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Mature Cystic Teratoma of Retro Peritoneum in a Three Years Old Female Child: A Case Report and Literature Review

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ABSTRACT
The most common type of germ cell tumor is teratoma, divided into three types: mature, immature (or malignant type) and monodermal. Retro peritoneal teratoma are rare lesions seen in 3.5-4% of all germ cell tumors in children and 1-11% of primary retro peritoneal neoplasms. Presenting a case of a three years old female child with abdominal pain and inability to pass urine. On general examination, the child had pallor. Abdominal examination showed a large palpable lump in the right hypochondrium, extending up to the right lumbar region. Computed tomography (CT) scan of the abdomen, showed, a large retro peritoneal teratoma in the right supra-renal region. This was surgically excised. Histopathological examination showed it to be mature cystic teratoma of the retro peritoneum.

INTRODUCTION

Germ cell tumors are congenital tumors containing derivatives of all the three germ layers. The most common type of germ cell tumor is teratoma, divided into three types: mature, immature (or malignant type) and monodermal. The mature type of teratoma is the most common type, which is a benign solid or cystic tumor[1]. They are frequently seen in gonads[2]. They may present in extra gonadal sites also like media stinum, sacrococcygeal region and retro peritoneum[3]. Retro peritoneal teratoma are rare lesions seen in 3.5-4% of all germ cell tumors in children and 1-11% of primary retro peritoneal neoplasms[2]. Due to the retro peritoneal location, these tumors can grow to be quite large before signs or symptoms are detected. Patients usually present with abdominal distension or a palpable mass. Radiological features include presence of calcification, teeth and fat., however, calcification cannot be considered an indicator of a benign tumor since 12.5% of calcified tumor are malignant[4]. The prognosis of neonatal teratoma is favourable with an 80-100% survival reported after surgical excision of the tumor and treatment of any recurrence[5]. Here we report a case of mature cystic teratoma of retro peritoneum in a three years old female child.

Case Report: A three years old female child presented with chief complaints of abdominal pain since the last 2-3 days and inability to pass urine in the last 1 day. There was no h/o vomiting, constipation, loose stools, fever, melaena, hematemesis or Koch's contact. On clinical examination, the child was having mild pallor. There was no icterus, cyanosis, or lymphadenopathy. Abdominal examination showed a large palpable lump in the right hypochondrium, extending up to the right lumbar region. It was firm to hard in consistency, non-mobile and bimanual palpable. The per-rectal examination was normal. Blood investigations showed hemoglobin 9.5 g/dl and normal routine biochemical tests. Serum alpha-fetoprotein and serum beta-HCG levels were normal.

Imaging Findings: Plain radiograph showed a large mass on the right side of the abdomen, displacing the bowel loops to the left side. Abdominal ultrasonography (USG) showed a large lobulated multi-cystic mass involving right hypochondrium and lumbar regions, with few solid components and calcification, in its superior part. Our impression was lymphangioma with hemorrhage or retro peritoneal cystic tumor like teratoma. Computed Tomography (CT) showed a large well-defined, multi-spatted, solid-cystic, retro peritoneal lesion, involving the right hypochondrium and lumbar regions, with dense calcification and fat component within. The right adrenal gland was not seen, separately, from this lesion. This lesion was seen to compress the IVC, throughout its extent. There was mild ascites. No significant abdominal lymphadenopathy was seen. There was moderate pleural effusion on the right side, with underlying passive sub-segmental collapse. Features were suggestive of retro peritoneal tumor, such as cystic teratoma.

Surgery: Exploratory Laparotomy was done with a transverse supra-umbilical incision. There was a large, firm, retro peritoneal mass in the right supra-renal region, causing extrinsic impression on the right kidney, with relative sparing of the right renal parenchyma. The small bowel loops were displaced to the left. The mass was excised completely and sent for histopathological study.

Histopathology: Gross pathological examination showed a large lobulated mass, with variegated solid, cystic areas on cut section. On microscopic examination, elements of ectoderm, endoderm and mesoderm were seen. Cystic areas were focally lined by areas of stratified squamous epithelium, focally by areas of ciliated pseudo stratified columnar epithelium and, at few places, by gastrointestinal epithelium, Areas of mature cartilage, adipose tissue, muscle, bony tissue and neural tissue identified. Histopathology and microscopy revealed the tumor to be a mature cystic teratoma of the retro peritoneum. Differential Diagnosis include, Cystic mesothelioma/Malignant fibrous histiocytoma/Dedifferentiated Liposarcoma. The post-operative period was uneventful and the child was discharged after 7 days.

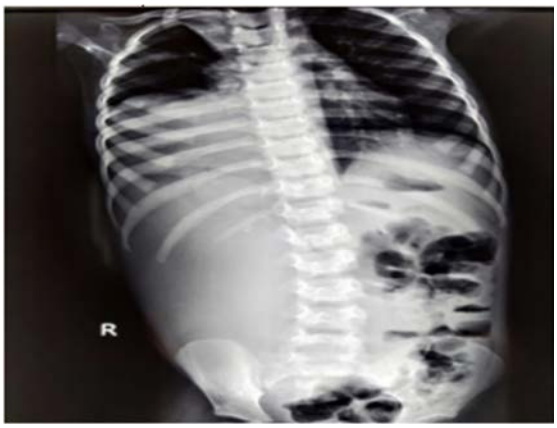

Fig. 1: Plain Radiograph Showed a Large Opacity Involving The Right Half of Abdomen, Displacing. The Bowel Loops to the Left

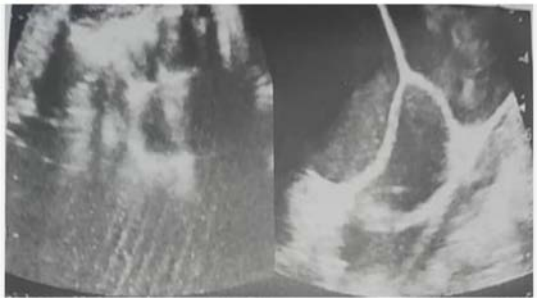

Fig. 2A

Fig. 2B

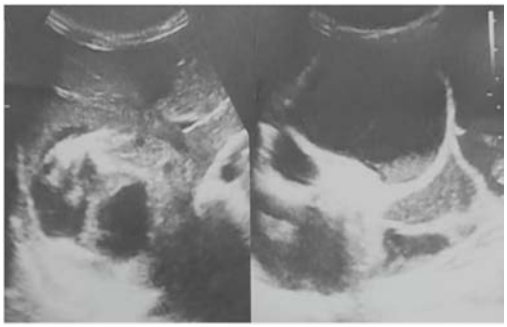

Fig. 2C

Fig. 2D

Abdominal Ultrasound (USG) Showed a Large, Lobulated Multi-Cystic Mass, Measuring 18.2×12.7× 8.3cms, in the Right Side of the Abdomen, Involving the Right Hypochondrium and Lumbar Regions (Fig. 2C, 2D). Few Solid Components with Calcification, are Seen, in Superior Part, Which is Indenting Liver and Pancreas (Fig. 2A, 2B) (Fig. 2A-Blue THICK ARROW). Few Cystic Components with Debris Seen, Suggesting Secondary Hemorrhage (Fig. 2D) (Blue Straight Arrow)

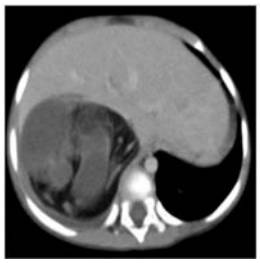

Fig. 3A

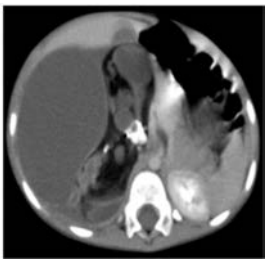

Fig. 3B

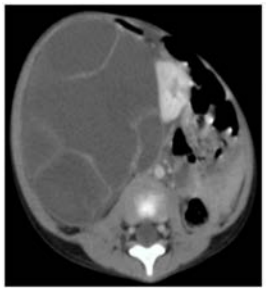

Fig. 3C

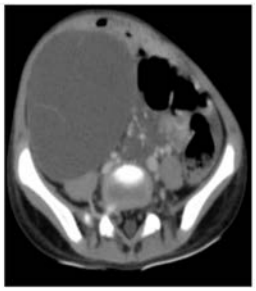

Fig. 3D

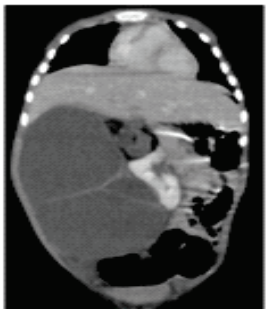

Fig. 3E

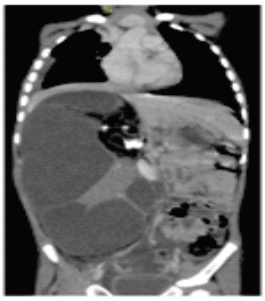

Fig. 3F

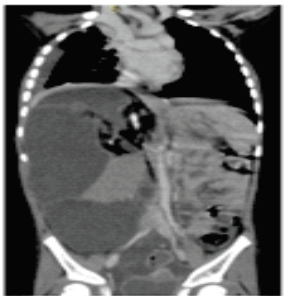

Fig. 3G

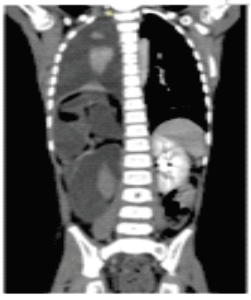

Fig. 3H

Axial Contrast Enhanced CT Scan of the Abdomen Showed a Large Well-Defined, Multi-Septated, Solid-Cystic Lesion (Predominantly Cystic with Enhancing Solid Component) measuring 10.9x8.3x 17.4cms (AP x TR X CC) in the Right Side of Retro Peritoneum (Fig. 3a-3d) (Fig. 3c-Blue Circle) Coronal Contrast Enhanced Scan Showed Dense Calcification and Fat Component within the Lesion (Fig. 3f) (Blue Thick Arrow) The Right Adrenal Gland was Not Seen Separately, from this Lesion (Fig. 3e-3h)

Retro peritoneal Teratoma (RPT)are the third most common primary retro peritoneal tumors in the pediatric population, after neuroblastoma and Wilms tumor 6 The majority of these tumors are benign and para renal in location. They are more common on the left side, though, occasionally, lesions can be bilateral[7,8]. They generally occur in the first 6 months of life and the occurrence of primary RPT is rare in adults, with only a few cases reported in medical literature[6]. RPT's often remain asymptomatic due to their location and become large, by the time, they are diagnosed[9]. Patients typically present with abdominal distension or a palpable mass, like in our case. The location of this tumor would coincide with the symptoms, such as vomiting, constipation, lumbar back pain, abdominal distention and edema. RPT may present with chemical peritonitis following rupture of the cyst, secondary infection (abscess formation) and can undergo malignant transformation. These tumors are also prone to infection[10]. Imaging in the form of ultrasound and cross-sectional imaging, such as Computed Tomography scan, are crucial, in order, to

arrive at a particular diagnosis or differential diagnosis, as CT provides better resolution of soft tissues as compared to USG and, can identify features, indicating malignancy (20, 23)[11,12]. Presence of fat and calcification, within the RPT, clinches the diagnosis[13]. This could be of great benefit to the clinician. However, surgical excision of the tumor has to be done, for definitive histopathological diagnosis[11]. Retro peritoneal Teratoma typically presents with abdominal distension and/or a palpable lump in the abdomen. Serum alfa-fetoprotein is a good biochemical marker, for diagnosing RPT and post-operatively, in assessing tumour recurrence[5,14]. In imaging, the diagnosis can be specifically, made, by, Computed Tomography (CT) scan of the abdomen and is confirmed by histopathologic examination of the rejected specimen. Complete surgical removal of tumour, irrespective, of its size, is the single most important factor in prognosis of RPT's[6,15].

CONCLUSION

The incidence of retro peritoneal teratoma in females is twice than that, in males, as in this case. Early diagnosis and complete resection are the mainstay of treatment for the primary RPT in children. Majority of them are benign. The prognosis is usually favourable and recurrence can be followed with tumours markers such as AFP.

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