Original Research Paper

Volume - 14 | Issue - 02 | February - 2024 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Paediatric Surgery

A COMMON TUMOR IN AN UNUSUAL LOCATION IN A 10-YEAR-OLD BOY: A CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT **Introduction:** Pre-sacral mature cystic teratoma is a rare entity in males and because of its low incidence amongst males at this site, its management is not clearly defined. **Case Report:** We report a case of a 10-year-old boy presenting with abdominal pain, abdominal distension, bowel bladder complaints and bilateral swelling of leg. There was a palpable lump in the right lower quadrant on abdomen examination. Ultrasonography and CECT confirmed a large pre-sacral lump compressing rectum and bladder outlet. The child was taken up for exploratory laparotomy. The pre-sacral mass was resected with great difficulty and since the colon was hugely distended, an ileostomy was made which was subsequently closed. **Conclusion:** Pre sacral teratomas in males are rare and may carry nonspecific complaints such as constipation and urinary obstructive symptoms. The location of the lump causes bowel and bladder symptoms to start early but clinically becomes difficult to diagnose due to which the management becomes delayed. The location of the tumor also makes it extremely difficult to remove specially if the size is big.

KEYWORDS : Pre-sacral teratoma, dermoid, pre-sacral tumor, pediatric tumor

INTRODUCTION

Various types of masses may affect the presacral region of children. Such masses often present with non-specific symptom and thorough clinical and radiological examination is important to decide the type of lesion and its management. Sacrococcygeal teratoma are the most common germ cell tumour originating from totipotent germ cells. (1)(12) More commonly seen in female than male (1:3-1:4). (2) There is a paucity of documented presacral teratoma cases in males in India. We report a case of a presacral MCT treated in our hospital.

Case Report

A previously healthy 10-year-old male presented to the outpatient department with complaints of abdominal distension for the past 1.5 years. The boy also had progressively increasing constipation and incontinence of urine. The increasing intensity of colicky pain, abdominal distension, need to press the abdomen to evacuate stools and bilateral leg swelling were the reasons for his presentation to the hospital.

The child was examined and was found to have bilateral pitting edema. Abdominal examination revealed a mass measuring 13 cm*1 cm (length*breadth) occupying hypogastrium, umbilical region and right iliac fossa. There was visible bowel peristalsis seen. The mass could also be appreciated on per rectal examination situated in the pre-sacral area.

Hematological examination revealed slightly elevated serum LDH and normal Serum HCG and Alfa-Feto protein. Subsequently, the boy was subjected radiological examination by sonography and CECT which revealed a mass of size 11.6X10X18 cm in the pre-sacral region and causing pressure effect on rectum and bladder outlet. There was also bilateral hydroureteronephrosis. In view of all of these findings a diagnosis of a benign pre-sacral mass lesion with mass effects on bladder outlet and rectum.

The patient was counselled and taken up for exploratory laparotomy. There was a hugely distended urinary bladder and equally distended left colon. The access to the recto-rectal space was restricted because of the abovementioned distensions. To gain access the sigmoid colon was divided and decompressed and then the recto-rectal space was approached. The lump was big, as already shown on radiological examination, it was difficult to dissect. The content was similar to dermoid cyst and therefore decompression was also not possible. A cystotomy was done and using a large bore suction and ovum forceps the content of the cyst was reduced and then slowly and carefully it was dissected completely.

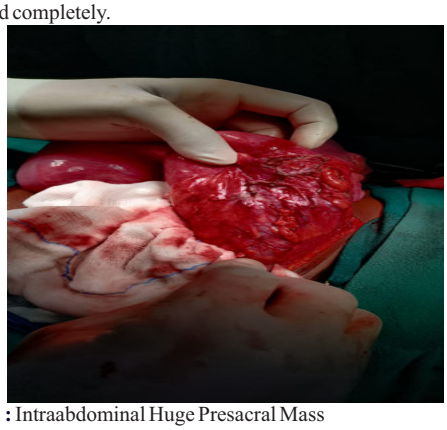


Figure1 : Intraabdominal Huge Presacral Mass

The colon was anastomosed side to side using staplers and due to the hugely distended colon an ileostomy was made. The resected mass was sent for histopathological examination which revealed a cystic mass that microscopically was lined by stratified squamous epithelium and adipose tissues, this was ultimately classified as a mature cystic teratoma. Post operatively, the patient had an uneventful recovery and was discharged on post-operative day 5. He was followed up closely and it was noted that all his obstructive symptoms of bowel and bladder had disappeared. The loop ileostomy was closed after 6 months. The child has been on follow up for the last 6 months and is asymptomatic.

DISCUSSION

Pre sacral masses arise in the pre-sacral region that is an embryological fusion of the hindgut, proctodeum, neural elements and bone. Anatomically, the pre-sacral space is the potential space between the rectum anteriorly and the lumbosacral spine posteriorly (3). These masses can contain tissues derived from all germinal layers and carry risks of malignant transformation. They are rare in pediatric population with most cases seen at a median age of 50 years and having a female predominance. (2) Our case is unique for the age of its

presentation and the sex of the patient.

Pre-sacral masses can be congenital or developmental or may even arise due to inflammation. These masses may contain neural, vascular, lymphatic or mesenchymal origins and can be uni-focal or multi-focal developments. A list of pediatric pre sacral masses is given in table 1. (4)

The presenting symptoms of the case that was peculiar was the bilateral swelling seen in the lower limbs. Most commonly, patients having a Mature Cystic Teratoma (MCT) in the abdomen present with pain in lower abdomen as the main symptom which was also seen in our patient. This is seen due to the compression of nerves and surrounding viscera.(5) Other symptoms include lower back pain, perineal pain, constipation, urinary and fecal incontinence, some of which was also present in our patient. In cases of presacral masses, a digital rectum examination is very important as it is seen that in 84-97% of the cases, the tumor is palpable and can help define the boundaries of the mass. (6) (7). The history of weight loss may be alarming in such cases as it could hint towards malignancies and since our patient did not have documented proof for the same, we had to factor in the chronicity of the complaint and ensure the mass didn't have multiple metastatic foci. Swelling of the lower limb and poor urinary output was indicative of the involvement of the renal system or urinary system.

The management flowchart for presacral masses is given as flow chart 1. Imaging would be required in this case and hence the patient was sent for radiological imaging. (7)

As for the investigations, Ultrasonography (USG) is a good primary imaging modality to define the extent of the mass and sonographically Mature Cystic Teratomas (MCT) may present as a cyst, containing a solid or complex tumor mass as seen in our case as well.

Further Computed Tomography (CT) scan is required to decide the surgical intervention to be taken by defining the anatomical boundaries, size and density of the mass.(8) Radiological investigations are very important due to their ability to correctly generate a pre-operative diagnosis of pre-sacral tumors. (7,9)

Surgical intervention is the mainstay in the treatment of MCTs. Studies have stated that the surgical approach in such cases depend on the nature and location of the lesion.

Previous studies have reported that lesions that extend up to S4 should be resected trans-abdominally, while those that extend below S4 and are <8 cm in diameter should be resected trans-sacrally, while those that in an intermediate position should be removed using a combined abdominal and sacral surgical procedure. (10,11) Since the mass did not extend to S4 and had no evidence of sacral involvement, a transabdominal approach was chosen in the form of a laparotomy.

On excision of the mass, it was observed that the descending colon was dilated due to the chronic pressure effect of the mass and was also adherent to the small bowel loops. So instead of doing adhesiolysis which would have prolonged the surgery and caused major metabolic changes in the patient, an ileostomy was done along with end-to-end anastomosis with the descending colon.

We are aware that laparoscopic surgery could be done in cases of pre-sacral teratoma, the dilated descending colon needed to be repaired and anastomosed as well. (10)

It is important to send the sample for histopathological investigation to find the degree of maturity of the teratoma and for correct identification of the specimen. Immature teratomas have a higher possibility of converting to malignant lesions.(6)

In our case, the histopathology was suggestive of mature cystic teratoma. The patient was followed up and the stoma was closed after 6 weeks. The further follow ups are uneventful.

CONCLUSION

Presacral Mature Cystic Teratomas (MCT) are rare and present with non-specific symptoms sometimes such as constipation and lower urinary tract symptoms as they did in our case. Owing to its location and its ability to cause obstruction early without being clinically detectable, diagnosis is delayed. In addition the resection of the mass in this region is also extremely challenging.

Through this article we want to emphasize that one must have a high degree of suspicion, especially in pediatric patients presenting with both bowel and bladder obstruction. and should do radiological evaluation early to diagnose these lesions early. The tumor must be completely resected during the operative procedure to prevent its recurrence as MCT can be premalignant.

Conflict of Interest: No conflicts of interest.

Source of Funding: the authors declare no source of funding.

Declaration of the patient consent form: Consent was taken from the patient for publishing the manuscript.

Acknowledgment: None.

Table 1 : Classification of pediatric pre-sacral masses

Classifications of Pediatric Pre-sacral Masses
Congenital and developmental masses Germ cell tumors (sacrococcygeal teratoma, germinomatous and nongerminomatous germ cell tumors) Anterior sacral meningocele Developmental cysts (epidermoid cyst, dermoid cyst, enteric cysts [rectal duplication cyst, tailgut cyst]) Cystic lymphangioma Lipoma
Neurogenic masses Neuroblastoma Ganglioneuroblastoma Ganglioneuroma Schwannoma Neurofibroma
Inflammatory masses Inflammatory bowel disease Granuloma Perirectal abscess
Mesenchymal masses Rhabdomyosarcoma Undifferentiated sarcoma Vascular masses Fibroma
Lymphomatous masses Lymphoma
Extension of sacral bone tumors Giant cell tumor Aneurysmal bone cyst Chordoma Osteoblastoma Ewing sarcoma family Osteogenic sarcoma
Other pre-sacral masses Hematoma Extension or metastasis to the pre-sacral space from another site

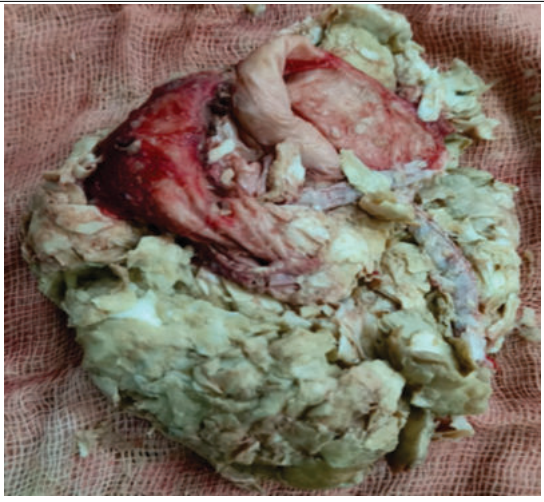
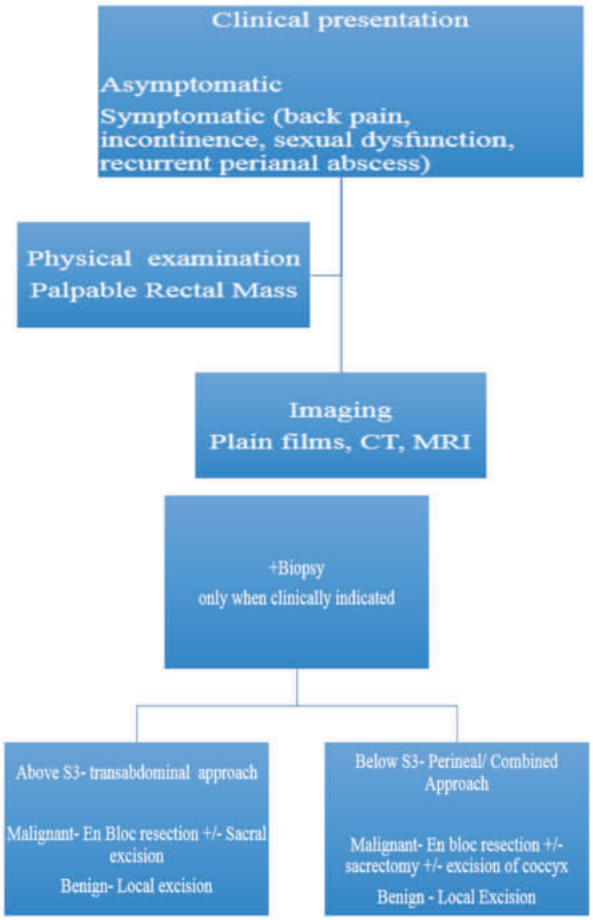


Figure 2: Evacuated Content Of Cyst



Flowchart 1: Management flow chart for sacral masses. (7)

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