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Case Report

Sacrococcygeal teratoma: a case report

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ABSTRACT

Teratomas are germ cell tumours composed of multiple types of cells derived from more than a single germ cell layer. The most common site of an extra gonadal teratoma is the sacrococcygeal region. We report a case of a 10-day-old female child with a large swelling in the sacrococcygeal region extending laterally into the buttocks. X-ray showed a large mass projecting from the lower pelvic region and delineate the extent of the tumour and the involvement of tissues. Image findings and clinical presentation led to the diagnosis of sacrococcygeal teratoma. Tumour was surgically excised with prior pre-anaesthetic checkup, post-operative period was uneventful. She was discharged with advice to come in follow-up surgical outpatient clinic. There was no evidence of tumour recurrence till 1 year of age. She had normal growth curve for her age with normal developmental milestones.

Keywords: Congenital tumour, Germ cell tumour, Sacrococcygeal teratoma

INTRODUCTION

Sacrococcygeal teratoma (SCT) has an incidence of about 1/40,000 live births and is the commonest congenital tumour in the neonate.[1] In 1973, Altman classified SCT into 4 types based on the external component and intra-pelvic/intra-abdominal extension of the tumour (American Academy of Pediatrics surgical section classification).[1] The SCTs seen at birth are usually Altman Type I and II (87%).[1] Rarely Type III can also be seen in neonates.[2] Type IV is typically seen later in life as there is no external component.[1] More common in female. It is caused by the retention of large amount of primitive totipotent cell in this region may be the reason of this tumour. Tumours of sacrococcygeal location originate ventral or dorsal to the sacrum, they may grow posteroinferiorly into the gluteal area or anterosuperiorly into the lesser pelvis. Specially in the ventral location these tumours may grow to a large size as they develop into the retrorectal or presacral space.[3] The majority of the cases they are benign tumors; the risk of recurrence or malignant transformation is low. SCT is usually discovered either because a blood test performed on the mother at 16 weeks shows a high alpha fetoprotein (AFP) amount, or because a sonogram is performed because the uterus is larger than it should be. The increased size of the uterus is often caused by extra amniotic fluid, called polyhydramnios. The diagnosis of SCT can be made by an ultrasound examination[4].

CASE REPORT

A 10 day old female child presented to general surgery opd of Bokaro General Hospital a tertiary care hospital in Bokaro steel city, Jharkhand, India with large swelling in the sacrococcygeal region. Baby was born premature to a healthy 27 year old mother after an uncomplicated gestation of 34 week though caesarean section.

Examination reveals a non tender mass of approximately 12.5 cm×13 cm in size in gluteal region (Figure 1). The overlying skin has ulceration on its posterior aspect.

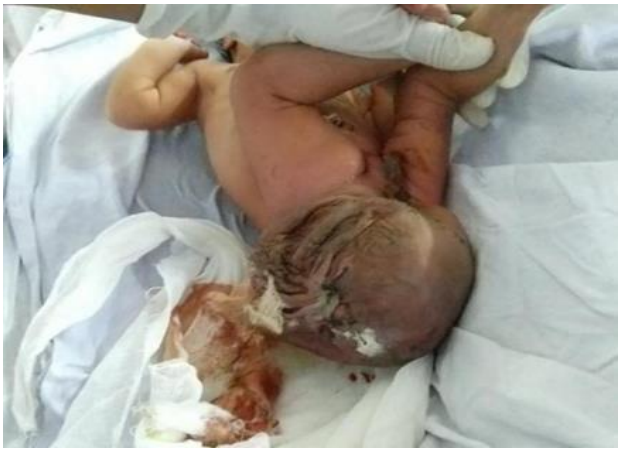

Figure 1: Pre operative: a large mass in sacrococcygeal region with ulceration.

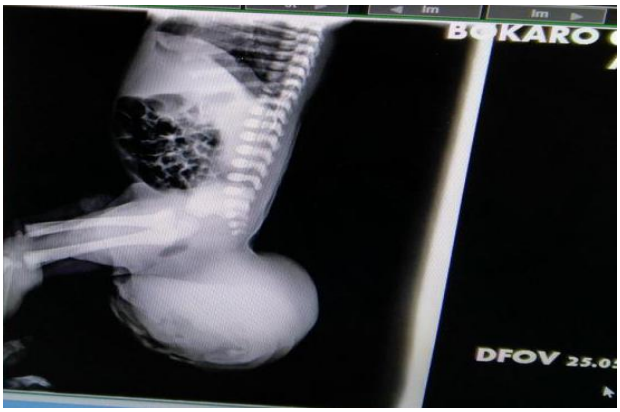

Figure 2: X-ray showing a large mass projecting from the lower pelvic region and the involvement of tissues.

Among investigations the laboratory investigations are within normal limits and the x-ray (Figure 2) delineate large mass projecting from the lower pelvic region and the extent of the tumour.

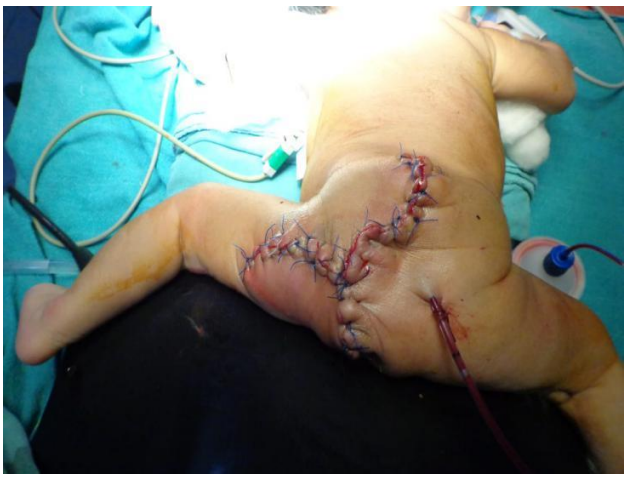

Figure 3: Post operative wound after excision of sacrococcygeal teratoma.

Tumour was surgically excised with prior pre-anaesthetic checkup, post operative period was uneventful (Figure 3). She was discharged with advice to come in follow-up surgical outpatient clinic. There was no evidence of tumour recurrence till 1 year of age. She had normal growth curve for her age with normal developmental milestones.

DISCUSSION

Sacrococcygeal teratomas are extra gonadal neoplasms arising in the presacral area. They have an incidence of 1 per 40,000 live births, and a prevalence of 1 in 21,000 births. Although sacrococcygeal teratoma is a rare tumor, it is the most common malignancy of germ cells in newborns and children under two years. Its presentation may be forming large cysts or as solid mass. They are usually located in the mid line of the body. In general, the order of frequency regarding to its location is as follows: sacrococcygeal, gonadal, retroperitoneal, cervical, mediastinal, oropharyngeal and other (gastric, hepatic, intracranial). It predominates in females, but in males malignancy is more recurrent.[5-7]

The term "teratoma," derives from the Greek word "teraton" meaning monster. In 1869 Virchow applied this term to a tumor originating in the sacrococcygeal region.[8]

The symptoms that occur with sacrococcygeal teratomas vary widely depending upon the size and specific location of the tumor. Small tumors often do not cause any symptoms and can usually be removed surgically after birth without difficulty.

However, larger sacrococcygeal tumors can cause a variety of complications before and after birth. Sacrococcygeal teratomas can grow rapidly in the fetus and require very high blood flow resulting in fetal heart failure, a condition known as hydrops. This is manifest as dilation of the heart, and the collection of fluid in tissues of the body, including the skin and body cavities such as pleural effusion, pericardial effusion, and/or ascites. In addition to hydrops, which can occur in approximately 15% of very large fetal sacrococcygeal teratomas, these tumors can cause polyhydramnios, hydronephrosis, bleeding into the tumor or rupture of the tumor with bleeding into the amniotic space, or dystocia. It is very important to have very close monitoring during pregnancy to recognize these symptoms as early as possible.

In adults, sacrococcygeal teratomas may not cause symptoms. In some cases, they may cause progressive lower back pain, weakness, and abnormalities due to obstruction of the genitourinary and gastrointestinal tracts.

In most cases, sacrococcygeal teratomas are diagnosed at birth when a large tumor is detected protruding from the sacral region. Many sacrococcygeal teratomas are found

incidentally on routine prenatal ultrasounds. If a sacrococcygeal teratoma is diagnosed prenatally a careful examination is usually done to rule out other anomalies. In some institutions a fetal MRI scan is also performed to better delineate the anatomy of the tumor and displaced structures. Other specialized imaging techniques may be used to diagnose a tumor as well as evaluate the size, placement, and extension of the tumor and to serve as an aid for future surgical procedures. After birth, such imaging techniques may include computerized tomography (CT) scanning and magnetic resonance imaging (MRI).[9]

Sacrococcygeal teratomas are classified according to the American Academy of Pediatrics Surgical Section:[10]

Type I - the tumor is predominantly external with a very minimal internal component. Type I is rarely associated with malignancy.

Type II - the tumor is predominantly external but has some internal extension into the presacral space.

Type III - the tumor is visible externally but is predominantly located in the pelvic area with some extension into the abdomen.

Type IV - the tumor is not visible externally and is located in the presacral space. Type IV has the highest rate of malignancy.

Benign teratomas and immature teratomas may produce small elevations of alpha-fetoprotein and human chorionic gonadotropin β. In case of malignant sacrococcygeal teratomas will have a component of yolk sac tumor, along with elevations in alpha-fetoprotein, which are monitored in series during treatment to help assess their response.[11]

The main treatment for sacrococcygeal teratoma is complete resection of the tumor and the coccyx. If this procedure is not performed, the risk of recurrence is extremely high. In patients with mature sacrococcygeal teratoma, the only recommended treatment is surgery, followed by rectal examinations and interval serum AFP levels to monitor for recurrence for 3 years before they are considered cured with no possibility of tumor recurrence.

CONCLUSION

Saccrococcygeal tumour is an uncommon tumour but most common of the large tumour in 1st 3 months of life. In the majority of the cases they are benign tumors; the risk of recurrence or malignant transformation is low. It is important to recognize the existence of this pathology in order to have the clinical expertise that offers timely diagnosis, an appropriate and multidisciplinary treatment. Monitoring with alpha-fetoprotein and ultrasound is a key to detect recurrence or postoperative complications.

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