CASE REPORT

Sacrococcygeal teratoma in adults: case report and literature review

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Sacrococcygeal teratoma is one of the most common tumours in infants but rare in adults. We present a case of sacrococcygeal teratoma in a female adult. The clinical presentation, radiological and histological findings, management, and outcome are described.

Introduction

Sacrococcygeal teratoma (SCT) is derived from embryonic germ cell layers. It is one of the commonest tumours in infants, with a prevalence of 1/40 000 births.[1] It is rare in adults however, most being located in the intrapelvic space,[2] and 1 to 12% were reported to undergo malignant transformation.[3] Radiological imaging is helpful in the diagnosis of these lesions and to delineating their extent. A mature SCT is potentially curable by complete surgical resection.[2] We present a case of adult SCT and a literature review of this entity.

Case report

When the patient was 21 years old (16 years prior to her latest admission), she presented to a gynaecology department with episodes of difficulty with micturition; at the time a pouch of Douglas mass was suspected. Ultrasound (US) examination of the pelvis showed a 10 cm x 5 cm x 6 cm lesion with mixed echogenicity on the left side of the pelvis posterior to the uterus. Open exploration revealed a right ovarian cyst, and a right cystectomy was performed; pathological examination confirmed it to be a lutein cyst.

Aged 31 years (10 years later), she presented again to the gynaecology department with episodes of difficulty with micturition and left lower abdominal pain. Gynaecological examination was unremarkable. Transvaginal US showed a 6.6 cm x 6.9 cm x 6.2 cm homogeneous mass in the pouch of Douglas. Laparoscopy was performed but no pelvic mass was found. Subsequently, she defaulted follow-up.

She was readmitted to the surgical department in November 2010 with left buttock swelling and fever. Physical examination confirmed that she was febrile and revealed a cystic lesion in the left buttock. Rectal examination revealed a presacral cystic lesion. At that time the preliminary diagnosis was a peri-anal/peri-rectal abscess. Bedside aspiration was performed and yielded 10 mL of old blood. She underwent urgent US of the buttock and plain computed tomography (CT) of the pelvis. The US revealed an irregular heterogeneous cystic lesion at the left posterior buttock, measuring about 7.4 cm x 8.9 cm x 7.4 cm (Figs 1a and b). The patient's CT scan confirmed the presence of a large irregular mass with fat densities and calcification, posterior to the rectum, and extending to the left ischiorectal fossa. Areas of homogeneous density near to that of water were seen. The lesion was distinct from the uterus, cervix, and vagina. The sacrum and coccyx were intact (Figs 1c and d).

After treatment with antibiotics the fever subsided and the patient was discharged. Magnetic resonance imaging (MRI) of the pelvis 2 weeks later showed a heterogeneous fat containing mass with cystic components in the retrorectal space extending to the left ischiorectal fossa. The mass was largely below the pelvic floor. Mixed T1 hypointense (in the superior aspect) and hyperintense components (more in the inferior aspect) were noted in the lesion. Moreover, T2 hyperintense signal components were noted, as were small areas of intralesional gadolinium enhancement. There was no evidence of invasion of the rectal or bladder wall, pelvic floor muscles or the sacrum and coccyx, and there was no perianal fistula. These features were all consistent with SCT (Figs 1e and f).

Serum alpha-fetoprotein (AFP) and human chorionic gonadotrophin (HCG) levels were normal. Transperineal excision of the SCT was performed. Intra-operatively, a 15 cm x 10 cm multilobulated cystic tumour with hair and sebum was encountered below the pelvic peritoneum. It was located between the coccyx and the rectum and more on the left side (Figs 2a and b), with dense adhesions around it. Laterally, the tumour reached

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成人骶尾部畸胎瘤病例報告和文獻回顧

骶尾部畸胎瘤是嬰孩最常見腫瘤的其中一種，很少在成年人身上發生。本文報告一名骶尾部畸胎瘤的成年女性患者，並描述這病的臨床表現、放射及組織學結果，以及醫治方法和結果。

the left ischial tuberosity and extended into the ischiorectal fossa over the right side. As the tumour was densely adherent to the coccyx and Waldeyer's fascia, it was considered to be a SCT, rather than its counterpart—a presacral teratoma that has no apparent tissue connection to the coccyx.

Histopathological examination confirmed the diagnosis of a benign mature teratoma (Figs 2c and d). Microscopy revealed the presence of various types of mature and differentiated tissues derived from all three germ layers. Endodermal origin tissue consisted of gastric mucosa, small intestinal mucosa, and respiratory type epithelium. Mesodermally derived tissues included: bone, cartilage, mature adipose tissue and skeletal muscle. Sebaceous glands and hair follicles formed the ectodermal component. There was no evidence of immature cell types or malignant transformation.

The patient's postoperative course was uneventful and she was discharged on postoperative day 6.

Discussion

Sacrococcygeal teratoma is a neoplasm arising in the sacrococcygeal region and contains tissue derived from more than one primitive germ layer.[4] Its cause remains unknown.[2,5] There were suggestions that it originated from multipotential cells in Henson's node, which migrates caudally to the coccyx. It is one of the commonest fetal neoplasms, but is rare in adults.[3,6] Review of the literature reveals case reports or case series only and there is a lack of information on its incidence in the Chinese population. Most adult SCTs are intrapelvic, whereas most are external in infants. In infants the liability to malignant

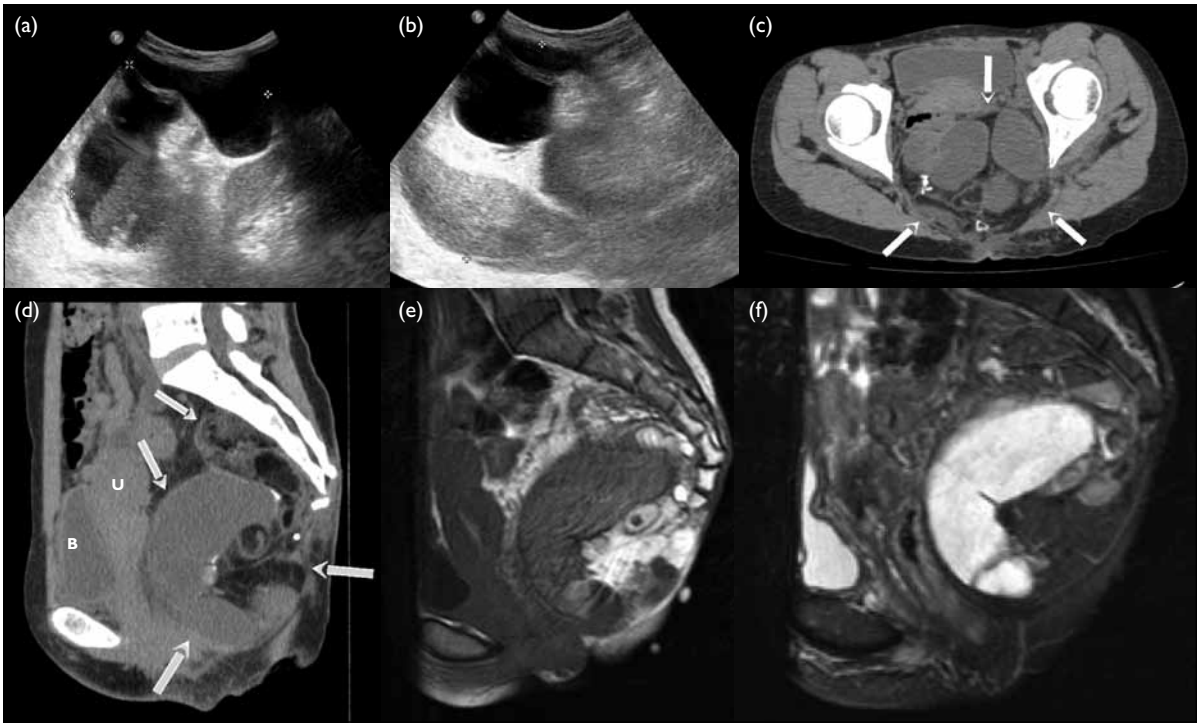

FIG 1. Ultrasound images of the left buttock showing (a) a large complex mass with cystic and soft tissue components, and (b) a heterogeneous lesion at the left buttock with cystic components (top layer), fat (middle echogenic layer) and soft tissue components (bottom layer). (c) Axial computed tomography (CT) pelvis obtained at the level of the acetabulum showing a heterogeneous mass (arrows) with cystic components in the retrorectal space extending into the left ischiorectal fossa. Fat, soft tissue, and calcification were noted within the lesion. Displacement of the rectum anteriorly and to the right was evident. (d) Midline sagittal CT pelvis showing an irregular mass lesion (arrows) anterior to the sacrum and coccyx extending inferiorly. It appeared to displace the vagina anteriorly and compressed the urinary bladder (U: uterus, B: urinary bladder). (e) Midline sagittal T1 magnetic resonance (MR) image of the pelvis and (f) midline sagittal fat saturated T2 MR image of the pelvis showing a heterogeneous mass occupying the presacral space, that seemed to displace the rectum anteriorly. Cystic areas with homogeneous contents were seen. The anterior cystic components showed T1 hypointense signals and T2 hyperintense signals, indicating serous fluid. T1 hyperintense areas, which were suppressed on the fat saturated sequence, were seen at the posterior aspect of the lesion, indicating fat content

transformation is much higher.[4] Most adult SCTs are cystic, and 1 to 2% are malignant.[7] Sacrococcygeal teratoma occurs more frequently in females; the female-to-male ratio is about 10:1.[1,7] One postulate is that this may be related to the relatively delayed differentiation of the ovaries compared to the testes.[4]

The clinical presentation of SCT depends on its location and extension. Incidental features have been reported during routine clinical assessment.[8] Symptoms may be related to mass effects of the tumour, such as low back pain, bowel or urinary symptoms, and venous engorgement of the lower limbs.[1,7,9] The episodes of difficulty with micturition experienced by our patient may have been related to the mass effect of the tumour. Patients may also present with an abscess or a perirectal fistula.[10] Extrinsic compression of the vagina, as well as displacement of the uterus and rectum, may also be detected on pelvic and rectal examination.

Since SCT has the potential for malignant transformation which increases with age,[4] early detection and management are important. Radiography of the pelvis is helpful by revealing calcification within the tumour and enables assessing the integrity of the sacrum and coccyx.[4] More than 50% of SCTs exhibit calcification or ossification.[11] Anterior displacement of the rectum may be revealed after barium enemas.[4] Sonographic features include a heterogeneous mass with solid and cystic areas.[11,12] Echogenic areas within the mass may represent fat or calcification if posterior acoustic shadows are present.[11] Examination by CT and MRI can help evaluate the relationship of the tumour with surrounding structures and define the excisional plane for surgery.[1] In CTs and MRIs, SCTs show up as complex masses, which may contain fat, calcification or solid nodules, as well as fat-fluid levels.[6] The sacrum is usually intact in mature SCTs, but pressure erosion and an increased angle may become evident in chronic cases.[11] Predominantly solid tumours are more likely to be malignant.[6] Invasion of surrounding structures, locoregional lymphadenopathy and distal metastases are radiological features of malignancy.[2] The most sensitive imaging modality is CT, which may demonstrate calcification and indicate the integrity of adjacent cortical bone. Magnetic resonance imaging is superior for evaluating the anatomical relationship to adjacent organs, such as spinal canal invasion. Also, fat, fluid, and other soft tissue components may be better seen using MRI.[11,13] Both CT and MRI of the abdomen and pelvis are helpful in screening for recurrences.[8]

Biochemical markers including AFP, carcinoembryonic antigen, and HCG are helpful,[7] and tend to be elevated in patients with malignant lesions and could also be used to detect recurrences after surgery.[2] Elevated tumour markers are not expected in the benign entity. In view of the risk of dissemination of the tumour and subsequent recurrence, transrectal and transcutaneous biopsy is contra-indicated.[7,9] Based on CT-guided biopsy, an extra-rectal or presacral approach can be considered for inoperable cases, locally advanced lesions, or high-risk surgical candidates. Macroscopic examination commonly shows partially cystic lesions.[8] Microscopic features include presence of derivatives of more than one germ layer.[4] Sacrococcygeal teratomas are classified into three categories based on their histopathological features: mature, immature, and malignant. Mature teratomas contain an epithelial-lined structure, mature cartilage, often together with striated or smooth muscle. Immature teratomas contain primitive mesoderm, endoderm or ectoderm mixed with more mature elements. Malignant teratomas contain malignant tissue of germ cell origin.[2]

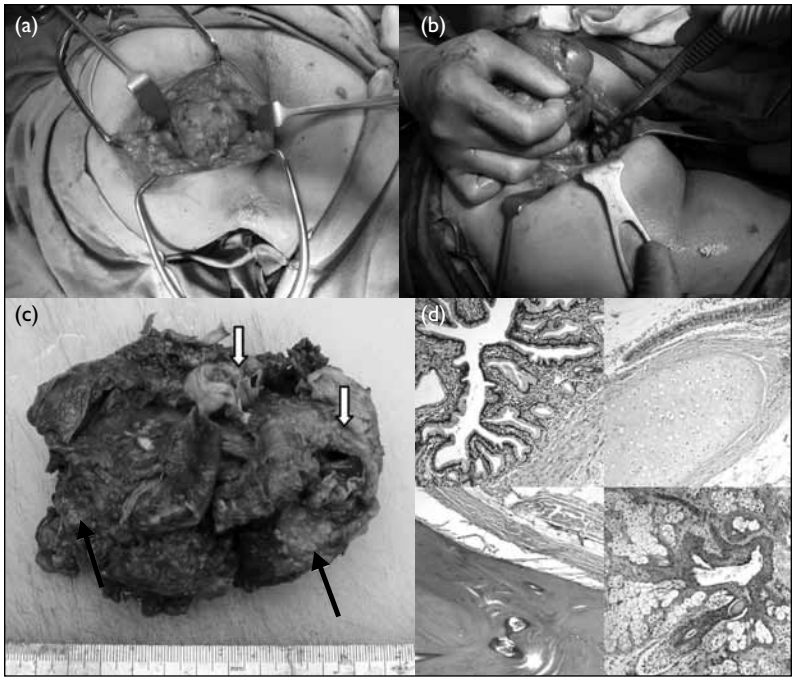

FIG 2. (a) Intra-operative photo showing the patient in the prone Jackknife position. Incision was made, exposing the tumour. (b) Intra-operative photo showing the tumour dissected from the surrounding structures. The tumour was located inferior to the pelvic floor (forceps pointing to the pelvic floor muscles). (c) Specimen photograph of the removed specimen showing a tumour with multiple cysts (white arrows) and foci of adipose tissue (black arrows). (d) Light microscopy images featuring: (clockwise from top left) intestine, bronchi, bone with skeletal muscle, and a pilosebaceous unit (H&E stain, x100)

In our case, localisation and characterisation of the pelvic mass was important, as it facilitated the surgical approach. In this special case, the tumour was retrorectal, involved the left ischiorectal fossa, and was below the peritoneal reflection and levator muscles. This special anatomical location was probably the reason why the mass was not visualised during the patient's prior laparotomy and laparoscopy. Complete excision through the perineal

approach was achieved. For an occult pelvic mass not visualised during laparotomy/laparoscopy, cross-sectional imaging by CT and/or MRI is/are advocated for better anatomical delineation.

Primary treatment of mature SCT consists of complete excision. Mature SCT is potentially curable and the prognosis is excellent.[7,8,11] The surgical approach may be transsacral or transabdomino-perineal, depending on its size and location.[1,4] Removal of the coccyx for better tumour clearance has been suggested.[2] Preoperative angiography may be considered for embolisation of large SCTs.[6] Chemotherapy and radiotherapy in addition to surgery may be indicated for malignant lesions.[8,14] Mortality is considerably higher in patients with malignant teratomas or benign teratomas that undergo malignant transformation.[2]

It has been suggested that the diagnosis of SCT should always be considered in patients complaining of pain in the sacrococcygeal region and a palpable presacral mass.[4] The differential diagnoses based on radiological features include: meningocele, rectal duplication cyst, lipoma, and liposarcoma.[9] Several distinguishing features have been identified. Meningoceles are usually noted superior to the level of the sacrococcygeal region, typically they are not multiloculated and they seldom contain fat or calcification.[11] Moreover, communication of the meningocele with the thecal sac may be demonstrable by MRI.[10] Rectal duplication cysts are not as multiloculated, seldom contain fat or calcification, and solid elements are unusual.[11] Calcification, fat, septum and soft tissue component may also be found in both lipomas and SCTs. Although the findings of fat, calcification, cystic and soft tissue component are not specific for teratomas, their presence makes the diagnosis more likely. Other differential diagnoses include: sacrococcygeal chordoma, retrorectal abscess, and tailgut cyst.

Conclusion

Although a rare tumour in adults, SCT should be considered in the differential diagnosis of patients with occult pelvic masses. For an occult pelvic mass not visualised by laparotomy/laparoscopy, cross-sectional imaging is advocated for better anatomical delineation.

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