarian vein syndrome continues to be a rare diagnosis that should be recognized. The diagnosis is mainly urographic. Owing to its simplicity, low morbidity, and good results attained, the laparoscopic approach will continue to advance the surgical management of ovarian vein syndrome.

Keywords: Ovarian vein syndrome, Urographic, Laparoscopic approach

INTRODUCTION

Ovarian vein syndrome is an uncommon cause of ureteral obstruction caused by an aberrant dilated ovarian vein [1]. It is a poorly understood clinicopathological condition first described in 1964 [2]. Typically occurring in young multiparous woman with a right-sided predilection. The symptoms are non-specific including acute or chronic lumbar pain. The diagnosis is urographic. The treatment gains great advances because of the development of laparoscopy.

CASE SERIES

We reported two cases of ovarian vein syndrome treated by laparoscopic approach.

Case 1: A 43-year-old female with obstetric history of three gestations, presented with a 24-month history of recurrent right flank pain. She denied having a history of gross hematuria or urinary tract infection. On physical examination, BMI was at 17. Laboratory tests revealed a creatinine 90 μmol/L. The urine analysis was normal.

Ultrasound followed by computed tomography (Figure 1) showed a moderate dilatation of the right upper urinary tract and confirmed the diagnosis of right ovarian vein syndrome.

Through a retroperitoneoscopic approach, the ovarian vein was dissected, ligated and resected. The mean operating time was one hour. Preoperative and
postoperative period were uneventful, and the patient was discharged after 48 hours of surgery.

In a follow-up of nine months the patient remained asymptomatic. Urine cultures repeated every three months were negative and there were no radiologic signs of ureteral obstruction.

**Case 2:** A 38-year-old female with obstetric history of two gestations, presented with a nine-month history of recurrent right lumbar pain. She reported that the pain was exacerbated in premenstrual period, but she did not have any history of hematuria or urinary tract infection. The physical examination did not reveal any abnormality. Laboratory tests showed a negative urine culture and a creatinine value of 65 μmol/L.

Imaging investigations showed moderate dilation of upper right urinary tract. The diagnosis was confirmed by computed tomography angiography revealing the compression of the right ureter by a dilated ovarian vein measuring 9 mm in diameter. The patient underwent a ligation of ovarian vein throughout a retroperitoneoscopic approach. The procedure was successfully performed and the mean operating time was 55 minutes. Postoperative outcomes were good and the patient was discharged after 48 hours. In a follow-up of two years, the patient was asymptomatic with resolution of obstruction in radiologic finding.

**DISCUSSION**

Ovarian vein syndrome is a rare cause of ureteral obstruction [1]. In 1964, Clark reported a series of 129 right-sided ovarian vein syndromes. Many authors have published case reports of the ovarian vein syndrome, but the largest study includes only eight cases [2]. It is classically described on the right side in 95% of cases [3, 4].

The pathophysiology of ovarian vein syndrome is still poorly understood. Several mechanisms have been suggested [2], mainly a ureteral compression by an aberrant ovarian vein draining into the right renal vein, hormonal changes associated with pregnancy may also explain ovarian vein syndrome [2, 5, 6]. It is probably a multifactorial syndrome [7].

The symptoms appear frequently in multiparous women, but have been also described in nulliparous women and children [2, 8–12]. The clinical features have no specificity including an acute or chronic lumbar pain, recurrent urinary tract infection, and frank hematuria [2, 9, 13]. Typically, the pain is exacerbated in premenstrual period or during pregnancy [7, 4]. In this cases, the two patients were presented with isolated right lumbar pain; one of the exacerbated in premenstrual period.

A careful preoperative evaluation must be required to eliminate other ureteral obstruction causes such as tumor compression and retroperitoneal fibrosis [1]. That is why some radiological examinations were indicated to confirm the diagnosis of ovarian vein syndrome [4]. The abdominal ultrasound showed generally moderate dilatation of the upper urinary tract. Transvaginal ultrasound can reveal dilated ovarian veins [7].

The gold standard for diagnosis was intravenous urography showing typically dilation and tortuosity of upper ureter with transverse defect at L3/L4 level [2, 7].

The computed tomography angiography revealed the crossing ovarian vein and excludes other causes of ureteral compression such as tumor compression retroperitoneal fibrosis [8].

In the case of our patients, the diagnosis was confirmed by computed tomography angiography.

Various management options for ovarian vein syndrome have been described; including conservative measures (medical treatment and embolization), and surgical excision, Wish represent the radical treatment of this entity [14].

Traditionally, ligation of ovarian vein has been performed through an open surgery. However, the laparoscopic approach has gained traction since the first report of transperitoneal laparoscopic ovarian vein ligation published by Elashry et al. in 1996.

The laparoscopic treatment, as a minimally invasive alternative to the open surgery [8]. It had progressed well recently to involve use of retroperitoneoscopic approach [2]. It had proven to be a valuable technique, offering superior visualization of the operative field. It permits careful assessment of the periureteral anatomy and identification of the ovarian vein [1].

It may limit the risk of hemorrhagic complications [1], reducing postoperative pain and analgesic requirement improvement in convalescence time and patient outcomes [1, 8, 15, 16]. As seen in these cases, the ligation of the ovarian vein and ureteral dissection by retroperitoneoscopic approach was easy, with an excellent immediate and long-term outcomes and the patients being discharged after a short hospital stay.
CONCLUSION

The ovarian vein syndrome continues to be a rare diagnosis that should be recognized. The diagnosis is mainly urographic. Owing to its simplicity, low morbidity, and good results attained, the retroperitoneoscopic approach will continue to advance the surgical management of ovarian vein syndrome.

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Sidiya Oueld Chavey – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Final approval of the version to be published

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The corresponding author is the guarantor of submission.

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The use of a novel neuromuscular electrical stimulation device in peripheral vascular disease

Katherine J. Williams, Alun H. Davies

ABSTRACT

Introduction: Enhancement of peripheral circulation using electrical devices has demonstrated benefit in many vascular disorders.

Case Series: Herein, we present a case series of three complex patients illustrating the successful use of a new single-use disposable neuromuscular electrical stimulation (NMES) device (geko™, Firstkind Ltd, UK). Cases include the management of recalcitrant ulcers, non-reconstructable critical limb ischemia, and an infected arterial bypass graft.

Conclusion: Neuromuscular electrical stimulation (NMES) can potentially enhance peripheral circulation in vascular patients. Difficult or recalcitrant vascular cases may benefit from it as an adjunct to best medical care. NMES has few side effects, and may be especially useful where polypharmacy is an issue. The incidence of a skin reaction may necessitate device discontinuation.

Keywords: Neuromuscular electrical stimulation, NMES, EMS, Vascular disease

INTRODUCTION

Neuromuscular electrical stimulation (NMES) can be used to enhance circulation, and may be a useful adjunct in the management of vascular disorders [1]. Artificial augmentation of blood flow, using intermittent pneumatic compression, has been shown to prevent in-patient venous thromboembolic events, aid vascular ulcer healing, increase walking distances in arteriopathics, and reduce limb swelling [2–5]. NMES can be used to enhance circulation, and may be a useful medical adjunct in both the in-patient and out-patient setting. We present a case series illustrating the use of an NMES device in complex patients.

CASE SERIES

Case 1: A 90-year-old female presented to outpatient clinic with a large ulcer in her right ankle gaiter area. She had a longstanding diagnosis of mixed peripheral venous and arterial disease, with recurrent hard to heal ulcers. The current wound had previously been treated with vacuum therapy, compression bandaging and honey dressings for six months. She had required repeated admissions to hospital for intravenous antibiotics for infection of this recalcitrant ulcer. Compression bandaging was poorly tolerated at home and had to be discontinued due to clinical progression of peripheral arterial disease. An NMES device was applied unilaterally...
to the right leg, and operated for 24 hours a day for 14 days. At the end of this period, ulcer characteristics were much more favorable, with active granulation tissue and a significant improvement in depth of ulcer. Wound size decreased in surface area from 64.6–18.9 cm² (Figure 1). Allergic reaction (simple rash) to device adhesive caused discontinuation of device use, but with simple dressings for a further four weeks the wound healed completely. The rash resolved spontaneously.

**Case 2:** A 64-year-old diabetic male with critical limb ischemia of his right leg (ABPI 0.45) was given NMES therapy for eight weeks. He was a unilateral above-knee amputee with a history of endocarditis, aortic and pulmonary valve repair, malaria and hepatitis. He had undergone multiple right lower limb angioplasties and a failed axillo-femoral bypass. Further reconstruction was deemed technically inappropriate. His regular medications included gliclazide, metformin, beta blockers, and regular high dose analgesia. He was trained in how to fit the device and wore it for 6–10 hours per day for six weeks.

Patient progress was followed using calculated leg volumes (perometer, from knee crease to sole of foot), the intermittent claudication questionnaire (ICQ), and patient reported quality of life questionnaires.

At six weeks leg swelling was reduced (3602–3480 mL), patient reported quality of life had improved (Visual Analogue Score, a subjective measure of health, increased from 75–90, and depression score, CES-D, decreased from 24–18), but ICQ showed a slight worsening of symptoms (78.5–81.6). The patient expressed pleasure in wearing the device, and reported increased motivation to leave the house in his wheelchair. No side effects were reported.

**Case 3:** A 78-year-old male with a history of peripheral vascular disease presented to the emergency department with wet gangrene of his right forefoot secondary to critical ischemia, complicated by systemic sepsis. He had a background of insulin-dependent diabetes, ischemic heart disease, atrial fibrillation, hypertension, and rheumatoid arthritis. The patient was resuscitated, admitted and right femoral to posterior tibial bypass was performed with PTFE graft and vein cuff. An angioplasty with drug eluting stent of the right posterior tibial artery was performed. Fasciotomies, were performed, as were amputations of the first, second, and third toes of the forefoot. An intravenous heparin infusion was administered overnight and warfarin commenced. His admission was complicated by severe respiratory tract infection requiring non-invasive ventilatory support, and difficult glycemic control. An episode of cholecystitis was treated by the general surgical team and the patient underwent a laparoscopic cholecystectomy. Postoperative limb swelling was problematic, forefoot wound healing and mobility were poor, and the arterial graft became infected. A multidisciplinary team decision was made to treat this conservatively with antibiotics. A negative pressure dressing (VAC pump, KCI) was applied to the forefoot wound. NMES devices were applied bilaterally and activated continuously for two weeks. Each device battery lasts for approximately 24 hours, and his wife was trained in how to remove the device and re-fit a new one (Figure 2).

Swelling reduced in both legs, maximal diameter right calf 41–33 cm, left calf 37–34 cm but returned to the initial postoperative values when NMES was discontinued. No device side effects were reported. His right forefoot amputation wound and a left heel pressure sore healed completely over six weeks. After a period of physiotherapy the patient was discharged with a walking aid. Graft patency was confirmed by ultrasound at discharge.

**The NMES Device**

All cases reported here were treated with the geko™ device (Firstkind Ltd, UK), applied transcutaneously over the common peroneal nerve at the knee (Figures 3 and 4). It is portable and disposable, with a self-adhesive strip on
Activation of the device causes excitation of the motor nerve supplying the muscles of the anterior and lateral compartments of the leg, and causes the foot to dorsiflex in a twitching motion, frequency 1 Hz. Pulse width can be adjusted for strength of desired effect. Treatment protocols are flexible according to indication, but we have found that longer periods of stimulation may predispose to contact allergy. Topical emollients are recommended for use on the application site after removal of the device. Correct application of the device requires training, and this normally takes an average of 10–15 minutes. Once trained the patient or their family can apply the device themselves each day.

DISCUSSION

The management of complex patients requires knowledge of many different treatment modalities, with tailoring to the individual patient to achieve optimum results. With this case series we have illustrated the utility of NMES as an adjunct to standard care, in different cohorts of patients. It has been shown to potentially aid healing of treatment-resistant ulcers, help reduce limb swelling, and increase quality of life in complex arteriopathies. By causing contraction of the leg muscles, increasing venous return, and removing dependent fluid from the limb, the potential clinical applications of this device are huge. In contrast to pharmaceutical and surgical interventions, this medical device is easy to apply and has few side effects. There are no reported drug interactions, which is important in vascular patients where polypharmacy is common. The efficacy of the device has not yet been evaluated in large scale clinical trials, but they are awaited eagerly.

CONCLUSION

Neuromuscular electrical stimulation (NMES) can potentially enhance peripheral circulation in vascular patients. Difficult or recalcitrant cases may benefit from NMES as an adjunct to best medical care. NMES has few side effects, but skin rash may necessitate device discontinuation.

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Katherine J. Williams – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published
Alun H. Davies – Conception and design, Critical revision of the article, Final approval of the version to be published

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Giant cell tumor of bone: Report of three cases with unusual locations

Nimisha Batra, Virinder Mohan

ABSTRACT

Introduction: Giant Cell tumor (GCT) is an uncommon neoplasm. The tumor typically affects the ends of long bones and involvement of flat bones of pelvis and greater trochanter is extremely rare. Case Series: In this case series, three cases suspected of having GCT on the basis of their characteristic radiological findings have been reported. However, they presented at unusual locations, i.e., ilium and greater trochanter of femur. Two of the cases were males and one was female. These patients presented with pain localized to the affected region, were examined clinically and relevant investigations were done. Clinical history, clinical examination, laboratory investigations and imaging findings have been discussed in detail. The patients were on follow-up and radiological diagnosis of GCT was confirmed by histopathology. Conclusion: The aim of this case series is that GCT at not so common locations can be diagnosed on the basis of their characteristic radiological findings and computed tomography scan is the choice of investigation to see the expansile nature of the lesion.

INTRODUCTION

Giant cell tumor (GCT) is a mostly benign and an uncommon neoplasm of bone of uncertain origin that accounts for approximately 4–5% of all bone tumors [1]. As previously described, 80% of GCTs have a benign course with a local recurrence rate of 20–50%. The tumor typically affects the ends of long bones, most commonly the distal femur, proximal tibia, distal radius, and proximal humerus in that order. Involvement of the flat bones of the pelvis or the greater trochanter is extremely rare [2, 3].

The purpose of this presentation is to report three radiologically diagnosed and histologically proven cases of GCT at unusual locations.

CASE SERIES

Case 1: A 55-year-old female was admitted in the surgical services with the chief complaints of lower abdominal pain and mass in the right iliac fossa since one month. The pain was continuous, diffuse, dull aching with no radiation to the back, shoulder, or groin. There was no history of fever, vomiting, loose stools, constipation or abdominal injury.

On general examination the pulse and blood pressure were within normal limits. There were no pallor, clubbing, cyanosis, icterus, lymphadenopathy and edema. Local examination revealed a soft tissue mass in the right iliac
fossa with localized tenderness. The mass was firm, non-mobile and had indistinct borders. There was no evidence of redness, rise in temperature or engorged vessels over the swelling. Abdominal examination was unremarkable with normal bowel sounds.

Laboratory investigations revealed hemoglobin level of 10 g/dL. The rest of the investigations were unremarkable.

With the clinical diagnosis of a right iliac fossa lump, the patient was sent for computed tomography (CT) scan of the lower abdomen which revealed an expansile lytic lesion involving the right iliac wing, causing cortical thinning but no breach in the cortex. There were no matrix calcifications. The lesion was associated with a large soft tissue component (Figure 1).

The characteristic features of the bony lesion were suggestive of GCT of the right iliac bone, which was confirmed on histopathology.

**Case 2:** A 49-year-old male was admitted in the orthopedic services with chief complaint of pain and swelling over the right upper thigh region. The patient had difficulty in walking. The right lower extremity was in spontaneous abduction and external rotation. There was no history of trauma or any other relevant history. The general physical examination was unremarkable. On local examination, there was a tender swelling over the right thigh in the region of the greater trochanter. The skin over the swelling was normal in color and temperature.

Laboratory investigations were unremarkable.

X-ray right hip joint revealed a lytic, expansile lesion of the greater trochanter, with cortical destruction and thinning of the cortex (Figure 2). There were no sclerotic margins or periosteal reaction. The lesion was associated with a large soft tissue mass.

Computed tomography scan of right upper thigh revealed a large expansile lesion of the greater trochanter causing cortical thinning and breach in the cortex and associated with soft tissue swelling (Figure 3).

![Figure 1: Computed tomography scan of lower abdomen with oral contrast showing an expansile lytic lesion in the right iliac blade with thinning of cortex and a large soft tissue component.](image1)

![Figure 2: X-ray right hip joint showing a lytic expansile lesion of the greater trochanter with thinning and destruction of cortex. There is no periosteal reaction or any calcification within the soft tissue.](image2)

![Figure 3: Computed tomography scan of right thigh showing an expansile lesion in the greater trochanter causing cortical thinning and breach with associated soft tissue component.](image3)

A Radiological diagnosis of GCT of the greater trochanter was confirmed on histopathology.

**Case 3:** A 45-year-old male presented with the chief complaints of pain and mass in left flank since last two months. The pain was continuous and dull aching in
nature with no radiation. However, pain increased while walking. There were no associated complaints of fever, loose stools, constipation or abdominal injury.

General physical examination was normal and local examination revealed a hard, non-tender, non-mobile swelling with smooth margins. The skin over the swelling was normal in color and temperature. Laboratory investigations were reported normal.

X-ray pelvis revealed a large lytic expansile lesion with few septations and well defined margins arising from the left ilium showing typical “soap bubble” appearance (Figure 4). Rest of the pelvic bones appeared normal.

Radiological diagnosis of GCT of left ilium was confirmed by histopathology.

DISCUSSION

Giant cell tumor is also known as osteoclastoma. It is an aggressive lesion of bone characterized by highly vascular tissue containing proliferating mononuclear stromal cells and numerous uniformly distributed giant cells of the osteoclast type [4]. It typically occurs in young and middle aged adults after the growth plate closes. Its frequency decreases in the later decades of life, and it is extremely rare in patients over 70 years of age [1]. The majority of these lesions (60%) occur in long bones, and mostly all are localized to the articular end of the bone. The most common sites include the proximal tibia, distal femur, distal radius, and proximal humerus [4]. GCT may rarely occur in flat bones or apophysis, which is an epiphyseal equivalent. However, when this happens, the lesion is less likely to demonstrate the classic appearance of a lytic lesion with a well-defined, non-sclerotic margin. Only few cases of Trochanteric location have been found as reported by Lichtinger (2004) and Gebhardt (2005) [5–7]. In a series of 70 giant cell tumors by Shankman et al. (1988), only two occurred in ischium, two occurred in ilium and none in the pubis. Overall, approximately 4% of all GCTs occur in innominate bones [1]. In the pelvis, the ilium is the most frequent site of giant cell (excluding the sacrum) [1]. Balke et al. (2009), in their case series of 20 patients of giant cell tumors of pelvic bone over a period of 20 years found only nine cases involving the ilium [8]. In another series of seven patients of GCT of innominate bones by Kattapuram et al. (1996), only one case involving the ilio-sacral region was identified [9]. Giant cell tumor of the pelvic bone mostly occurs in third or fourth decade of life with a clear female predilection [2].

Giant cell tumor may occur in the skull or pelvic secondary to Paget disease. The bones of the hands and feet are uncommon locations, with a prevalence of less than 2% [10]. Multicentric GCT has been reported in less than 1% of cases, with lesions often located in the distal extremities. Lung metastasis have been reported in 1–6% of cases. Rarely, GCT may undergo malignant transformation. This may occur as a result of differentiation of the primary tumor or secondary to prior radiation therapy. The overall prevalence is less than 1% [5].

Clinical symptoms in patients with solitary lesions are non-specific. These include pain, local swelling and limitation of range of motion in the adjacent joint [4].

Giant cell tumor of bone is characterized radiographically as an expansile lytic lesion with well defined margins. It may extend beyond the cortex. Most tumors demonstrate absence of a sclerotic border (unless the tumor has been presented for a long time), a lack of periosteal reaction, and the absence of calcified or ossified matrix [1]. The GCT of pelvic bones are generally lytic and are associated with large soft tissue mass which resembles an aggressive lesion demonstrating increased vascularity [9]. Computed tomography is the best modality to show the expansile nature of the tumor, while magnetic resonance imaging scan is superior in showing the soft tissue extension [2].

When a giant cell tumor occurs in unusual locations in elderly individual, it may easily be confused radiographically with expansile metastatic disease, plasmacytoma, malignant fibrous histiocytosis, or fibrosarcoma [11]. However a slow growing lesion with expansion and typical location with or without associated soft tissue mass in the above described clinical setting is almost diagnostic of the disease.

The treatment of choice of GCT involving pelvic bones is curettage with or without resection and/or with adjuvant therapy such as cryotherapy, phenol, or polymethylmethacrylate (PMMA) to minimize the incidence of recurrence [2, 7]. Reconstructive surgery using a bone allograft after complete resection have been reported [6]. In advanced cases of aggressive lesions and extensive soft tissue involvement, wide resection even up to the extent of amputation may be necessary for complete cure [2].
CONCLUSION

Giant cell tumor (GCT) is an uncommon, benign tumor of the bone. Pelvic GCT is not commonly encountered. However, when an expansile, lytic lesion with cortical thinning and destruction is seen associated with a large soft tissue component, a radiological diagnosis of GCT can be made. The ilium is the most commonly affected among pelvic bones. Computed tomography is the investigation of choice to see the expansile nature whereas magnetic resonance imaging scan shows soft tissue extension.

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Author Contributions

Nimisha Batra – Substantial contribution to concept and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Virinder Mohan – Substantial contribution to concept and design, Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published

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REFERENCES

A rare case of huge intrahepatic portal vein aneurysm

Muhammet Battal, Mustafa Ozer Ulukan, Burhan Akdana, Rabia Karasu, Uğur Temel

ABSTRACT

Introduction: Our case was incidentally diagnosed large aneurysm of the main portal vein. The incidence of intrahepatic and extrahepatic portal vein aneurysms (PVAs) is not clear. Portal vein aneurysm usually occurs at the junction of the superior mesenteric vein and splenic veins or at the hepatic hilus at the bifurcation of the right and left portal veins [1]. Case Report: Color Doppler and contrast-enhanced dynamic computed tomography scan and computed tomography angiography clearly showed a well-circumscribed, 44x34 mm intrahepatic portal vein aneurysm. Conclusion: The aim of this study is to evaluate the imaging features of portal vein aneurysm.

Keywords: Portal vein, Aneurysm, Liver, Angiography, Intrahepatic
DISCUSSION

Portal vein aneurysms are most often diagnosed sporadically, and the incidence is ambiguous because most patients are asymptomatic. The reported frequency in the ultrasonography is 0.067% [2]. Although the cause of portal vein aneurysm is unknown, two origins, congenital and acquired, have been proposed. Hepatocellular disease and portal hypertension are etiologies of acquired origin portal vein aneurysm. A portal venous system aneurysm may be congenital or may be acquired as a result of weakening of the vascular wall [5]. Congenital factors include an abnormality of the internal walls of the vessel, incomplete regression of the distal right primitive vitellin vein or a variant branching pattern of the portal vein [6]. Portal vein aneurysms may lead urgent surgical operations if they make complications as thrombosis, portal hypertension, rupture, embolism, and compression of the duodenum and inferior vena cava [7]. In this case, the hypothesis of a congenital origin can be suggested because no other cause was found.

Figure 1: Portal vein aneurysm was seen as a well-circumscribed enhanced mass on magnetic resonance imaging scanner.

Figure 2: Portal vein aneurysm. Color Doppler ultrasonography showing color flow in the lesion.

Figure 3: (A) Portal vein aneurysm was seen as a well-circumscribed enhanced mass on computed tomography scan, (B) Aneurysm was seen in axial section of computed tomography scan.

Figure 4: Computed tomography angiography images clearly showing aneurismal dilatation of the umbilical portion of the portal vein.
diagnosis is generally based on color Doppler ultrasound. Color Doppler sonography showed a constant hepatopetal flow along the aneurysmal wall, which immediately led to the diagnosis. We stress the usefulness of color Doppler sonography for studying the hemodynamics of this vascular anomaly and contrast-enhanced dynamic computed tomography and angiography clearly showed a well-circumscribed, 44x34 mm portal vein aneurysm. Large aneurysms may give rise to various complications including right upper quadrant pain, jaundice due to compression of adjacent organs such as the duodenum and the bile duct, rupture, or complete occlusion of the portal vein by thrombosis [3, 4]. Operative treatment is required in these complications. The prognosis depends on complications and underlying liver disease. Therefore, this patient requires careful follow-up.

CONCLUSION

As having complications such as thrombosis, portal hypertension, rupture, embolism, and compression of the duodenum and inferior vena cava, these aneurysms should be appreciated in emergency situations.

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Author Contributions

Muharrem Battal – 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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Alobar holoprosencephaly with unfused thalami: A rare variety of holoprosencephaly

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ABSTRACT

Introduction: Holoprosencephaly with unfused thalami is a rare malformation involving the forebrain and the face. The epidemiology of the disease is poorly known due to paucity of population based studies. Case Report: A 32-year-old grand multipara at 27th week gestation found on routine ultrasound examination to have a single live fetus with the fetal head showing dilated single cerebral ventricle, with no evidence of anterior midline echo (falx, inter hemispheric cistern and septum pellucidum). The thalami appear relatively small but not fused with a thin midline linear echoic septum separating them. Two subsequent sonograms at 30th and 33rd weeks of pregnancy, including coronal sonograms of the fetal head, correctly identified a dilated single cerebral ventricle. There was no history of diabetes mellitus, hypertension or previously affected child. Pregnancy termination was done on the couple’s request, because of the poor fetal prognosis. Postmortem clinical examination revealed a female newborn with normal body structure. The couple declined consent for autopsy. Conclusion: Alobar holoprosencephaly with unfused thalami is a rare and severe variety of holoprosencephaly with poorly understood aetiology and poor prognosis.

Keywords: Alobar holoprosencephaly, Unfused thalami, Pregnancy termination

INTRODUCTION

Holoprosencephaly, the most common malformation of the forebrain in humans, is a structural anomaly of the brain resulting from failed or incomplete forebrain division in the third to fourth weeks of gestation [1, 2]. Its incidence is estimated to be 1 in 16,000 live births and 1 in 250 spontaneous abortions and with a prevalence of 1:250 in embryos [2] and approximately 1:10,000 among live-born infants [3–5]. Holoprosencephaly with unfused thalami is very rare. The epidemiology of the disease is poorly known due to the paucity of population based studies [1, 6]. Environmental, mechanical, and genetic factors have been mentioned as possible causes [1, 6]. Such factors include chromosome aneuploidy, structural abnormality, autosomal recessive and dominant syndromes and maternal diabetes [4]. The role of maternal and paternal ages, parental consanguinity, maternal smoking and drinking habits is controversial [3].

The imaging study of choice in prenatal assessment of holoprosencephaly includes meticulous transvaginal or transabdominal ultrasound and Magnetic resonance imaging (MRI) scan. However, with continued refinement in ultrasonic imaging devices, it is now assuming an increasingly important role in the diagnosis [7, 8].

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Three distinct types of holoprosencephaly have been documented in the literature, which include lobar, semilobar and alobar. The latter is the severest with characteristic intracranial findings of monoventricular cavity, fused thalami, and absence of midline structures such as the corpus callosum and falx cerebri [7]. However, the alobar variety with unfused thalami is rare, which informed this presentation.

CASE REPORT

A 32-year-old gravida 6 para 5+0 female referred at 27th week gestation, for routine antenatal ultrasonography. There were no symptoms or signs suggestive of any clinical condition. The patient had no history of diabetes mellitus, hypertension or previously affected child. General physical examination was unremarkable. The uterus was gravid with symphysiofundal height of 29 cm. There was singleton fetus in cephalic presentation. The fetal heart sound was 110 beats per minute.

Sonographic examination of the gravid uterus revealed a single live fetus. Axial sonograms of the fetal head (Figures 1 and 2) showed dilated single cerebral ventricle, with no evidence of anterior midline echo (falx, inter hemispheric cistern and septum pellucidum). The thalami appear relatively small but not fused; with a thin midline linear echoic septum separating them. It protrudes into the single ventricular cavity. The third ventricle was not visualized. The spine and cranium appeared to be well formed. Biparietal diameter, head circumference, abdominal circumference, and femur length were all consistent with the clinically estimated gestational age of 27 weeks. The interorbital distance was 1.3 cm; no evidence of facial anomalies was noted sonographically (Figure 3). The amniotic fluid index was normal. Two subsequent sonograms at 30 and 33 weeks of pregnancy, including coronal sonograms of the fetal head, correctly identified a dilated single cerebral ventricle.

The couple requested for the termination of pregnancy because of the poor fetal prognosis. This was performed by administration of vaginal prostaglandin (misoprostol). Postmortem clinical examination revealed a female newborn with normal body structure. The couple declined consent for autopsy.

DISCUSSION

Alobar holoprosencephaly with unfused thalami is a very rare congenital malformation of intracranial and
midfacial structures, which may be part of a Smith–Lemli–Opitz syndrome. It may be also an isolated finding or may occur in combination with other extra cephalic defects [8]. Although autopsy and maternal genetic testing were not conducted in the case cited above, prenatal history, detailed family history and focused physical examination of the parents and stillborn to identify microform of holoprosencephaly showed no definitive link to any syndrome or extra cerebral anomalies. The average gestational age at diagnosis is 21.9 weeks (range, 10.5–32.3 weeks) in most literature and in our patient the fetal age is about 27 weeks, which is relatively late, but this can be explained by delay in assessing the hospital by the patient [9].

The spectrum of prenatal cranial ultrasonographic anomalies in the three forms of holoprosencephaly include monoventricle with fused thalamus and corpus striatum, absent corpus callosum, fornix, and falx in alobar form. The features in semilobar include monoventricle with rudimentary occipital horns, falx, and interhemispheric fissure and fused thalamus and basal ganglia. The lobar form is characterized by separated lateral ventricle, absent septum pellucidum; the basal ganglia and thalamus may be fused or separated and the corpus callosum may or may not be present [7, 8, 10]. The prenatal axial fetal head sonogram of the index case showed dilated single cerebral ventricle, with no evidence of anterior midline echo (falx, interhemispheric cistern and septum pellucidum) which is consistent with the alobar holoprosencephaly. However, the thalami appear relatively small but not fused with a thin midline linear echoic septum separating them. This finding has only been reported in a few literatures [10–12].

Greene et al. proposed the use of two criteria for the prenatal sonographic diagnosis of alobar holoprosencephaly [10]. First, the intracranial criterion of a large central fluid collection in the fetal head, with no visible midline structures but with the presence of a mantle around the fluid collection and fusion of the thalami and corpus striatum. Second, sonographic facial abnormalities including hypotelorism, central clefts, facial asymmetry and abnormal orbits [10]. Similarly, Chervenak et al. considered that both hypotelorism and absence of the midline should be observed sonographically to diagnose holoprosencephaly with certainty [11]. The same criteria were used by Parant et al. [12]. In the present case, the first of these criteria was satisfied but no convincing evidence of the second criteria was noted. Demyers and other researchers have also found reasonable percentage of patients with a lobar prosencephaly with a normal face, which is in keeping with our patient [6, 13, 14].

Nerberg et al. had demonstrated that the absence of falx and fusion of the thalami are independent diagnostic features of alobar and semilobar holoprosencephaly irrespective of facial feature [15].

Wengohefer et al. found that exact prenatal assignation of holoprosencephaly into lobar, alobar, and semilobar was inconsistent in 41% of the cases and the ultrasound diagnosis was not confirmed in 19% of the cases [9]. The ongoing improvement of ultrasound equipment is not very likely to improve the ultrasound diagnosis but will allow for a reduction in the number of cases with a wrong assignment to the three groups. Distinguishing the alobar holoprosencephaly from other causes of large intracranial fluid collections such as semilobar holoprosencephaly, ventriculomegaly, hydranencephaly, and large Dandy–Walker cyst is very challenging but may not help in the fetal outcome because all of these malformations have a poor prognosis [9, 15, 16].

**CONCLUSION**

Alobar holoprosencephaly with unfused thalami is a rare and severe variety of holoprosencephaly with poorly understood aetiology and poor prognosis. During routine obstetric sonographic examination, attention should be paid to excluding possible malformations.

**Author Contributions**

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**Guarantor**

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**Conflict of Interest**

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ABSTRACT

Introduction: Capillary hemangioma of testis is extremely rare. Less than 20 cases have been reported and all of these were single lesion. To our knowledge this is the first case report of a multifocal testicular capillary hemangioma.

Case Report: An elderly patient presented with pain in the right testis. Ultrasound showed a mass in the upper pole with increased vascularity at the periphery of lesion. The left testicle and testicular tumor markers were normal. Radical orchidectomy was performed that showed a well-defined cystic area within the testis. Histology showed an organizing hematoma in the cystic area with benign capillary proliferations in the periphery. There were multiple well-demarcated foci of benign capillary proliferations in the adjacent testicular parenchyma. These proliferative lesions were positive for CD34 and factor VIII. This case was diagnosed as multifocal capillary hemangioma. A review of color Doppler imaging of the right testis showed an area of hypervascularity inferior to the mass lesion that corresponded with multifocal capillary hemangiomas on histology. Conclusion: Vascular neoplasms of testis are rare and mainly reported in young adults. Preoperative imaging and frozen sections are vital for appropriate management of these patients. Conservative management or incomplete excision may cause recurrence or hemorrhage but malignant transformation of these lesions has not been reported. This case of multifocal capillary hemangioma in the testis emphasizes the need for an astute radiological examination with frozen section for the appropriate management of patients with a clinically suspicious testicular mass.

Keywords: Testis, Multifocal, Capillary, Hemangioma

INTRODUCTION

Testicular capillary hemangioma is a very rare neoplasm. Less than 20 cases have been reported in literature [1, 2]. All of these were single lesions and most of them were reported in children and younger adults [1–5]. They are often misdiagnosed as malignant tumors clinically. Ultrasound and color Doppler can be inconclusive [3]. Previous case reports have emphasized the importance of frozen section in deciding the appropriate management for these patients [1, 6]. We report a rare case of multifocal capillary hemangioma of the testis in an elderly male that was missed on color Doppler examination and was diagnosed on histology. Awareness of this entity is important so that appropriate management can be given to the patient after an astute
clinico-radiological correlation and frozen section examination.

CASE REPORT

A 74-year-old male presented with pain in the right testis. He had no significant past medical history. Ultrasound of the right scrotum showed a well-demarcated mass with mixed echogenicity in the upper pole measuring 1.91 cm (Figure 1). Color Doppler sonography showed a focus of hypervascularity at the upper pole of lesion (Figure 2). Left testicle showed no mass lesion or areas of increased vascularity. Testicular tumor markers such as alpha-fetoprotein, beta human chorionic gonadotropin and lactate dehydrogenase were within normal limits.

The patient underwent radical orchidectomy in view of a suspicious lesion in the testis. Macroscopically, there was a well-defined cystic area measuring 20 mm, in the upper pole of the testis. The rest of the testicular parenchyma was unremarkable. Whole of the testicular parenchyma was examined and the histology showed an organizing hematoma with granulation tissue in the cystic area (Figure 3). On several deeper levels multiple well-defined foci of benign capillary proliferations ranging from 0.2 mm to 1 mm were seen with intervening atrophic seminiferous tubules inferior to the hemorrhage that confirmed the multifocal nature of the lesion (Figures 3 and 4). Benign endothelial cells lined these capillaries (Figure 5). These endothelial cells were positive for CD34 (Figure 6) and factor VIII and negative for cytokeratin, confirming the vascular nature of the lesion. None of the smaller foci showed hemorrhage. Intervening seminiferous tubules were negative for PLAP and showed no evidence of intratubular germ cell neoplasia. A similar proliferation of benign capillaries was identified adjacent to the organized hematoma which may have represented a remnant of a pre-existing large capillary hemangioma. This case was diagnosed as multifocal

Figure 1: Ultrasound of the right testis showing a mass lesion measuring 1.91 cm in the upper pole.

Figure 2: Color Doppler imaging of right testis. Areas of increased blood flow around the mass lesion (short arrow). Separate area of increased blood flow inferior to the mass lesion measuring 1.07 cm (long arrow).

Figure 3: Hemorrhagic area with cystic changes and granulation tissue (small arrow), benign capillary proliferations inferior to the hemorrhage (long arrow) (H&E stain, x200).

Figure 4: Multiple foci of capillary proliferations (arrows) in right testis with intervening atrophic seminiferous tubules (H&E stain, x400).
capillary hemangioma.

A retrospective review of color Doppler imaging showed an area of increased blood flow inferior to the mass lesion in the right testis measuring 1.07 cm (Figure 2). This corresponded with the area of multiple capillary hemangiomas on histology. This lesion had not been described preoperatively and was not evident on conventional sonography.

DISCUSSION

Capillary hemangiomas are common soft tissue tumors, but they are reported rarely in the testis. They are mainly seen in children and young adults with only two cases reported in males aged more than 70 years old [7, 8]. There are approximately 51 reported cases of vascular hemangiomas of the testis in literature and of these less than 20 cases were unequivocally reported as capillary hemangioma [2]. All of these were reported as solitary lesions on ultrasonography and on histology. There are two case reports in which testicular sparing surgery could be performed in young patients due to frozen section diagnosis [1, 6].

In this case, capillary hemangiomas of testis were seen as separate areas of increased blood flow on preoperative imaging. The sizes of these foci of vascular proliferations ranged from 0.2 mm to 20 mm. This patient presented with pain in the testis due the hemorrhage in the largest focus.

Testicular hemangioma can mimic germ cell or other malignant tumors of testis clinically and radiologically [3]. There have been previous case reports on the radiological findings that will help differentiate between hemangiomas and malignant tumors [9, 10]. Ricci et al. have suggested that an extensive hypervascularity with areas of low resistance velocity on spectral Doppler imaging in a testicular mass should raise the possibility of hemangiomas which should be confirmed on frozen section for appropriate surgical management [10].

In our case, two foci of hypervascularity were seen on color Doppler imaging. Although a frozen section of these areas may have helped in diagnosis of a benign vascular lesion, enucleation or testicular sparing surgery may have been inadequate in this patient due to the presence of multiple foci of hemangiomas in the parenchyma. Incomplete excision of these lesions may result in possible recurrence [1], however no unequivocal malignant transformation has been reported [1, 3]. If managed conservatively or incompletely excised, they can present as hemorrhage and pain in the residual lesion.

CONCLUSION

This is the first case of multifocal capillary hemangioma of the testis in an elderly male and emphasize the need for recognition and awareness of this entity by radiologists, clinicians and pathologists. Testicular vascular tumors are rare and lack a definite preoperative tool that can help differentiate between these benign tumors from malignancy. Therefore in a patient with a testicular mass and normal tumor marker, a color Doppler ultrasound correlation with frozen section can help manage the patient appropriately.

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Shikha Singhal – Substantial contributions to conception and design, Acquisition of data, Analysis
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Unusual imaging of pancreatic metastasis: A case report of tumor-to-tumor metastasis

Rossella Graziani, Paola Spaggiari, Silvia Carrara, Giovanna Lo Bue, Alessandro Zerbi, Luca Balzarini

ABSTRACT

Introduction: Metastasis of one tumor to another tumor is a very rare and controversial phenomenon. Solitary renal cell carcinoma metastasis to a preexisting pancreatic endocrine tumor is distinctly uncommon. We report atypical imaging findings of pancreatic metastasis from renal cell carcinoma, due to tumor-to-tumor metastasis for presence of renal cell carcinoma metastasizing to a pancreatic endocrine tumor. Case Report: A 78-year-old male suffering from mild anemia underwent to multidetector computed tomography scan showing renal cell carcinoma and solid-cystic pancreatic mass, both resectable, treated with right radical nephrectomy and spleno-distal pancreatectomy. Histopathology of the resected renal and pancreatic specimens confirmed a clear cells right renal cell carcinoma metastatic to endocrine neoplasm of pancreatic body-tail. We compared multidetector computed tomography scan findings and histopathological pancreatic specimen. The imaging finding of peripheral rim enhancement coincided in pancreatic pathologic specimen with presence of pancreatic endocrine tumor. The imaging finding of solid trabeculae inside the mass corresponded in pancreatic pathologic specimen to presence of pancreatic endocrine tumor mixed with lobules of typical renal carcinoma metastatic cells. Finally, the imaging finding of hypoenhancing central area of lesion coincided in pancreatic pathologic specimen with presence of large necrotic component. Conclusion: We describe an unusual multidetector computed tomography scan finding of renal cell carcinoma metastasizing to pancreatic endocrine tumor and emphasize the knowledge of rare phenomena of tumor-to-tumor metastasis.

Keywords: Tumor-to-tumor metastasis, Pancreatic Endocrine Tumor, Renal cell carcinoma, Pancreatic metastasis

INTRODUCTION

Renal cell carcinoma (RCC) metastases to the pancreas is especially rare. Their incidence in autopsy series has been reported as 1–3% in patients with primary RCC and their diagnosis is often radiological [1]. Early detection of RCC pancreatic metastases, frequently performed by multidetector computed tomography (MDCT) due to the imaging pattern of hyperenhancing lesions allows for appropriate treatment and improved outcomes for metastatic disease [2–4].

Metastasis of one tumor to another tumor is a very rare phenomenon in which one tumor metastasizes into another tumor [5].
The aim of this case report is to describe atypical MDCT picture of pancreatic metastasis from RCC due to pancreatic endocrine tumor (PET) metastasized by renal cell carcinoma. We emphasize the knowledge of this rare phenomenon in order to avoid an incorrect imaging diagnosis and to planning a relevant treatment.

CASE REPORT

Asymptomatic 78-year-old male, non-smoker and non-drinker, with unremarkable past surgical history, was admitted to our hospital for occasional finding at check-up laboratory tests of persistent iron deficiency mild anemia during the last six months and ultrasound detection of pancreatic and renal masses. The physical examination was noncontributory.

Laboratory investigations on admission showed a normal white blood cells count 10.0x10^9/L (reference range: 4.0–11.0x10^9/L) and a reduced serum level of hemoglobin 11.8 g/dL (reference range: 12–16 g/dL). His serum levels of lipase, amylase, CA19-9, liver enzymes and renal function tests were within the normal range. Moderate increased blood level of endocrine tumor markers was present with Chromogranin A of 115.87 U/L (reference range: 19–98 U/L) and NSE of 14.73 ng/mL (normal value inferior to 12.5 ng/mL).

A 64-slice MDCT scan examination with quadriphasic study (pre-contrast enhanced, contrast enhanced pancreatic, venous and delayed phases) was performed. A contrast enhanced MDCT scan showed focal enlargement of pancreatic body due to the presence of large solid-cystic mass (Figure 1), well-delimited but not encapsulated, measuring 80 mm in maximum diameter, mainly solid with some irregular, large, low-density central areas, suggesting presence of necrosis, and solid trabeculae inside central areas, hypoenhancing before contrast medium administration (Figure 1A). A thin peripheral rim enhancement was present during the pancreatic phase of contrast enhanced MDCT study (Figure 1D), showing wash-out in venous (Figure 1E) and late (Figure 1F) phases. The central areas and solid trabeculae of this mass remained hypoenhanced (Figure 1E–F) during all phases of MDCT study. The dilatation of upstream main pancreatic duct associated to parenchyma atrophy (Figure 1C) was present. There was no evidence of local invasion and peripancreatic vessels were preserved.

Figure 1(A–F): Multidetector computed tomography imaging. Pre-contrast (A) and contrast-enhanced pancreatic (A–D), venous (E) and late (F) phases. Axial images. Multi-detector computed tomography examination showed a pancreatic body well-delimited mass (A, C–F) with maximum diameter of 80 mm, a peripheral rim enhancement (arrow), irregular low-density central areas and solid trabeculae (arrowhead), both hypoenhanced compared to the unaffected parenchyma of pancreatic tail (B) in pancreatic phase (C–D). During the portal venous (E) and delayed (F) phases, the mass remained hypodense and peripheral rim of enhancement showed wash-out. The upstream main pancreatic duct was dilated (C). Perinodular solid masses (short arrow) bulging from the upper pole of right kidney with maximum diameter of 70 mm, homogeneously hyperenhancing during pancreatic phase (B–D) with wash-out during venous (E) and late (F) phases of examination were visible. There was no evidence of local peripancreatic and perirenal invasion, liver metastasis. Superior mesenteric, splenic vessels, portal vein, and renal vessels were preserved.
A multinodular solid mass bulging from the upper pole of right kidney with maximum diameter of 60 mm was visible (Figure 1). This renal lesion was homogeneously hyperenhancing during pancreatic phase of examination (Figure 1B-D), showing wash-out during venous (Figure 1E) and late (Figure 1F) phases of MDCT study. Extra-renal involvement was absent and right renal vessels were preserved.

There was no evidence of abdominal lymphadenopathy, free fluid or metastatic lesions in the liver and in left kidney. Computed tomography scan of the chest was normal.

Endoscopic ultrasound (EUS) confirmed the presence of a well-demarcated solid-cystic mass of pancreatic body, hypoechoic with central fluid and hyperechoic areas, indicating intratumoral necrosis or hemorrhage (Figure 2A). Fine-needle aspiration biopsy endoscopic ultrasound guided (EUS-FNAB) of pancreatic lesion, performed using a 22-gauge needle (Figure 2B) revealed presence of malignant cells. Fine-needle aspiration ultrasound guided of one lesion bulging from the right renal upper pole revealed presence of malignant cells of RCC.

The imaging findings were suggestive for presence of right kidney RCC associated with primary malignant lesion of pancreatic body without usual imaging pattern of pancreatic metastasis form renal cancer. All renal and pancreatic lesions were resectable. The patients underwent right radical nephrectomy and distal pancreatectomy. On gross pathologic examination, in the pancreatic specimen of resected body-tail, a well-circumscribed red and yellow variegated lesion measuring 6 cm in the greatest dimension was present. The lymph nodes identified separately were free of cancer, and the spleen was unremarkable.

Histology of the pancreatic lesion showed two different cells population. In peripheral portion of the mass (Figure 3A) pancreatic endocrine tumor (PET) cells were exclusively observed, infiltrating even solid trabeculae inside pancreatic lesion, mixed with lobules of RCC composed of clear cells (Figure 3B). Large areas of necrosis were found in central portion of the mass, separated by solid trabeculae (Figure 3C). Immunohistochemistry confirmed the histologic picture: CD10 immunoperoxidase showed staining of RCC with no uptake of stain by PEN and synaptophysin immunoperoxidase of an adjacent section demonstrated of PEN only (Figure 3D), which was also positive at Chromogranin A and CD56 staining. PET resulted NET G1 according to WHO classification (2010), with Ki 67 of 1% and pT2N0 according TNM stage. This histological picture can be entirely consistent with a pancreatic endocrine neoplasm surrounding a well-defined nodule of metastatic renal cell carcinoma.

On gross pathologic examination, in the renal specimen of resected right kidney a multinodular, circumscribed and exophytic lesion measuring 7 cm in the greatest dimension was present in the upper pole. Histopathology of the resected renal specimen confirmed a renal cell carcinoma of the kidney, composed mainly of clear cells. The TNM stage was pT3b, pNx, pM1.

Adjuvant therapy were recommended after surgery but the patient declined.

Follow-up with physical examination, laboratory tests, thoracic and abdominal MDCT scan were done every six months.

The patient remain without evidence of disease 12 months from the original diagnosis.
DISCUSSION

Metastasis of one tumor to another tumor is a very rare phenomenon. The criteria for satisfying a true tumor-to-tumor metastasis are as follows [5, 6]:

1. more than one tumor must exist
2. the recipient tumor is a true neoplasm
3. the metastatic neoplasm is a true metastasis with established growth in the host tumor and not the result of contiguous growth
4. tumors that have metastasized to the lymphatic system where lymphoreticular malignant tumors already exist are excluded.

This case report showed two distinct neoplasms and histologic evidence of encasement of an RCC by a PET. The comprehensive criteria that must be fulfilled for the diagnosis of a true tumor-to-tumor metastasis were present in our patient.

Several authors have reported in literature lung cancer is the most common donor tumor, whereas RCC is the most common recipient [5–9]. The reason for tumor-to-tumor metastasis favoring specific tumors is still unknown. The RCC’s rich vascularity, high content of glycogen and lipid, tendency to be localized without infiltration or metastasis could explain its favorable environment for receiving metastases from other cancers [6, 7, 9].

A solitary RCC metastasis to a preexisting pancreatic endocrine tumor (PET) is very uncommon. It is known in literature that PET are frequently hypervascular neoplasms.

Matsukuma investigating 47 autopsy cases of lung cancer concomitant with other tumors found tumor-to-tumor metastasis in only one pancreatic endocrine microadenoma [7].

Cenkowski first described one case of RCC metastasizing to a preexisting PET, reporting MDCT and histopathologic findings [10].

In both cases, reported by Cenkowski and in our patient, MDCT findings of pancreatic lesion due to tumor-to-tumor metastasis from RCC are different from MDCT typical picture of pancreatic metastasis from RCC, which appears as enhancing lesions [1, 2], reflecting hypervascularity of the of primary tumor.

We found a well-delimited solid-cystic mass, with MDCT peripheral rim enhancement, low-density central areas and solid trabeculae, both hypoenhancing after contrast medium administration during pancreatic phase of MDCT study.

We assessed MDCT imaging findings and histopathological pancreatic specimen, comparing them. We have found that MDCT imaging finding of peripheral rim enhancement coincided in pancreatic pathologic specimen with presence of pancreatic endocrine tumor. The imaging finding of solid trabeculae inside the mass corresponded in pathologic specimen to the presence of pancreatic endocrine tumor mixed with lobules of typical renal carcinoma metastatic cells. Finally, the MDCT finding of hypoenhancing central area of lesion coincided in pathologic specimen with presence of large necrotic component.

Early detection of metastases to the pancreas allows for appropriate treatment and improved outcomes of disease. In patients with pancreatic metastases from RCC, absence of extrapancreatic metastases and limited vascular involvement, 2 and 5 years survival rates of 78% and 65%, respectively after resection of pancreatic disease are reported [3, 4]. In our patients, radical nephrectomy and distal pancreatectomy were performed without complications. Clinical, laboratory, imaging follow-up after one year are all negative. However, the true prognosis of tumor-to-tumor metastasis remains unknown because this phenomenon is rare and most of the articles in literature about this disease are sporadic cases reports.

CONCLUSION

A knowledge of rare phenomenon of RCC metastasizing to a preexisting pancreatic endocrine tumor is useful to avoid an incorrect diagnosis in the presence of unusual imaging findings in the pancreatic metastasis from renal cancer. On the basis of these reports, the mechanisms for RCC to pancreatic endocrine tumor specific metastasis, as well as correct treatment and prognosis of this rare disease may be elucidated in the future.

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Author Contributions

Rossella Graziani – 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
Paola Spaggiari – Analysis and interpretation of data, 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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ABSTRACT

Introduction: Systemic lupus erythematosus (SLE) is an autoimmune disorder involving multiple organs, predominantly seen in women of child-bearing age. Erythroderma is described in patients with subacute cutaneous lupus erythematosus (SCLE) but is rare in SLE. Among the cardiac manifestations, pericarditis is common but myocarditis is rare. We report a case of severe SLE presenting with erythroderma, pancytopenia, arthralgia and myocarditis.

Case Report: A 67-year-old male with a background history of hypertension, transient ischemic attack and polymyalgia rheumatica presented with severe erythroderma, malaise, arthralgia, weight loss and was found to be pancytopenic. His antinuclear antibody (ANA) was positive and double stranded DNA (dsDNA) was more than 200 IU/mL with very low complement levels. He developed lupus associated myocarditis with moderately impaired global left ventricular systolic function. He was initially started on steroids and hydroxychloroquine. But as he developed steroid induced myopathy, it was gradually tapered off and mycophenolate mofetil was started. He responded well to the treatment. He satisfied 8 of the 17 systemic lupus international collaborating clinics (SLICC) criteria establishing a diagnosis of systemic lupus erythematosus (SLE). His systemic lupus erythematosus disease activity index (SLEDAI) score came down from 17 to 2 over a period of six months of follow-up.

Conclusion: Systemic lupus erythematosus may present with erythroderma. Careful clinical examination is important in all potentially multi-system diseases.

Keywords: Erythroderma, Systemic lupus erythematosus, Myocarditis, Pancytopenia, Arthralgia

INTRODUCTION

Systemic lupus erythematosus (SLE) is an autoimmune disease involving multiple organs. Ninety percent of patients are women of child-bearing age although both sexes, all ages and all ethnic groups may be affected. It is characterized by immunological abnormalities with production of number of antinuclear antibodies. Erythroderma is rarely associated with SLE, though described with subacute cutaneous lupus erythematosus (SCLE) [1–3]. Premature atherosclerosis is a significant cause of morbidity and mortality in SLE [4]. Pericarditis is a frequent manifestation, whereas myocarditis and Libman-Sacks endocarditis are less common but serious manifestations of SLE [5]. We report a case of severe SLE...
presenting with erythroderma, pancytopenia, arthralgia and myocarditis.

CASE REPORT

A 67-year-old male was presented with symptoms of malaise for two weeks, a generalized rash for one week and joint pain involving the ankles and knees. He also reported weight loss of 5 kg over last six months. His past medical history included hypertension, transient ischemic attack and polymyalgia rheumatica for 16 years on continuous steroids. He was a smoker but did not drink alcohol. There was no significant family history. On clinical examination, he had widespread erythematous rash which initially started on the back and then involved trunk and limbs sparing hands, feet and mucosa. The rashes were photosensitive with a clear demarcation between exposed and covered skin (Figures 1–4). He complained of pain in the small joints of hands, both knees and left ankle. There was no lymphadenopathy and chest, heart and abdominal examination were unremarkable at the initial presentation. But later on he developed third heart sound on cardiac auscultation and an echocardiogram was requested. Although initially apyrexial, he developed high fever on second day of admission.

Investigations and Differential Diagnosis

Full blood count revealed hemoglobin of 11.9 g/dL with a mean corpuscular volume (MCV) of 91 fl, white blood cell count (WBC) 3.3x10^9/L (neutrophil 2.5x10^9/L) and platelets 48x10^9/L. Blood glucose, urea and electrolytes, and liver function test were normal on admission. C-reactive protein (CRP) was 4 μg/mL on admission. He continued to spike temperature intermittently during the admission with no clear source of infection evident on clinical examination. CRP climbed to 216 μg/mL on seventh day. Four sets of blood culture were sent at different times which were all negative. Urine culture was also negative. On sixth day of admission, he became neutropenic with WBC dropping to 1.4x10^9/L (neutrophil 0.8x10^9/L) and he was started on piperacillin and tazobactam. Hemoglobin dropped to 9.9 g/dL on 15th day of admission. There was no history of blood loss. Direct Coombs test was mildly positive with presence of IgG and absence of C3 on the red cells. Haptoglobin and hemopexin levels were normal. LDH was normal at 548 IU/L. Bone
marrow aspirate showed trilineage dysplastic changes thought to represent myelodysplastic changes (Figure 5). Bone marrow trephine biopsy revealed presence of trilineage hematopoiesis which was disorderly, but no evidence of any abnormal infiltrate. Skin biopsy showed prominent exocytosis of lymphocytes with keratinocyte necrosis present together with papillary dermal patchy lymphocytic infiltrate. Focal colloid body formation was present at the dermoeidermal interface but no lichenoid infiltrate was present. These findings were in keeping with graft versus host disease but not typical of an acute phase. The features while not classical also warrant exclusion of lupus erythematosus and dermatomyositis. Serum protein electrophoresis and urine for Bence Jones protein were negative. Urine protein/creatinine ratio was 58.5 mg/mmol (normal range <45 mg/mmol). Serum creatinine was 66 μmol/L with an eGFR of >90 mL/min. Hepatitis B&C and HIV serology were negative. ANA was positive with a titre of >1:2560 with a homogeneous pattern. Anti-dsDNA was more than 200 IU/mL (normal range 10–20 IU/mL). Anticardiolipin antibodies and antibodies to extractable nuclear antigen (ENA) were negative. There was severe hypocomplementia with C3 level of 0.24g/L and C4 level of 0.02g/L. Computed tomography (CT) scan of chest, abdomen and pelvis showed two hemangiomas in liver, gallstones and indeterminate subpleural nodule in the lingular segment of the left lung.

A diagnosis of SLE with erythroderma was made. His disease activity score (SLEDAI) was 17 initially [6]. Systemic steroids (prednisolone 70 mg/day) and hydroxychloroquine (400 mg OD) were started. Skin lesions started improving, white blood cell and platelets gradually normalized. Routine examination during his stay in the hospital revealed third heart sound with no signs of heart failure. Echocardiogram showed globally impaired moderate left ventricular systolic dysfunction with an ejection fraction 40% and he was started on ACE inhibitors and beta blockers. Troponin I was raised at 0.07 (normal range <0.04). These findings were suggestive of myocarditis. He developed pain and weakness of the proximal muscles of both legs within seven days of starting steroids, so his steroid dose was reduced to 40 mg/day and then gradually tapered and was started on mycophenolate mofetil (500 mg BD). His creatinine kinase (CK) was normal at 24 IU/L. Magnetic resonance imaging (MRI) scan of both thighs was done. It showed atrophy of the flexors and the adductors of both hip joints but there was no focal areas of inflammation to guide biopsy. Electromyography (EMG) was normal. Acetylcholine receptor antibodies were negative. Within a week of reducing the dose of steroids his muscle pain improved. He continued to improve while being followed-up as out-patient. The dose of mycophenolate mofetil was increased to 500 mg TDS after two weeks. His exercise tolerance significantly improved, some mild residual erythema was present on the back at second month of follow-up but cleared at sixth month. His repeat echocardiogram at sixth month showed improvement of left ventricular function with an ejection fraction of 48%. At sixth month, he was on 15 mg of prednisolone.

DISCUSSION

The diagnosis of SLE in this case is based on the patient fulfilling 8 out of the 17 SLICC criteria [7]. There are case reports of subacute cutaneous lupus erythematosus (SCLE) presenting with erythroderma, not SLE. Erythroderma is a severe life-threatening condition that presents with diffuse erythema and scaling involving all or most of the skin surface area. A wide range of cutaneous or systemic diseases can present with erythroderma. The most common causes of erythroderma are exacerbation of psoriasis or atopic dermatitis [8]. It can also be secondary to drugs most commonly penicillins, sulfonamides, carbamazepine, phenytoin and allopurinol. Uncommon causes of erythroderma include cutaneous T cell lymphoma and other hematologic and systemic malignancies, connective tissue diseases and infections. Among the connective tissue diseases, subacute cutaneous lupus erythematosus and dermatomyositis have been described to be associated with erythroderma. Patients with SLE frequently develop abnormalities in one or more of the three blood cell lines. In this case, the pancytopenia gradually responded with the treatment. His Direct Coombs test was mildly positive with other markers of hemolysis being normal suggestive of mild hemolytic anemia. Due to the timing of onset of muscle pain and weakness in the proximal muscles of legs after glucocorticoid exposure, he likely had steroid induced myopathy. Muscle enzymes and EMG are usually normal in glucocorticoid myopathy. There are a variety of cardiac manifestations of SLE. Pericarditis is relatively common, but myocarditis is uncommon. Verrucous (Libman-Sacks) endocarditis is usually clinically silent, but it can produce valvular insufficiency and can serve as a source of emboli. In this case, the
myocarditis was severe enough to cause moderate global systolic dysfunction that produced no symptoms, but came to light by vigilant cardiovascular examination. Antimalarials such as hydroxychloroquine is helpful in patients with skin and musculoskeletal involvement. They have also shown to prevent damage to kidneys and central nervous system [9]. Systemic glucocorticoids are usually reserved for patients with organ involvement. In this case, it was considered because of the hematological and severe skin involvement. Immunosuppressive agents like methotrexate, azathioprine, cyclophosphamide and mycophenolate mofetil are considered in patients who are unresponsive or intolerant to steroids. Mycophenolate mofetil was added in this patient because of the side effects of steroids. In patients unresponsive to well-establish treatment, biologic agents like rituximab or belimumab may be considered.

CONCLUSION

We describe a very severe case of systemic lupus erythematosus initially presenting as erythroderma and later on developed pancytopenia and myocarditis. The diagnosis was based on features fulfilling the systemic lupus international collaborating clinics (SLICC) criteria. Though rare, systemic lupus erythematosus may present with erythroderma. Careful clinical examination is important in all potentially multi-system diseases.

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Kingsuk Mukherji – 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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Adrenal cortical carcinoma presenting with secondary amyloidosis: A case report

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ABSTRACT

Introduction: Adrenal cortical carcinoma is a rare aggressive tumor with a poor prognosis. Associated systemic amyloidosis is a rare phenomenon with only single case reported in English literature. Case Report: We report a case of a 50-year-old female harboring a huge adrenal cortical tumor along with systemic amyloidosis who underwent primary curative surgical resection of the tumor. Our patient had a Weiss score of 6 which is indicative of bad prognosis. Conclusion: The presence of proteinuria in patients with adrenal cortical carcinoma may be indicative of amyloidosis. Further, the high risk of metastasis in adrenal cortical cancer mandates a close clinical follow-up.

Keywords: Adrenal cortical carcinoma, Amyloidosis, Cancer, Histopathology

INTRODUCTION

Adrenal cortical carcinoma (ACC) is a rare aggressive endocrine malignancy accounting for 0.2% of all cancers [1]. An ACC has a bimodal age distribution with a first peak in childhood and a second higher peak in the fourth and fifth decades of life [2]. Females are more often affected than males. Majority of cases are sporadic while some show a familial predisposition. Certain malignancies are associated with systemic deposition of amyloid, peculiarly the myelomatosis, Hodgkin's lymphoma and renal cell carcinoma [3]. Systemic amyloidosis in association with ACC is exceptional as in the indexed case and only one such case has been reported in English literature [4].

CASE REPORT

A 50-year-old female presented to the surgery outpatient clinic with a history of weight loss, fever, fatigue and vague pain abdomen for one year. Her blood pressure (BP) records showed fluctuant BP with lowest of 110/70 mmHg and highest of 160/134 mmHg. The patient did not have any history of bone pains, tuberculosis, rheumatoid arthritis or any other chronic ailment. Complete hemogram showed hemoglobin 10.8 g/dL, leukocyte count 4100/μL, differential count 68/25/05/02, platelet count 2.2x10^5/μL and ESR 72 mm at 1st h. Urine examination revealed proteinuria (3+). The serum and urine protein electrophoresis were negative for M (monoclonal) band. Urine was negative for Bence Jones proteins. Biochemical parameters were
as follows: sodium 132 mmol/L, potassium 3.9 mmol/L, serum calcium 9.4 mg/dL, urea 34 mg/dL, creatinine 0.8 mg/dL, total serum protein 3.6 g/dL and albumin 1.2 g/dL. Blood glucose, liver function tests and lipid profile were within normal limits.

On examination a lump was felt per abdomen and computed tomography (CT) scan was advised. A large mass 13x12x10 cm in gastro-splenic region, likely from adrenal, was detected on CT scan which had heterogeneous enhancement with areas of calcification and necrosis. It compressed and displaced the pancreas. The left kidney was also displaced posteriorly. The mass was well delineated from the left kidney by a plane of fat. There was no lymphadenopathy/suggestion of metastasis. The radiologic impression was of a malignant tumor with a possible origin from left adrenal with no evidence of metastasis in the abdominal viscera. Fine-needle aspiration cytology (FNAC) and/or histopathological correlation were advised.

The FNAC was done and smears were cellular with loose clusters and singly scattered tumor cells in a necrotic background. The tumor cells were large, polygonal with plasmacytoid appearance at places. Nuclei were pleomorphic, hyperchromatic with coarse chromatin and cytoplasm was moderate to abundant and granular with vacuolization in some tumor cells. Binucleated cells were also seen. Mitotic figures were noted. Immunohistochemistry (IHC) for vimentin, chromogranin, cytokeratin, S-100 and HMB-45 was done on the cell block. Immunopositivity for vimentin was seen while the other markers were negative. The FNAC report of adrenal cortical carcinoma was rendered.

Based on the FNAC report, complete steroid hormone profile (testosterone, dehydroepiandrosterone, androstenedione, aldosterone, estradiol, 11-deoxycortisol, cortisol, pregnenolone and 17-hydroxypregnenolone) of the patient was done which was within normal limits. Radiologic workup was done to rule out metastasis which showed no evidence of metastasis. The FNAC report of adrenal cortical carcinoma was rendered.

A primary curative radical surgery comprising left adrenalectomy, left nephrectomy and splenectomy was done. The tumor was bulky and compressed the kidney, hence nephrectomy was done. Splenectomy was done because the splenic artery and vein were entangled by the tumor. On gross examination, the left adrenal tumor measured 15.2x11.1x10.0 cm and weighed 770 grams. The mass was well separated from the attached kidney by perirenal fat (Figure 1). Externally, the tumor was grey brown, globular and well encapsulated. Cut section showed a variegated appearance with grey brown to grey yellow to grey tan areas. Areas of hemorrhage and necrosis were seen. Normal adrenal tissue was not identified.

Left kidney measured 13.5x6.5x4 cm and weighed 130 grams. Cut section was pale waxy and translucent (Figure 1). Spleen was enlarged measuring 14x10.5x5.2 cm and weighing 380 grams. Cut section was grey tan and firm. Representative sections examined from the adrenal mass showed a well-encapsulated tumor with organoid and lobular pattern (Figure 2A). The tumor cells were large with plasmacytoid appearance at places (Figure 2B) showing considerable nuclear pleomorphism, and atypia with moderate to abundant eosinophilic cytoplasm (Figure 2C). Clear cells (15%) were seen. The mitotic count was 12/50 high power field including atypical mitoses. Areas of hemorrhage and confluent necrosis were seen. Formation of cholesterol clefts and fibrosis was noted. Lymphovascular invasion was seen (Figure 2D). The capsule was devoid of any breach. Nine histological criteria proposed by Weiss were assessed and a Weiss score of 6 was obtained. Noteworthy were foci showing deposition of intratumoral perivascular homogeneous acellular eosinophilic material (Figure 3A). A panel of immunostains was applied for confirmation of the tumor type. Tumor cells were positive for vimentin and negative for S-100, HMB-45, chromogranin and cytokeratin.

Sections from the kidney were free of tumor and showed deposition of homogeneous acellular eosinophilic material in the glomerular tuft, mesangium and the peritubular capillary walls (Figure 3B). Sections from spleen were free of tumor and showed deposition of eosinophilic material in the arteriolar walls with replacement of follicles at places (Figure 3C).

The acellular eosinophilic material deposits seen in tumor per se, kidney and spleen showed congophilia and apple green birefringence on polarization (Figure 3D), thus confirming it to be amyloid. Multiple organ involvement; kidney, spleen and adrenal gland indicated systemic involvement and metastasis.
amyloidosis. Hence rectal, gingival and subcutaneous adipose tissue biopsies were not done. Furthermore, the sections were treated with potassium permanganate and conghophilia was not seen subsequently thus confirming it to be secondary amyloid. The final histopathological diagnosis was of an adrenal cortical carcinoma with amyloid deposition and amyloidosis kidney and spleen.

After surgery the patient was advised for follow-up in the oncology clinic but the patient did not come for follow-up.

Systemic amyloidosis in association with carcinomas is rare and perhaps with adrenal cortical tumors is exceptional. It is mostly accompanied with immunocyte dyscrasias and sometimes malignant neoplasms [5]. Amongst malignancies, Hodgkin’s lymphoma and renal cell carcinoma have a greater predilection for such an association with a reported prevalence of 4% and 3.2%, respectively [2]. Amyloidosis associated with cancers shows hepatic, splenic and renal involvement. The kidney and spleen were involved in our case. The amyloid A (AA) type of protein has a wide distribution in systemic amyloidosis associated with cancer. The serum amyloid A (SAA), the serum precursor of AA type amyloid is produced by the liver after stimulation by a factor released from activated macrophages. Tumor cells are believed to be effective macrophage activators [4, 5]. It is plausible that tumors initiate amyloid formation by such a mechanism.

Adrenal cortical carcinomas can be functional by producing hormones or non-functional. Cushing’s syndrome is most common manifestation of hormone secreting neoplasms [6]. Rarely, feminization and hyperaldosteronism may be seen in isolation. Incidentally, the adrenal mass may be detected on imaging studies done for an unrelated cause and these incidentalomas are commonplace with hormonally inactive tumors [7]. These patients usually present with pain and/or lump abdomen as in the current case in which the tumor was non-functional. Weight loss, fever, fatigue, anorexia, nausea and myalgias are other symptoms [6]. A few patients may present with the metastatic disease.

In this case, the cytologic diagnosis of ACC was based on combination of characteristic cytology, immunostaining, clinical and radiological information. Cytomorphologic features include good cellularity, scattered and loose clusters of large pleomorphic tumor cells and tumoral fragments with traversing capillaries. Tumor cells are mostly plasmacytoid or polygonal, with moderate to abundant finely granular cytoplasm. Nuclei are usually eccentric with coarse hyperchromatic chromatin and conspicuous nucleoli. Multi-nucleation, bizarre nuclei, frequent mitoses including atypical ones and necrosis are seen in high grade tumors [8, 9]. On cytologic grounds alone, it is extremely difficult to differentiate adrenal cortical carcinoma from other adrenal and extra-adrenal tumors. Immunopositivity for vimentin and negative staining for S-100, HMB-45, chromogranin and cytokeratin helped in cytodiagnosis in the present case. On histopathology our case had a Weiss score of 6. A score of 3 or more is a criterion for malignancy [10]. A diligent histomorphologic assessment, along with staging and curative resection is important for prognosticating the disease.
CONCLUSION

We have reported a case of adrenal cortical carcinoma with secondary amyloidosis which is extremely rare. Weiss score of 6 in our patient correlates with a poorer survival. Given the higher risk of metastasis in adrenal cortical cancer a close follow-up and careful clinical assessment is imperative.

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Author Contributions
Reetu Kundu – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published
Ujjawal Khurana – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published
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REFERENCES

CASE REPORT

Gastric outlet obstruction: An unusual complication of feeding jejunostomy

Umesh Jethwani, Nikhil Bansal

ABSTRACT

Introduction: Enteral nutrition is very important part of management of surgical patients. In cases of corrosive esophageal stricture feeding jejunostomy plays important role in providing nutrition, maintenance of gut structure and function. Though commonly performed surgery, various complications (mechanical, infectious, gastrointestinal and metabolic) have been reported. Case Report: We are reporting a rare complication of gastric outlet obstruction developing in postoperative period of feeding jejunostomy done for corrosive oesophageal stricture. Conclusion: High index of suspicion is required to diagnose this condition in a patient of feeding jejunostomy in postoperative period. In such case operative intervention is required to correct the underlying pathology.

Keywords: Gastric outlet obstruction, Feeding jejunostomy, Esophageal stricture

INTRODUCTION

Feeding jejunostomy is a very important route of providing enteral nutrition in cases of corrosive esophageal stricture. Feeding jejunostomy can be made by–open method, percutaneous method or laparoendoscopic assisted method [1]. Various indications-laparotomy patients in whom a complicated postoperative recovery is expected, those with a prolonged fasting period, those in a hyper catabolic state, or those who will subsequently need chemotherapy or radiotherapy [2]. Although it is an excellent method for surgeons for providing nutritional support, maintenance of gut structure and function, various complications have been associated with this surgery [3]. Among these complications (mechanical, infectious, metabolic) gastric outlet obstruction due to kinked feeding tube is very rare and never reported. The purpose of this article is to report this unusual case we have encountered.

CASE REPORT

A 36-year-old male presented to emergency room with progressive abdominal distension and obstipation from nine days. Patient had history of corrosive induced esophageal stricture, for which feeding jejunostomy was done at a peripheral hospital 10 days back. On examination, he was dehydrated, pulse 96/minute, blood pressure 80/60 mmHg. Distension was present in upper abdomen, bowel sounds were absent and succession splash was present. There was also a midline scar of previous surgery with feeding jejunostomy in situ (non-functioning) (Figure 1). Laboratory investigations were within normal limits. Provisional diagnosis of gastric outlet obstruction was made. After resuscitation patient was planned for exploratory laparotomy. On laparotomy, stomach was grossly distended, with kinking at the jejunostomy site (due to tight suturing) leading to closed loop obstruction (Figure 2). Feeding tube was removed.
and site was repaired. New feeding jejunostomy was made 10 cm distal to it. Postoperative period was uneventful, feeding was started on second postoperative day and patient was discharged on seventh postoperative day.

DISCUSSION

Jejunostomy is a very simple surgery for providing nutritional support in patients of esophageal stricture till the definitive surgery is done. It can be done by various methods, e.g., Witzel (Longitudinal/Transverse), Stamm, needle catheter, percutaneous endoscopy and laparoscopy [1].

However, various complications [2–7] have been reported with this procedure which can be categorized as

1. Mechanical—tube dislocation, obstruction or migration of tube
2. Infectious—cutaneous or intraabdominal abscess, peritonitis or aspiration pneumonia
3. Gastrointestinal symptoms—nausea, vomiting, diarrhea, constipation, abdominal distension
4. Metabolic—hyperglycemia, hypokalemia, hypophosphatemia, hypomagnesaemia, electrolyte imbalance.

Percutaneous placed or laparoscopic jejunostomy tube is preferred because it is minimally invasive and unnecessary laparotomy is avoided. Success and complication rates of these procedures compare favorably with those of the corresponding open surgical procedure. A rare complication of gastric outlet obstruction secondary to feeding jejunostomy was detected in our institute. Various factors suspected for this complication are

1. Tight closure at entry of jejunostomy catheter.
2. Kinking due to single anchoring suture taken at parietal wall to fix the tube instead of three stitches.
3. Tunnel formation in Witzel technique leading to closed loop obstruction.

Due to obstruction jejunostomy site there was closed loop obstruction in our case (esophageal stricture at other end) leading to gastric outlet obstruction. Since it is first time reported complication so no specific guidelines are available to treat this entity. We have taken the patient for emergency laparotomy and corrected the kinking at the feeding tube site.

Mostly these types of complications are due to faulty surgical technique of feeding jejunostomy and can be prevented by adhering to the basic principles of surgery.

CONCLUSION

Enteral nutrition by feeding jejunostomy is very important part of management of esophageal stricture till the patient is nutritionally fit for surgery. We are reporting a rare complication of gastric outlet obstruction following feeding jejunostomy. Although rare, high index of suspicion is required to diagnose this condition in a patient of feeding jejunostomy in postoperative period. In such case operative intervention is required to correct the underlying pathology.

*********

Author Contributions

Umesh Jethwani – 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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REFERENCES


Surgical treatment of high-dysplastic developmental spondylolisthesis in a child: A case report

Nalli Ramanathan Uvaraj, Aju Bosco

ABSTRACT

Introduction: High-dysplastic developmental spondylolisthesis (HDDS) is extremely rare, comprising 5% of the total cases of spondylolisthesis. It can remain asymptomatic for a long time and can progress to a more severe grade of olisthesis and spondyloptosis. Opinions regarding the surgical management of high-grade dysplastic spondylolisthesis in children and adolescents still remain conflicting and controversial. Case Report: This case describes a 12-year-old girl with high-grade dysplastic L5-S1 spondylolisthesis with instability pain, managed with uninstrumented in situ circumferential fusion. The patient showed excellent clinical, functional and radiological outcomes at follow-up of 48th month. Since, plain radiographs and conventional computed tomography (CT) scan failed to show convincing anterior intercorporal fusion, we did fusion analysis with a multi-slice helical tomography scan with multiplanar reconstruction. The reconstructed multiplanar images defined a good posterolateral and anterior intercorporal fusion. Conclusion: Uninstrumented in situ circumferential fusion is a safe and effective surgical option in the management of high-grade L5-S1 dysplastic spondylolisthesis in children and adolescents. Multi-slice helical CT scan with multiplanar reconstruction is the modality of choice in the evaluation of interbody fusion and its progression.

Keywords: Spondylolisthesis, Child, High-grade, Uninstrumented in situ circumferential fusion

INTRODUCTION

Congenital (developmental/dysplastic) spondylolisthesis comprises 15% of spondylolisthesis presenting before adulthood of which only 5% are of high-dysplastic (HDDS) type. Dysplastic spondylolisthesis occurs only at the L5-S1 level and usually presents in females [1, 2]. The significant causative association of genetic factors and the presence of a strong familial preponderance has been established. It is attributed primarily to congenital dysplasia of the L5-S1 facet joints. The pars interarticularis may be poorly developed or elongated. It may be associated with spina bifida occulta of L5 and/or the S1 vertebra and lumbosacral segmentation defects [3]. There is rounding of the superior anterior aspect of the sacrum (sacral doming). Classically, the L5 vertebra is trapezoidal in shape and acquires a kyphotic tilt in relation to the vertically oriented sacrum resulting in an increased lumbosacral angle. It can remain asymptomatic for a long...
time and can progress to a more severe grade of olisthesis and spondyloptosis [4]. Patients usually present during adolescence with low back pain and spinal stiffness. They may also present with lumbar hyperlordosis, hamstring shortening, flexed-hip and knee walking and toe gait. Patients do not always present with radicular symptoms due to adaptability of the nerves to compression or tension.

Treatment of high-grade dysplastic spondylolisthesis (Meyerding grades III, IV and V) in a growing child is operative. Opinions regarding the surgical management of high-grade dysplastic spondylolisthesis in children and adolescents still remain controversial. We present a case of high-dysplastic developmental spondylolisthesis (HDDS) treated operatively, with a note on the surgical technique along with a review of literature.

CASE REPORT

A 12-year-old girl presented with pain in the lower back since 12 months radicular pain without any history of radicular pain. Pain affected her activities of daily living. Clinically, she had an exaggerated lumbar lordosis and painful restriction of movements of the lumbar spine. Her neurological examination was remarkably normal.

Plain radiographs revealed a Meyerding grade III anterolisthesis of L5, an elongated intact pars interarticularis, vertically oriented sacrum with doming of the sacral end plate, trapezoidal L5 and lumbosacral kyphosis (Figure 1), consistent with the diagnosis of a HDDS as classified by Marchetti and Bartolozzi [5]. Computed tomography scan revealed dysplasia of the posterior elements at L5, S1 levels and magnetic resonance imaging (MRI) scan showed decreased spinal canal dimensions at the L5-S1 level (Figure 2).

A detailed family history revealed that the girl’s mother was having an untreated L5-S1 dysplastic spondylolisthesis, emphasizing the need for screening the family members of patients with dysplastic spondylolisthesis.

Operative procedure

Taking into consideration various factors, it was decided to manage this case with an uninstrumented circumferential in situ fusion as a staged procedure. In the first stage, through a posterior midline approach, decortication and autografting (posterior iliac crest graft) of the posterior elements of L4, L5, S1 and the transverse processes of L4 and L5 were done. In the second stage, through a left sided retroperitoneal approach (as against a traditionally performed transperitoneal approach) the bodies of L5 and S1 were exposed. Femoral head allograft was placed between the bodies of L5 and S1, to accomplish an anterior intercorporal fusion. No intraoperative complication was noted and the postoperative period was uneventful. Postoperatively, a three-point support brace was used for six months.

Results-Analysis of outcome

Patient reported complete resolution of her symptoms at third month and resumed full activity at sixth month postoperatively. On final follow-up at 48th month, she had pain free range of movements of the spine with some restriction of forward flexion. A comparative analysis of the preoperative radiographs and radiographs at latest follow-up showed a marginal reduction in slip angle, slip percentage and lumbosacral angle (Table 1). Follow-up radiographs showed some evidences of intertransverse fusion from L4 to S1 (Figure 3). Plain radiographs and conventional CT failed to show a convincing anterior intercorporal fusion between L5 and S1 (Figure 4). We therefore did fusion analysis with a multi-slice helical tomography scan with multiplanar image reconstruction. The reconstructed multiplanar images defined a good
posterolateral fusion mass extending from L4 to S1 and solid anterior interbody fusion mass bridging L5 and S1 (Figure 5A–D). Her functional outcomes with regard to pain, postoperative function, level of daily activity, and social life were assessed at 48th month using Oswestry Disability Index (ODI) and Scoliosis Research Society (SRS) scores [6, 7]. The ODI score improved from 37.7% to 58%.

Table 1: Results from radiographic analysis

<table>
<thead>
<tr>
<th>Variable</th>
<th>Preoperative</th>
<th>Postoperative (At 48th month follow-up)</th>
</tr>
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<tbody>
<tr>
<td>Slip percentage</td>
<td>69%</td>
<td>58%</td>
</tr>
<tr>
<td>Slip angle (degrees)</td>
<td>26</td>
<td>20</td>
</tr>
<tr>
<td>Lumbosacral angle (degrees)</td>
<td>18</td>
<td>12</td>
</tr>
<tr>
<td>Lumbar lordosis (degrees)</td>
<td>72</td>
<td>65</td>
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</tbody>
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Figure 2: T2-weighted sagittal magnetic resonance imaging scan of the lumbosacral spine showing degeneration of the L5-S1 disc with severe spinal canal narrowing, anterolisthesis of L5 over S1 with doming of the sacral end plate.

Figure 3: Follow-up plain lateral radiograph of the lumbosacral spine (taken at 14th month postoperatively) showing the posterior fusion mass from L4-S1 and some evidences of anterior interbody fusion.

Figure 4: Computed tomography scan of the lumbosacral spine showing anterior intercorporal fusion between L5 and S1 and a good posterior fusion mass extending from L4 to S1.
slip and of lumbosacral kyphosis. If the patient presents later with radicular symptoms, decompression may be accomplished in the presence of a circumferentially fused stable lumbosacral spine. We, therefore, recommend decompression only when patients present with radicular symptoms.

Though there is no significant reduction of slip in insitu fusion as against instrumented reduction, the ODI and SRS scores were significantly better in the fusion in situ group than in the reduction group [10]. Disc degeneration above the fusion was more common in the reduction group than in the fusion in situ group on long-term MRI follow-up [13].

**DISCUSSION**

The surgical management of high-grade dysplastic spondylolisthesis in children and adolescents still remains conflicting and controversial. The spectrum of available surgical options include, instrumented in situ fusion, instrumented reduction and fusion or uninstrumented in situ fusion which may be posterolateral, anterior or circumferential and vertebrectomy (Gaines procedure).

Though modern surgical techniques and instrumentation permit reduction or even removal of a severely slipped fifth lumbar vertebra, yet the benefits of reduction still remain controversial [8, 9]. Despite modern neurophysiologic monitoring (somatosensory and motor evoked potentials), these instrumented reduction procedures present the possibility of severe neurological complications [10]. According to available literature, instrumented reduction of L5–S1 high-grade spondylolisthesis is associated with an 8–30% rate of postoperative neurological compromise, mostly consisting of nerve root injuries or cauda equina syndrome. There is a relatively high incidence of well-documented complications associated with reduction, including instrumentation failure, loss of reduction and pseudoarthrosis [11]. Though reduction may reduce the slip, restore the sagittal plane balance and normal biomechanics, we believe that the risks of reduction in the setting of pediatric high-grade spondylolisthesis outweigh the benefits.

Despite no reduction in the translational deformity, uninstrumented in situ fusion offers better long-term clinical, radiological and functional outcomes than instrumented reduction has in the same patient population [10]. In situ posterolateral fusion in children with severe spondylolisthesis was followed by deterioration of olisthesis in 25% of the cases [12]. Anterior interbody fusion saves levels, offers better fusion rates, provides axial load sharing and halts slip progression. Furthermore, circumferential in situ fusion offers better long-term results than posterolateral or anterior fusion alone [13].

Despite a severe neural canal and neural foraminal narrowing seen in the MRI images, the patient under study had no radicular symptoms. Hence, a primary decompression was not done in our patient. This is consistent with the fact that patients do not always present with radicular symptoms due to adaptability of the nerves to compression or tension. In addition, decompression in the presence of instability will further weaken the posterior buttress with postoperative progression of slip and of lumbosacral kyphosis. If the patient presents later with radicular symptoms, decompression may be accomplished in the presence of a circumferentially fused stable lumbosacral spine. We, therefore, recommend decompression only when patients present with radicular symptoms.

Though there is no significant reduction of slip in insitu fusion as against instrumented reduction, the ODI and SRS scores were significantly better in the fusion in situ group than in the reduction group [10]. Disc degeneration above the fusion was more common in the reduction group than in the fusion in situ group on long-term MRI follow-up [13]. Therefore, uninstrumented

**Table 2: Functional outcome analysis**

| Variable | Preoperative | Postoperative (at 48th month) 
<table>
<thead>
<tr>
<th></th>
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<tbody>
<tr>
<td>ODI Score</td>
<td>37.7% (moderate disability)</td>
<td>8.1% (mild disability)</td>
</tr>
<tr>
<td>SRS Score</td>
<td>36.0</td>
<td>103.0</td>
</tr>
</tbody>
</table>

Abbreviations: ODI, Oswestry Disability Index; SRS, Scoliosis Research Society

Figure 5: (A, B) Multi-slice helical tomography scan of the lumbosacral spine with reconstructed multiplanar images defining a good posterolateral fusion mass (white arrows) extending from L4 to S1, and (C, D) a solid anterior interbody fusion mass (black arrows) bridging L5 and S1.
in situ circumferential fusion can be considered as a method of choice in the management of high-grade L5-S1 dysplastic spondylolisthesis in children and adolescents.

Although reduction may improve the cosmetic appearance postoperatively, very few patients complain about their cosmetic appearance in the long-term and indeed their appearance is improved by relief of hamstring tightness and correction of sciatic scoliosis. Though reduction was not attempted intraoperatively, the observation of a marginal reduction of slip angle and slip percentage at follow-up may be explained by the fact that corrective remodeling occurs during growth in patients operated on at an early age.

Traditionally, interbody fusion has been assessed with plain radiographs, which have significant intra- and interobserver variation [14]. Sandhu et al. found that although plain radiographs showed evidence of interbody fusion at sixth month, only 33% were subsequently judged fused on histologic examination [15]. When human allograft bone is used for interbody fusion, it is again difficult to interpret fusion progression or allograft resorption, replacement or graft incorporation on plain films. The CT scan provides better evaluation of fusion progression and has evolved as the preferred method of assessing interbody fusion. It offers the potential for high-quality reformatted images in the coronal and sagittal planes, and provides exquisite bone detail [14]. Three dimensional multi-slice thin section CT scans with multiplanar reconstruction have been demonstrated to show fusion maturation and bone growth and have been effective in fusion evaluation unlike with plain CT films [14].

The retroperitoneal approach that has been used to obtain anterior interbody fusion between L5 and S1 offers an alternate safer approach to the commonly used transperitoneal approach. In addition, ethylene oxide sterilized allograft used in this patient, can be used to achieve anterior intercorporal fusion.

CONCLUSION

Uninstrumented in situ circumferential fusion is a safe and effective surgical option in the management of high-grade L5-S1 dysplastic spondylolisthesis in children and adolescents. Multi-slice helical computed tomography scan with multiplanar reconstruction is the modality of choice in the evaluation of interbody fusion and its progression.

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Author Contributions

Nalli Ramanathan Uvaraj – 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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Successful cesarean delivery following prenatal diagnosis of giant sacrococcygeal teratoma in the fetus

Murat Bakacak, Salih Serin, Aykut Urfaloğu, Mehmet Akif Sarıca

ABSTRACT

Introduction: Sacrococcygeal teratomas are the most common congenital neoplasia in the newborn, which are seen on the frontal surface of coccyx and sacrum. As a result of ultrasonographic developments in recent years, sacrococcygeal teratoma can be easily diagnosed in the prenatal period. Case Report: We report a case of a 29-year-old patient with a singleton pregnancy of 38 weeks, with a giant sacrococcygeal teratoma of 20x18 cm in the sacrococcygeal zone. The infant was successfully delivered by classical cesarean section. Conclusion: It is quite important to diagnose sacrococcygeal teratomas in the early weeks of pregnancy and to monitor prenatal management and prenatal complications to be able to determine the mode and place of delivery. When planning delivery, the least traumatic method should be preferred.

Keywords: Sacrococcygeal teratoma, C-section, Delivery, Fetus, Prenatal diagnosis

INTRODUCTION

Teratomas, which are mostly observed in the sacrococcygeal zone in the fetus, are the most common neoplasia in the newborn [1]. The prevalence rate of this disorder is 1: 40,000 live birth. The clinical significance of (SCT) results from its concurrence with other pathologies, thereby increasing prenatal and perinatal mortality and morbidity [2]. Mortality and morbidity rates are relatively high in patients suffering from SCT as a result of high output heart failure, polyhydramnios, hydrops, preterm delivery, anemia, and tumor rupture [3]. SCT can be easily diagnosed in the prenatal period due to ultrasonographic development in recent years [4]. SCT is diagnosed in the early period in the form of a cystic, solid or mixed mass extending from the sacral zone to the perineum or hip [5]. Ultrasonography is valuable in monitoring the size of the tumor, early diagnosis of complications, and in the determination of the most favorable time and method of delivery as well as prenatal diagnosis [4].

In this study, a 29-year-old patient presented with a singleton pregnancy of 38 weeks, with a giant sacrococcygeal teratoma of 20x18 cm in the sacrococcygeal zone and subsequent delivery of the infant by classical cesarean section.

CASE REPORT

A 29-year-old Gravida 2, Parite 1 was referred to our clinic with the pre-diagnosis of intra-uterine fetal tumor.
The pregnancy was calculated as 38 weeks according to the date of her last menstrual period. Antenatal follow-up had not been regular. No abnormality was observed in the medical history of her previous child.

Ultrasonographic examination of the patient showed an intrauterine, single and alive fetus in transverse presentation with biometry compatible with 38 weeks. In the fetus, a heterogeneous mass was observed with a smooth surface, extending from the sacral zone to the perineum, with solid and cystic zones of 20x18 cm, and no intra-abdominal extension (Figure 1). Cesarean delivery under regional anesthesia was planned after the consideration of the complications that might arise during the removal of the fetus.

Combined spinal-epidural anesthesia (CSE) was selected as the cesarean delivery anesthesia. After block application was confirmed, surgery was started.

Taking into account the dimension of the fetal mass, a mid-line incision under the belly was applied to the abdomen, and a longitudinal incision was applied to the uterus in order to decrease the risk of rupturing the tumor. The head of the fetus was turned towards the opening and the fetus was delivered without complications with a mean APGAR score of 9-10. In the examination of the newborn, a mass of approximately 20x22x22 cm with a smooth surface and prominent venous congestion was detected in the fetal perineum (Figure 2). Sacrococcygeal teratoma was considered as a result of the localization and appearance of the mass. In the magnetic resonance imaging (MRI), which was applied in the postnatal period, the mass was observed arising from the sacrococcygeal region in the sagittal imaging and the mass had hypointense solid areas and hyperintense cystic septations on the T2-sequence (Figure 3). The mother was discharged from the hospital on postoperative day-2. The newborn was admitted to the pediatric surgery clinic for an operation.

**Figure 1:** Heterogeneous mass with a smooth surface, extending from the sacral zone to the perineum.

**Figure 2:** View of the mass in the newborn.

**Figure 3:** T2-sequence of the magnetic resonance imaging scan of the mass applied in the postnatal period.
DISCUSSION

Although teratomas, which are the most common congenital tumors, may be observed in different parts of the body, they are generally located in the sacrococcygeal zone at reported rates of approximately 50% [6]. As a result of ultrasonographic developments in recent years, sacrococcygeal teratoma is easily diagnosed in the antenatal period [7, 8]. In ultrasonography examination, a mass in cystic, solid or mixed form including both components, is observed with different echogenicity according to the content and sometimes showing intra-abdominal and pelvic extension, present especially in the sacral zone. In addition, it can also define the relationship between vascular structures and fetal circulation and can differentiate SCT from meningomyelocele by demonstrating the SCT vascular structures in the mass during Doppler examination [5, 9].

Large vascular structures in SCT and the development of arteriovenous shunts increase certain prenatal complications such as high output heart failure, fetal hydrops, polyhydramnios, and cardiomegaly [8].

Diagnosis of SCT in the prenatal period is significant for prenatal management, prenatal follow-up and planning of the delivery method. In vaginal delivery, death may occur as a result of serious dystocia and bleeding originating from a large vascular tumor. Vaginal route delivery was reported in 8 of 10 patients, but it was also noted that cystic tumor aspiration developed in one case [10]. Although vaginal delivery is possible in sacrococcygeal teratomas <5 cm and which do not include additional abnormalities, cesarean delivery is preferred for lesions >5 cm because of dystocia or hemorrhage risks [11]. Chuileannain et al. published two cases in which classical cesarean section procedure was applied to the patients, four cases in which sub-uterine segment cesarean procedure was applied, and one case in which delivery was realized through the vaginal tract, and it was stated that perinatal death was observed only in the case in which vaginal delivery was preferred [12]. On the other hand, Kay et al. reported vaginal deliveries without complication after prenatal percutaneous drainage in two cases with cystic sacrococcygeal teratomas, and percutaneous needle drainage was reported to be a potential alternative to cesarean delivery [13]. As Hoehn et al. stated, cesarean delivery should be preferred in cases with large tumors in order to avoid dystocia, tumor rupture, hemorrhage and traumatic delivery. Nevertheless, it should be remembered that difficulties may be encountered even in cesarean section. It is well-known that extensive hysterotomy applied due to a large teratoma, will increase morbidity [14]. This current case, cesarean section was applied since dystocia might occur due to the sacrococcygeal tumor having a large volume, and a classical uterine incision was preferred to prevent teratoma rupture.

Since SCTs are generally diagnosed via ultrasonography in the prenatal period, intrauterine cyst aspiration, open fetal surgery, radiofrequency ablation method and thermoregulation are alternative options in the treatment. However, there are varying success rates of these operations, and more comprehensive studies are needed on those subjects [11]. Specialist physicians in the fields of gynecology, pediatrics and pediatric surgery should be present in the centers where the deliveries of patients with SCT will be performed in order to create a multi-disciplinary team approach [6].

CONCLUSION

In conclusion, diagnosis of sacrococcygeal teratoma in the early weeks of pregnancy may contribute to prenatal management, follow-up of prenatal complications and determination of the mode and place of delivery. When the delivery is planned, the least traumatic method should be preferred for the delivery.

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Murat Bakacak – 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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Mehmet Akif Sarıca – Acquisition of data, 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.

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Early bilateral hippocampal lesions in transient global amnesia: Evidence against delayed ischemia?

Kelsey Flynn, Pascale Lavoie, Robert Laforce Jr

CASE REPORT

A 65-year-old right-handed female was admitted to neurology after she suddenly began experiencing problems with her memory. Her past medical history was significant for osteoarthritis and a fractured clavicle, but there were no vascular risk factors, head trauma, migraine or epilepsy. She did not smoke, occasionally drank alcohol and denied illicit drug use. Her mother had passed away at 98 years of age without any sign of Alzheimer’s disease but one of her aunts was diagnosed with Alzheimer’s disease. She has four siblings, all were in good health, except for one brother with cardiac problems.

On the day of the event, at 11:15 a.m. she began showing symptoms after a short bout of intense running for almost six minutes. She reported a vague sensation of confusion as to where she was and what she was doing, but nonetheless called her husband to pick her up. When he arrived, he noted her confusion in space and time, and she admitted to feeling dizzy. Upon arrival at their home, her confusion persisted. For example, she had forgotten about her sister’s birthday, and that they were renovating their kitchen. She repeatedly asked her husband “Why is there a present on the counter?”, or “Why is our kitchen such a mess?” After a few hours, she returned to normal level of functioning with no recollection of what had just happened.

Neurological examination was normal. Blood pressure was 122/68 mmHg, heart rate 76 bpm. Basic blood work, ions, urea/creatinine, TSH, calcium/magnesium/phosphorus, B12/folates were unremarkable. She scored 26/30 on a brief cognitive screening measure, losing four points in the free recall section, but showing intact recognition with verbal cues. The rest of her cognitive examination was normal. Magnetic resonance imaging (MRI) scan of the brain conducted 19 hours after symptom onset revealed bilateral lesions of the hippocampi (Figure 1).

DISCUSSION

The patient was diagnosed with transient global amnesia (TGA), a benign syndrome characterized by the sudden onset of severe anterograde and mild retrograde amnesia [1]. Transient global amnesia is generally accompanied by repetitive questioning and resolves within 24 hours, leaving no sequelae. Recurrence is rare. Several possible etiologies have been suggested as underlying mechanisms for TGA including migraine-related mechanisms, venous-flow abnormalities, epileptic seizures, and ischemic events [2–4]. However,
the fundamental etiology of TGA remains unclear and may be highly heterogeneous.

Recent neuroimaging studies have revealed that a large portion of individuals with TGA present with small, reversible, restricted lesions on diffusion MRI [5]. This has been used to support an underlying ischemic event that could be the result of venous overflow. Furthermore, these lesions often tend to appear when patients are scanned after a 48-hour delay, hence supporting the delayed ischemic hypothesis [5]. Interestingly, our patient’s ischemic event was detected in the acute phase (0–24 hours) of TGA where most other authors have failed to report any changes. Whether this is attributable to lesion size is unclear, but nonetheless does not support the notion of delayed ischemia.

We herein report a case of transient global amnesia (TGA) secondary to bilateral ischemic hippocampal lesions 19 hours after symptom onset. In accord with the hypothesized etiology of venous overflow, our findings replicate other studies showing reversible ischemia in hippocampal regions associated with this syndrome. Interestingly, lesions to other structures known to be involved in declarative memory (e.g., thalamus, cingulate gyrus, and basal ganglia) have been associated with TGA [6]. In addition to being rare because of their bilateral nature, the lesions shown here are unique in that very few authors have reported such changes in the acute phase of TGA. Moreover, they raise the possibility that delayed ischemia may not be the sole mechanism behind TGA. Indeed, ischemia may be present early in the process but simply not detected [5].

CONCLUSION

Newly developed imaging techniques such as functional Magnetic resonance imaging scan may allow earlier detection of cerebral changes associated with transient global amnesia. This in turn may help clinicians in the differential diagnosis of other transient amnestic syndromes, as well as more rapid targeted interventions.

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

Pascale Lavoie – Acquisition of data, Analysis and interpretation of data, Critical revision of the article, 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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**REFERENCES**


Gallbladder agenesis diagnosed intraoperatively

Atul Kumar Mittal, Pinakin Patel, Gajendra Anuragi, Bhairu Gurjar, Suresh Singh, Rajgovind Sharma

To the Editor,

Gallbladder agenesis is a rare congenital anomaly with very low incidence of 0.01–0.06% [1]. Female predominance seen in ratio of 3:1 [2]. Symptoms usually present in second or third decade of life [2]. Some patients present with a clinical picture suggestive of gallbladder disease. The inability of ultrasonography abdomen to convincingly diagnose agenesis of the gallbladder the diagnosis is infrequently made preoperatively. This study describes a case of gallbladder agenesis diagnosed on laparoscopy.

A 30-year-old female presented with pain (on and off) right hypochondrium since two years in surgery outdoor with reports of ultrasonography abdomen showing cholelithiasis. Clinical examination revealed a tender right hypochondrium with a positive Murphy's sign. The diagnosis of cholecystitis with cholelithiasis made on further ultrasonography and she was scheduled for laparoscopic cholecystectomy.

Intraoperative findings were:
1. The gallbladder could not be seen even after meticulous search.
2. The procedure converted to open procedure.
3. In open exploration; gallbladder could not be seen, common bile duct was dilated and stones were absent.
4. Postoperative magnetic resonance cholangiopancreatography confirmed the diagnosis of congenital absence of the gallbladder (Figures 1 and 2). Postoperative diagnosis was costochondritis, and managed with topical and oral analgesics and anti-inflammatory drugs.

Gallbladder agenesis can present with an unpleasant surprise to the surgeon intraoperatively [1]. Gallbladder agenesis was first reported by Lemery and Bergman in 1701 and 1702. Agenesis resulted from failure of the gallbladder and cystic duct to bud off from the common bile duct during the fifth week of gestation [3].

Gallbladder agenesis clinically presents with:
(1) Asymptomatic (incidental finding at laparotomy for another reason) (35%)
(2) Symptomatic (50%)
(3) In children with multiple fetal anomalies (such as tetralogy of Fallot and agenesis of the lungs) [1].

Symptomatic patients present with clinical features, similar to those of biliary tract conditions symptomatic group should be managed conservatively with smooth muscle relaxants and if this fails, sphincterotomy is done [4, 5].

Unable to find out the gallbladder at laparoscopy prompts surgeons to open exploration of the biliary tracts frequently, it is of no benefit and adds morbidity of the procedure, like iatrogenic injury to biliary tract. Recent non-invasive imaging techniques such as magnetic resonance cholangiopancreatography and endoscopic ultrasonography provide an excellent alternative to open exploration [6–9].

Gallbladder agenesis is a rare congenital anomaly with very low incidence. Cases in which ultrasound suggests non-visualization of the gallbladder or suspicion of biliary tract anomaly, preoperative magnetic resonance cholangiopancreatography should be considered [7]. It is also helpful in demonstrating an ectopic gallbladder along with other possible anomalies of the biliary tract.
system [8, 9]. Proceed to immediate open exploration should be avoided in order to prevent iatrogenic biliary tree injuries [8].

Figure 1 (A–C): Magnetic resonance cholangiopancreatography films showing dilated common bile duct and agenesis of gallbladder with normal extrahepatic biliary system.

Figure 2 (A, B): Open exploration showing absence of gallbladder.

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