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Supra Renal Teratoma- A Case Report

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abstract

Teratomas are neoplasm originating from tot potent cells- developing from more than one germ cell layers-ectoderm, mesoderm, endoderm. Primary retroperitoneal teratomas are rare and suprarenal location is extremely rare. They usually present in childhood with non specific symptoms and abdominal lump. Complete surgical excision cures the benign neoplasm but immature teratoma may need adjuvant chemo/radiotherapy. It needs careful follow up as even mature teratomas may take a malignant course. Here in we discuss a case of Benign cystic teratoma in supra renal location in a 15months old female child.

Key words: *Infant, Teratoma, Totipotent, Suprarenal.*

Introduction

Teratomas (Greek *teras*, meaning "monster," and *-oma*, a suffix denoting a tumor) are the most common form of germ cell tumors present in neonates and infants ^[1,2]. They are encapsulated neoplasm originating from totipotential cells, composed of different types of parenchymal tissues of varying degrees of differentiation derived from more than one germ cell layer (ectoderm, mesoderm, endoderm)^[3,4,5]. The Incidence of germ cell tumor is about 0.9/100,000 population of which teratoma constitutes the

majority of neoplasm^[6]. They are mostly gonadal, however they can be found in extra gonadal sites such as sacro-coccygeal, presacral, mediastinal, pineal, bowel, tongue and retro-peritoneum. Primary extra-gonadal teratomas are common in neonates and infants, where as in adults; particularly in males it is secondary to gonadal tumors ^[7]. The most common site of extra gonadal teratoma in children is sacro-coccygeal or pre sacral region. Retro peritoneal supra renal location is extremely rare and very few cases have been reported so far.

They usually present as an asymptomatic abdominal mass. Plain X-ray, USG followed by CECT /MRI of abdomen are the investigations to detect the nature and extent of the mass. It can also be detected antenatally by fetal ultra sound. Serum alpha fetoprotein and β -Hcg levels may be elevated and are useful markers for follow ups. Complete surgical excision is the treatment of choice in mature teratoma. Histopathology differentiates benign mature from immature and malignant teratomas.

Case History

A 15 month old female child first order born from non consanguineous marriage was presented to us with complain of a swelling in left side of abdomen and low grade fever for last 20 days. There were no other associated symptoms. The swelling was gradually increasing in size for which parents sought medical attention and was referred to our hospital with a provisional diagnosis Neurblastoma. On examination there was a firm non tender swelling approximately 9cm x10cm in left flank. There was no other significant physical finding.

All routine investigations to look for the causes of fever were within normal limits. Her 24 hr urinary VMA was normal. Serum α -feto protein and HCG were not raised. USG and CECT of abdomen revealed a left supra renal mass measuring 92x75x105mm, predominantly cystic with calcifications, displacing the left kidney, pancreas, splenic vein and left renal vein. (Figure-1)

On laparotomy a thick walled cyst of size 8x6x5 cm was found in left supra renal region overlying IVC/Descending Aorta, with left Supra renal vein overlying the mass. (Figure-2) The mass was displacing and rotating the left kidney and also displacing colon/pancreas to right and upwards. The mass was separated from the adjacent structures and adrenal tissues and complete excision was performed. On gross examination, mass was multilobulated. (Figure-3) The cyst was filled with a turbid fluid and a pultaceous material along with hair strands and teeth. (Figure-4) The

histopathological diagnosis was benign cystic teratoma. She had an uneventful recovery and was doing well on follow ups after 6 months.

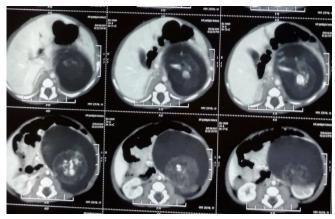


Figure-1: CECT showing Suprarenal mass



Figure-2: Operative photograph of the mass



Figure-3: Excised multilobulated mass



Figure-4: Cut section of the mass

Discussion

Teratomas are neoplasm developing from pluripotent cells of either germ cell or embryonic origin ^[8], deriving tissues from all the germ cell layers. Those arising from germ cells are found to be involving gonads, usually seen in adults and older children. Teratomas arising from embryonic cells are located in extra gonadal sites, particularly in pediatric age group.

Many extra-gonadal sites have been reported, including the tongue [9], face, cranium [2], the large bowel [10], mediastinum, retro peritoneum, sacro-coccygeal and presacral region [11]. Most common sites of teratomas in neonates are the presacral sacro-coccygeal and regions. Retroperitoneal teratomas in supra renal location are extremely rare (3.5-4% of all germ cell tumors.)^[12], seen more in females than in males ^{[6,} ^{13]}. Retroperitoneal teratomas are often located near the upper pole of the kidney with preponderance on the left side [13]. Our patient had a left sided supra renal cystic mass and was a female infant of 15 months age.

Histologically, Teratoma can be classified as mature, immature. Mature teratoma is benign and cystic and are classically called dermoid cyst. The cyst is mainly composed of squamous epithelium, fat, tooth and hair. Dermoid is a benign neoplasm with malignant potential [7]. Whereas immature

teratomas are solid mass composed of undifferentiated tissues with more chance of malignant transformation.

Retroperitoneal teratomas are mostly asymptomatic and as the tumor grows, may compress the surrounding structures resulting in colicky abdominal pain, intestinal obstruction or peritonitis. The usual presentation is a palpable mass or distension of abdomen [14]. Our patient presented with abdominal mass on left side with low grade fever, no other pressure symptoms observed.

The diagnosis of a retroperitoneal teratoma cannot be made on clinical grounds alone. Ultrasound is the first modality of investigation to identify the nature of the mass, whether cystic, solid or mixed and can also differentiate fluid, fat, sebum and calcifications inside cystic mass^[15]. CT abdomen is superior to USG in distinguishing fluid, fat, sebum or calcification. Detection of sebum as fatty portion with dependent fluid in horizontal interface is characteristic of a teratoma [15, 16]. MRI is considered as a superior modality of investigation which delineates proper anatomy ,differentiates solid from cystic ,detects calcification and tumor invasion to adjacent tissues [17, 18]. Angiography, though not commonly done for evaluation may be used for assessing vascularity of tumor as well as infiltration to adjacent organs and encasement of vessels ^[19]. A postoperative biopsy of the specimen is often required for a definitive diagnosis. In children, it should be differentiated from, neuroblastoma, lipomatous tumors and other adrenal tumors Increased serum alpha-feto protein level indicates recurrence of tumor in follow up^[14]. After ultrasonogram of abdomen, we did CECT abdomen to delineate the mass. Alpha feto protein and Beta HCG levels were normal in our case. Possibility of neuroblastoma was ruled out as 24 hrs urinary VMA level was not raised in our case.

Chances of malignancy are uncommon in retroperitoneal teratoma and non mutilating excision is possible, hence complete surgical excision is the treatment of choice [14, 20]. Prognosis

is generally good after complete resection of the tumor, although malignant recurrence were also reported in few cases. Hence careful follow up is needed. In the case of malignant or immature teratoma, adjuvant therapy, such as chemotherapy, radiotherapy, or concurrent chemo-radiotherapy, will also be necessary, provided that the primary tumor has been completely removed [21].

In our case the patient was a 15 month old child with retroperitoneal teratoma, which is rare in this age group. It was benign in nature and complete excision of the tumor was done successfully. Post operative period was uneventful and follow up after six months shows no recurrence.

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