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Surgery

KEY WORDS:

SACROCOCCYGEAL TERATOMA: A CASE REPORT

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ABSTRACT

The sacrococcygeal teratomas are rare benign tumors derived from totipotent embryonic cells of the caudal region. The purpose of this article is to report our experience in management of two cases of giant sacrococcygeal teratomas in Ziguinchor Regional Hospital and emphasize the need for prenatal diagnosis and management before complications.

INTRODUCTION

Sacrococcygeal teratomas are benign tumors derived from totipotent cells of the caudal region during embryogenesis (1).

The incidence of sacrococcygeal teratoma is between 1/35000 and 1/40000 births (2-4). Diagnosis of these tumors are ideally done in the antenatal period (5). Depending upon the size and vascularity of tumor complications varies from intra partum rupture of tumor during vaginal delivery and degenerative changes in tumor especially after the age of 4 months of these tumors (6-8).

Case Report

Herewith presenting a case of 10 months old baby girl brought with huge swelling in the sacral region since birth. At the time of birth mass was of lemon size for which parents have been advised further investigations but due to poor compliance of parents it has been missed. Before presenting to current size, during last 9 months swelling rapidly progressed in size involving lumbosacral region and significant shifting of anal opening ahead (Fig.1). The swelling is painless, hard in consistency with approximate size of 30x30cm and not associated with any bladder and bowel abnormalities. Baby was investigated with MRI spine showed huge solid mass extending towards presacral region involving coccyx with suspected involvement of spinal canal. During last 6 months parents visited multiple hospitals for treatment but were denied due to risk of loss of bladder and bowel function postoperatively secondary to damage to spinal cord. Finally, baby was brought to Mimer medical college, Talegaon hoping to get treated. Baby was thoroughly investigated radiologically and haematologically diagnosed with type 2 of huge sacrococcygeal teratoma and planned for exploration of tumour. MRI spine was suggestive of a large well-defined 21.2x15.8x11.3 cm sized mass lesion seen arising from the pelvis extending inferiorly into the perineum causing a large swelling inferiorly.

The lesion shows irregular fluid intensity and fat intensity areas within. The lesion shows septae and few irregular areas within showing mild enhancement. The lesion shows a cystic area within the pelvis anterior to sacrum at the superior aspect of the lesion. The lesion is abutting the sacrum with no bony defect in the sacrum. The lesion was compressing the urinary bladder anteriorly. The coccyx is not visualized separately from the lesion. Features are most likely suggestive of cystic variant of type 2 saccrococcygeal teratroma. Surgical exploration was done in jack knife position with inverted y incision (Fig.2). On raising the skin flaps on either side tumour is excised along with coccyx. Tumour was excised properly avoiding damage to rectum and sphincters. After in toto excision of tumour, wound is closed after putting room-vac drains in situ and gluteal reconstruction was done (Fig.3). On postoperative day 5 drain was removed and stitches were removed on postoperative day 10. Patients had no loss of bladder and bowel function postoperatively and the baby recovered well. Histopathology study confirmed findings of Matured Cystic Teratoma with free margins of tumour.

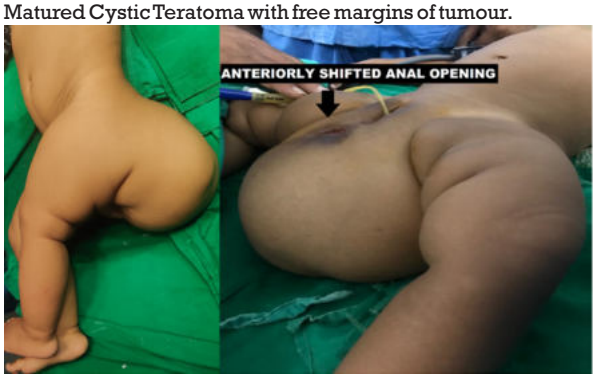

Fig 1. Rapidly Progressing Swelling Involving Lumbosacral Region And Significant Shifting Of Anal Opening Ahead.

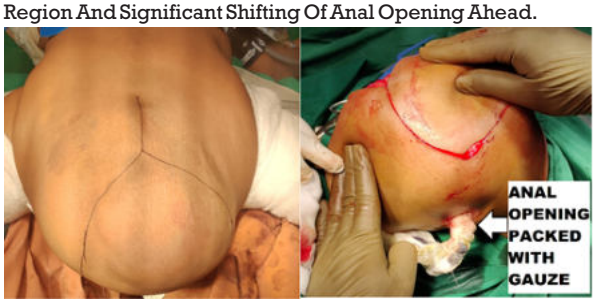

Fig 2. Inverted Y Shaped Incision To Raise The Flaps.

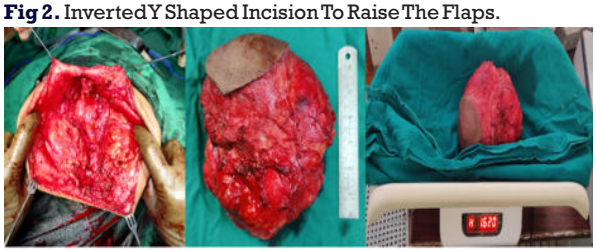

Fig 3. In Toto Excision Of Tumour Along With Coccyx Weighing 1.620kg

DISCUSSION:-

On Histopathological examination of the resected tumor and coccyx demonstrated matured cystic teratoma with free

resected margins. Sacrococcygeal teratoma is the most common neoplasm found in newborns, incidence higher female predominance being 4 to 6:1 (9). During embryogenesis regression of normally present embryonic blastema cells present in human tail bud occurs by 32 to 35 days of gestation. Hyperplasia of these embryonic blastema cells leads to sacrococcygeal teratoma (10). Coccygectomy is performed at the time of tumor resection to reduce the rate of recurrence of tumor. The chances of recurrence is reported up to 37 % if concomitant coccygectomy is not performed during resection of sacrococcygeal teratoma (11). Sacrococcygeal teratomas are classified into 4 types as per most widely accepted the Altman classification of the American Academy of Paediatrics. Type 1 is entirely external to the pelvis, type 2 has an external component as well as a small presacral component, type 3 has an external component but a large internal component extending into the abdomen and type 4 is entirely pelvic without an external mass. The majority of patients have type 1 or 2 SCTF approximately 36% of cases are type 1, 27% type 2, 18% type 3 and 18% type 4 (12). It is also important to identify the giant or rapidly growing, Hyper vascular sacrococcygeal teratoma. These hyper vascular lesions are associated with a very high fetal mortality and a high risk of exsanguinations at the time of surgical resection unless the tumor is de-vascularized by ligation of the sacral artery (13). The hyper vascular lesions often lead to congestive heart failure from high output shunting and coagulopathy from intra tumoral bleeding and consumption of clotting factors. Cross-sectional imaging has an important role in the evaluation of SCT, particularly of the rapidly growing hyper vascular SCT. Both sonography and MRI can provide assessment of the vascularity, size and Altman grade of the tumor. Magnetic resonance angiography can be used to demonstrate the course and size of the median sacral artery, which is the usual source of blood supply to SCT. The high contrast resolution of MRI and its ability to identify tumor margins and vessels makes it preferable to computed tomography for assessing the tumor extent. The mortality in giant hyper vascular SCT is particularly high in the presence of placentomegaly and hydrops. If the affected fetus is a singleton, with normal chromosomes, less than 30 weeks gestation and has high-output congestive heart failure from a type1 or 2 SCT, then fetal debulking can be successfully performed and can be lifesaving (14).

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