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CNS Metastasis of Sacrococxygeal Teratoma- A Cytologic Presentation

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ABSTRACT

Sacrococcygeal teratoma (SCT) is a congenital neoplasm that develop embryologically from totipotent cells and enlarge as a mass in sacrococcygeal region. SCT is usually benign but in a significant number of patients it presents with metastatic disease. A 1-year-old female child presented to our hospital with a history of a sacrococcygeal swelling since birth. Investigations revealed the lump to be a sacrococcygeal teratoma. The size of swelling increased gradually over time to a size of 12 cm × 9 cm at presentation. On further work up the patient was found to have metastasis in cerebrospinal fluid (CSF). We present a very rare case of CNS metastasis in a girl with sacrococcygeal teratoma reported on cytology.

Keywords: Cerebrospinal fluid, CNS metastasis, Cytology, Sacrococcygeal teratoma.

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INTRODUCTION

Sacrococcygeal teratoma (SCT) is a relatively uncommon tumour with a reported incidence of one in 21,700 live births.[1] However, in neonates sacrococcygeal region is the commonest site for teratoma to occur and SCT is the most common congenital neoplasm.[2] The term "sacrococcygeal" is a misnomer because SCT almost always arise from the coccyx and not from the sacrum with a female preponderance in the ratio of 4:1 to 10;1. [3] SCT are congenital neoplasms composed of diverse tissues and are believed to originate, early in foetal life, from totipotential cells of Hansen's node in the coccygeal origin.[4] SCT have tissue derived from all the three germinal layers i.e. endoderm, ectoderm and mesoderm and contain squamous and intestinal epithelium, skin appendages, neural elements, teeth and calcium.

They clinically present as a large midline exophytic mass in the sacrococcygeal region. Most of the SCTs are benign but should always be looked with suspicion because they may present as a malignant tumor. They may metastasize to spine, liver, lung, lymph nodes and very rarely to some other sites. [5]

CASE SUMMARY

A one year-old female patient was brought to our hospital with a swelling in the sacrococcygeal region that had been present since birth. Initially the swelling was small in size and it gradually increased to its present size. Routine antenatal check-up was not done and the baby was full term, born by normal vaginal delivery by traditional birth attendant at home. There was no other birth complication apart from the swelling. The developmental milestones were within normal range. Family history was unremarkable.

An incisional biopsy of the swelling was performed earlier at the age of 12 weeks, findings of which were consistent with benign sacrococcygeal teratoma. Since last 15 days, there is history of irritability, head banging, poor feeding, high pitched crying and hematochezia. On examination, swelling was 12cm x 9cm, globular, firm to hard, non mobile and non-tender with bluish discolouration of overlying skin. Cough impulse and transillumination tests were negative. There was no apparent neurological deficit. Other relevant findings were bulging anterior frontenelle, distended scalp veins and hematocolpos on per speculum examination.

Imaging by computed tomography was advised which revealed an irregular solid mass with cystic areas arising from the sacrococcyx with sacral bone defects.(Figure 1) Serum AFP was raised to 1100 ng/ml. On the basis of clinical signs and symptoms of increased intracranial tension, a lumbar puncture was performed and CSF was sent for cytopathological examination. CSF cytology was performed which revealed the presence of mature squamous epithelial cells and anucleate squames (Figure 2).

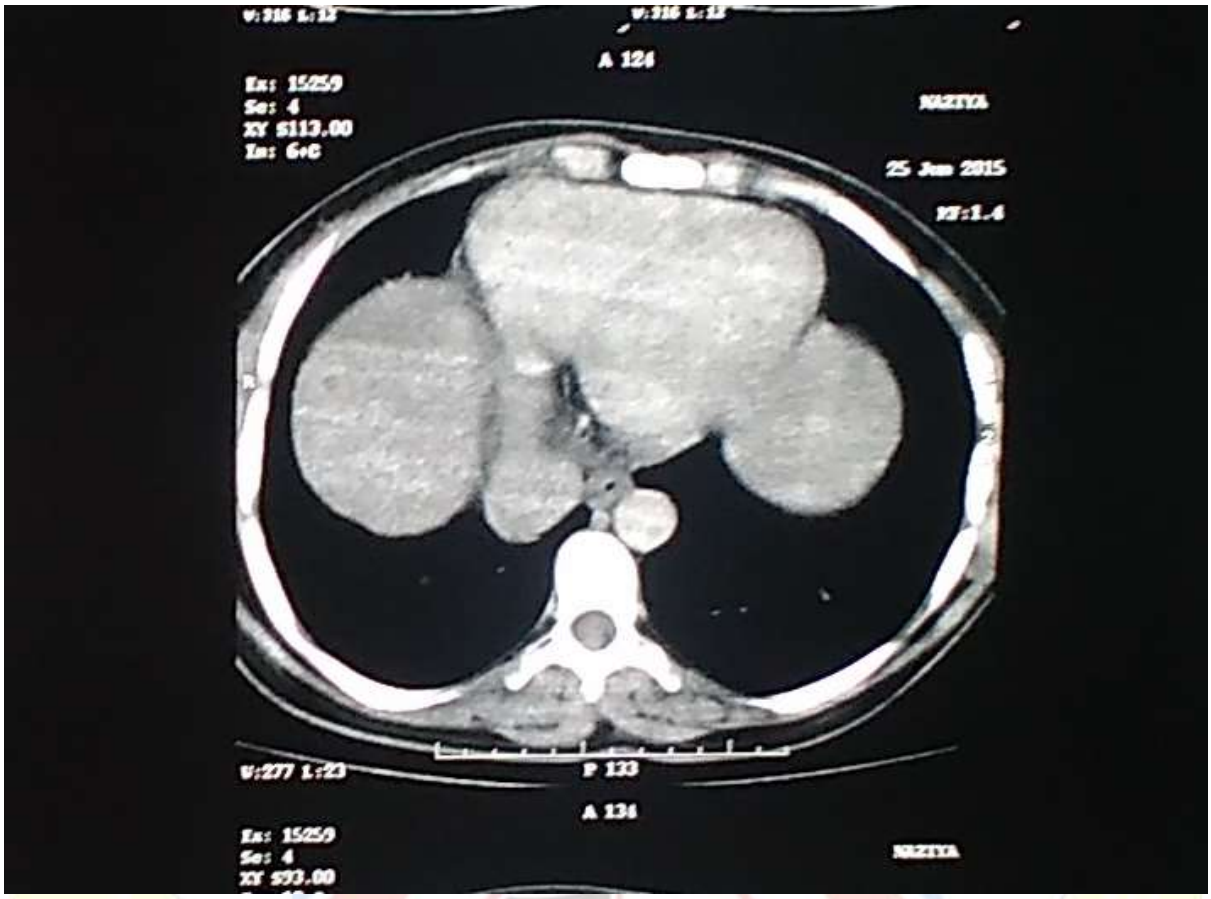

Figure 1: Imaging by computed tomography revealed an irregular solid mass with cystic areas arising from the sacrococcyx with sacral bone defects.

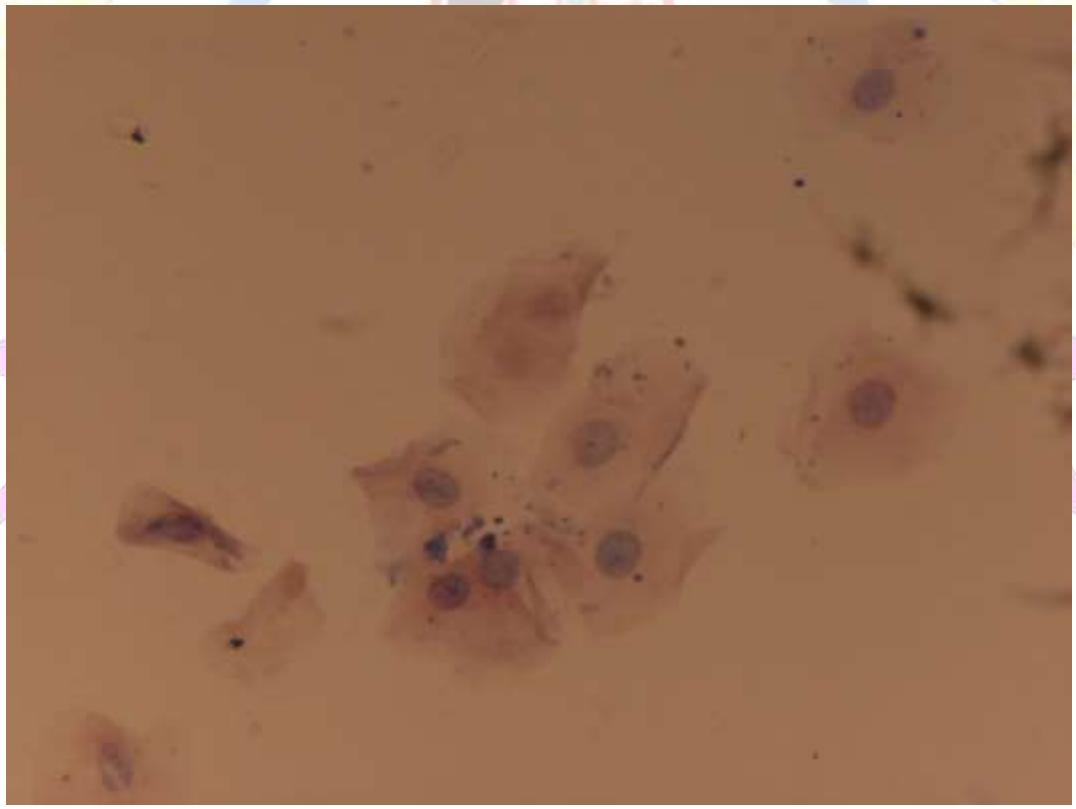

Figure 2: CSF cytology revealed the presence of mature squamous epithelial cells and anucleate squames. Haematoxylin and Eosin x 40X.

DISCUSSION

Teratoma accounts for 32.0% to 66.0% of extra gonadal germ cell tumours.[6] SCT are congenital tumors and are usually diagnosed in neonates, infants and younger children but rarely present in adults.[7] As much as 15.0% of SCTs are associated with congenital anomalies like sacral bone defects, imperforate anus, duplication of uterus or vagina, meningomyelocele and spina bifida. Scimitar sacrum, anorectal malformation and presacral mass when present together are referred to as Currarino's triad.[8] Teratomas usually present as mass in the sacrococcygeal region. Their predilection for this particular area is because of the fact that there are large number of pluripotent cells in the caudal region of the embryo, which is embryologically associated with the distal sacrum and coccyx. On the basis of presentation of mass, American Association of Pediatric Surgery has classified SCT into four types: exophytic and external (47.0%), dumbbell shaped, with equal internal and external components (34.0%), primarily located within the abdomen or pelvis (9.0%), entirely internal without any external components (10.0%).[9] In our patient, the mass was typical dumbbell shaped.

Differential diagnosis of SCT includes neural tube defects specifically meningoceles or meningomyeloceles, epidermoid cysts, pilonidal cysts, rectal abscesses, lymphangiomas and congenital lipoma. The histopathologic examination is the mainstay of diagnosis of a teratoma. Investigations like magnetic resonance imaging (MRI) and Computerized Tomography scan (CT scan) are helpful in knowing the extent of tumor in the pelvis and abdomen and also its relation to adjacent structures. [10] Prenatally, teratomas are diagnosed usually by routine obstetric ultrasonography. They appear as a mixture of cystic and solid components. Now a days, prenatal MRI has is also being used for diagnosing fetal anomalies. In our case, there was no history of any antenatal ultrasonography since the mother was illiterate and was unaware of its importance. In a case of suspected SCT, transrectal needle biopsy is contraindicated because of the possibility of dissemination or leakage of teratoma and subsequent recurrence, whether the tumor is malignant or not. [10]

Most of the SCTs are cystic and benign but there is a risk of malignancy which increases directly with age, intrapelvic component and the ratio of solid to cystic tissue. [10] If the age at the time of diagnosis is less than 2 months, the risk of malignancy is 7 to 10.0%; at 1 year of age it is 37.0% and at 2 years, it is 50.0%. [9] SCT which persists into adulthood are mostly benign. Tumors which are cystic and well differentiated with mature elements are usually benign while solid tumors with embryonic elements tend to be malignant. SCTs are more common in females but are more often malignant in males.[9] In SCT, malignancy usually arises from a single germ line and generally develops from embryonic tissues. Serum levels

of the fetal alpha fetoprotein, carcinoembryonic antigen and human chorionic gonadotrophin are raised in patients with malignant SCT. [10,11]

Well encapsulated and benign SCTs can sometimes undergo malignant transformation, usually into squamous cell carcinoma.[12] The potential for late development of a malignancy in previously benign lesion is probably because of the retained capacity for continued embryonic growth. This point is further emphasized by the fact that rate of malignancy is higher when the tumor is diagnosed at an older age.[5] It is therefore recommended that they be excised as soon as possible because surgery is the principal mode of treatment in SCT. Sometimes the patient presents with a metastatic disease at the time of diagnosis, similar to our case which presented with CNS metastasis. Complete removal of the mass should be attempted as recurrence has been reported in upto 37.0% cases, if the coccyx has not been excised.[13] It has been reported that malignant tumors have more chances of recurrence and the incidence of malignancy is higher in recurrent than in primary tumors. So it is imperative to follow up the patients postoperatively, with thorough physical examination, serum alpha-fetoprotein and ultrasonography for at least first three years to detect a likely recurrence.[14]

CONCLUSION

Sacrococcygeal teratomas constitutes a significant pediatric surgical problem. They are usually benign but they sometimes undergo malignant transformation and may present with metastasis. Early diagnosis and immediate surgical excision of the tumor is highly recommended to prevent any metastasis. This rare presentation will help clinicians in diagnosing such cases and providing immediate treatment and better patient care.

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