

Case Report

A sacrococcygeal teratoma-a rare entity among adult males: a case report and review of literature

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ABSTRACT

Sacrococcygeal teratoma (SCT) is derived from embryonic germ cell layers and is one of the commonest tumours in infants. It is the most commonly occurring solid congenital tumours in the foetus and the new born. It is very rare in adults with less than a hundred cases documented in literature, with even rarer cases of male presentation. We report a case of a 26-year-old adult male presenting with a sacrococcygeal teratoma who was treated in our hospital along with its literature review.

Keywords: Adult, Male, Mature, Sacrococcygeal, Teratoma, Treatment

INTRODUCTION

Teratomas are defined as tumours consisting of different cell types which are derived from more than one germ layer. Teratomas arise from totipotential cells, which are normally found in the ovary and testes and are sometimes abnormally present in sequestered midline embryonic rests. Hence, most teratomas are located in ovaries and testes of adolescents. The most common site for extragonadal teratoma is the coccyx.

Sacrococcygeal teratoma (SCT) is derived from embryonic germ cell layers and is one of the commonest tumours in infants, with a prevalence of 1/40,000 births with a female to male ratio of 10:1.¹ Most changes can be diagnosed prenatally, 50–70% are found during first few days of life, less than 10% are diagnosed beyond the age of two years.² It is rare in adults. There is a female preponderance; most series report a female-to-male ratio of 3-4:1.^{3,4} Less than a hundred cases of teratoma in

adults have been documented in literature.⁵ With even rarer cases of male presentation. We report a case of a 26-year-old male presenting with an adult sacrococcygeal teratoma who was treated in our hospital along with its literature review.

CASE REPORT

A 26-year-old caucasian male from a rural area of Jammu and Kashmir, India presented to our hospitals surgical OPD with chief complaints of a huge mass on the left gluteal region close to the natal cleft since birth and recurrent pain in the gluteal region which was intermittent, dull in nature, occasionally colicky and relieved with ingestion of analgesics. There was associated discharge from an opening close to the mass. The mass was present since birth and gradually grew in size and due to social implications, he did not get medical advice till pain grew and he was unable to sit. He was medically fit and had no neurological symptoms. Family history was unremarkable. His physical examination

showed mass at left gluteal region measuring approximately 16×11×8 cm extending to natal cleft (Figure 2). Surface was smooth and irregular with no prominent veins. It was non-tender, firm to hard in consistency, non-pulsatile and non-compressible. There was an opening just above the mass which drained puss on pressure. Per rectal examination was normal. Neurovascular examination was unremarkable. His base line laboratory work up included alpha fetoprotein which was normal. Plain X-ray on the pelvis showed abnormal calcification on the left side of pelvis over the pubic bone.

Computed tomography revealed non-enhancing pelvic mass suggestive of sacrococcygeal teratoma. It seemed to originate from the tip of the coccyx. It was a mixture of bone, cyst and small solid component with fat predominating (Figure 1). Fistulogram was done for the opening but did not reveal any communication with the rectum.

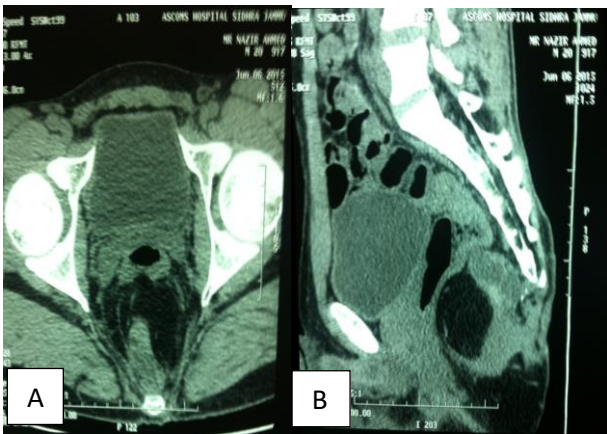


Figure 1: Abdominopelvic CT scan (A) Showing a mixed density mass with moderate peripheral enhancement and a non-enhancing central region in the pelvis; (B) Showing a heterogeneous mass occupying the presacral area.

The patient was prepared for surgery and posterior sacral approach was used for the excision of tumour. At operation, the tumour was found to be multi lobulated, arising from the sacrococcygeal region. It was not involving the major neurovascular bundles and was excised completely along with coccyx. Soft tissues and neurovascular structures were well preserved.

Adequate skin was left to cover up the wound pocket, rectum was preserved. Postoperatively dressing was done on alternate days and antibiotic cover given for 5 days. Post-operative period was uneventful and the patient was discharged after 5 days.

The tumour was 13×8.5×9.5 cm in size. Also, present were respiratory epithelium and foci of gastric-type glandular mucosa (Figure 3). A post sacral mass was also

resected; it showed fibromuscular tissue with smooth muscle (Figure 4-6).



Figure 2: Gross appearance of sacro coccygeal teratoma.

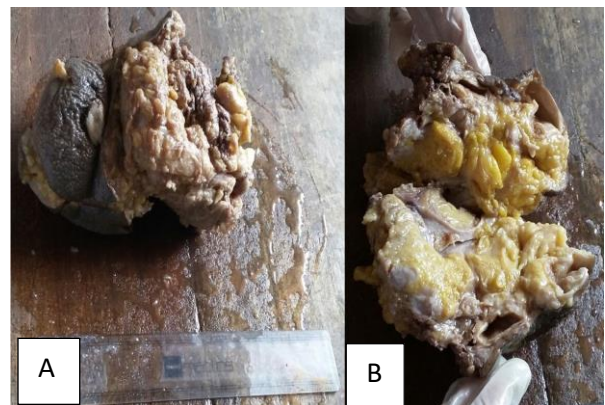


Figure 3: (A, B) There were several cysts, mature adipose tissue, and connective tissue containing numerous cysts lined by cuboidal epithelium containing keratin.

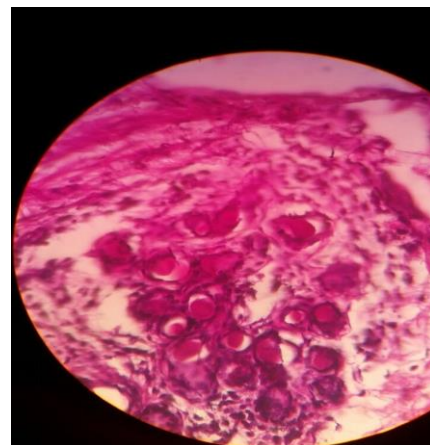


Figure 4: Showing cysts lined by cuboidal epithelium.

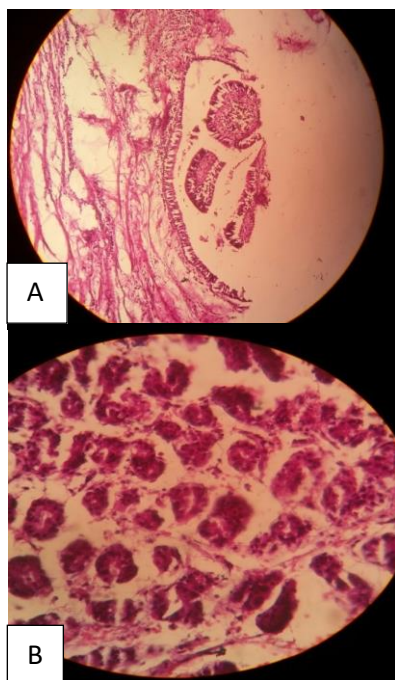


Figure 5: (A) respiratory epithelium (B) gastric glands.

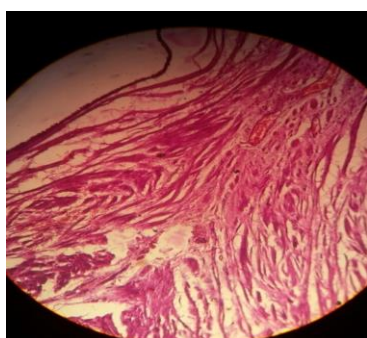


Figure 6: A cyst lined lined by cuboidal epithelium.

DISCUSSION

Teratomas are tumours which consist of different cell types that are derived from more than one germ cell layer. Teratomas do not usually arise as a result of metaplasia but contain tissue foreign to their anatomic site. They may be inherently malignant or have the potential for malignant degeneration. The most teratomas are located in ovaries and testes of adolescents. The most common site for extragonadal teratoma is the coccyx. Sacrococcygeal teratomas (SCT) are known to be the most commonly occurring solid congenital tumours in the foetus and the new born. Sacrococcygeal teratomas are rare in adults with less than a hundred cases documented in literature with even rarer cases of male presentation.⁵ There have been many theories postulated about the development of teratomas. These include derivation of the teratoma from multipotential cells of the primitive knot or Hensen's node, which are the embryonic entities that contribute to the gonadal ridge and eventually rest in

the coccygeal region. Other theories include parthenogenetic development of germ cells within the gonads or in extragonadal sites; "wandering" germ cells of non-parthenogenetic origin left behind during the migration of embryonic germ cells from yolk sac to gonad; or origin in other totipotent embryonic cells.^{6,7} A theory of incomplete twinning has also been proposed.⁸

In comparison to the pattern of presentation in neonates in which 90% of sacrococcygeal teratomas are externally visible, most of adult sacrococcygeal teratomas present as intra pelvic masses. Adult Sacrococcygeal teratomas are mostly asymptomatic in adults; usually detected by chance during imaging studies or routine clinical examination. Symptoms may be subtle and nonspecific usually related to mass effects or bulk of the tumour, such as low back pain, bowel or urinary symptoms, venous engorgement of the lower limbs and lower extremity motor power losses as result of compression of adjacent structures: rectum, bladder or uterus. A small percentage of patients may experience neurologic symptoms with lower extremity paresis or paresthesias (similar to our second patient), particularly in the late stages of malignant invasion by the tumour.⁷ A small percentage of patients may experience neurologic symptoms with lower extremity paresis or paresthesias, particularly in the late stages of malignant invasion by the tumour.⁷ Calcifications in the coccygeal region on radiography or an anterior displacement of the rectum due to the mass effect in the barium enema are findings suggestive of sacrococcygeal teratoma.⁹ More than 50% of sacrococcygeal teratomas exhibit calcification or ossification.¹⁰ The current standard includes computerized tomogram (CT) and Magnetic resonance imaging (MRI) as the most significant tools to characterize the mass, to evaluate the intrapelvic extension and relationship to other structures. The CT defines the mixed cystic-solid nature of the mass however MRI gives better tomographic evaluation and cyst evaluation which enables better pre-operation staging and planning.¹⁰ Most commonly, teratomas tend to appear as a complex mass with roughly equal amounts of solid heterogeneous and cystic areas with or without septations. They frequently present as thick-walled cystic masses, sometimes multiloculated, containing fat, calcified elements and/or small solid nodules. Complex, predominantly solid tumours with significant areas of necrosis within the tumour itself are most likely to be malignant. Invasion of adjacent structures, sacral destruction and secondary findings such as loco regional lymph node and distant metastases are clearly indicative of malignancy.¹⁰ Serum markers such as alpha-fetoproteins and human chorionic gonadotropins are not helpful in the diagnosis of sacrococcygeal teratoma, but tend to be elevated in patients with malignant lesions differentiating them from benign ones, and could also be used to detect recurrences after surgery.^{9,11} In most of the teratomas, macroscopic examination reveals partially cystic and partially solid masses, although completely solid teratomas do occur. While on microscopy presence

of derivatives of more than one germ layer are found. On the basis of microscopic examination of teratomas, they are classified as: mature, immature, and malignant. Mature teratomas (often referred to as benign 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 in addition to mature and/or embryonic tissues.⁷

The differential diagnoses based on radiological features in adults include: meningocele, rectal duplication cyst, lipoma, and liposarcoma, chordoma, neurofibroma, fibrosarcoma, giant cell tumour of sacrum, pilonidal cysts, osteomyelitis of sacrum, fistula with presacral extension and abscess formation, post injection granuloma, and tuberculosis.^{4,9} The primary treatment for all primary sacrococcygeal teratomas is complete surgical excision. Excision of the coccyx may be necessary because the bone may contain a nidus of pluripotent cells with a risk of recurrence. Most of these tumours can be removed through a sacral incision. However, if the tumour extends considerably to the pelvis and the retroperitoneum, an additional abdominal incision may be necessary to excise the tumour completely along with the coccyx bone.⁷ Pre-operative angiography may be utilized for embolisation to reduce intra-operative haemorrhage especially in large tumours. If the tumour is histologically benign (mature tissues only) or immature teratoma without frankly malignant tissue, complete excision is adequate and provides excellent results. For malignant teratomas, surgical excision alone is inadequate and patients require additional treatment with chemotherapy and/or radiotherapy. Because of rarity of these tumours, there has been no standard recommendation for the use of chemotherapy or radiation. Postoperative follow-up is very crucial and of great importance as far as the prognosis of the tumour is concerned. For patients with complete resection, a complete physical examination should be done periodically (every 1–2 months for 1 year, every 3–4 months during the second year, and then every 6 months for 3 years) with emphasis on the perineal and presacral area by a complete rectal examination. CT scans or MRI of the abdomen and pelvis are accurate in detecting recurrences. The usefulness of serum markers (AFP and HCG) to screen for recurrences is unknown in benign teratomas but are important in teratomas with malignant transformation. If these markers are elevated preoperatively, measurement of them should be repeated with follow-up examinations to detect recurrence if any.

CONCLUSION

Sacrococcygeal teratomas although very rare in adult and even rarer in adult males, should be considered in the

differential diagnosis of a pelvic mass in adults. For an occult pelvic mass not visualised by laparotomy/laparoscopy, cross-sectional imaging is advocated for better anatomical delineation. Long-term prognosis is good with complete excision of the tumour along with the coccyx bone and in tumours which have benign histology. The presence of malignant transformation and incomplete excision are associated with a less favourable prognosis and outcome.

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