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Large Mature (Benign) Cystic Retroperitoneal Teratoma Encircled by IVC in Young Male – Challenge in Complete Surgical Resection.

KEYWORDS: Retroperitoneum, Benign cystic Teratoma, Surgical challenge, Complete resection.

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ABSTRACT *Retroperitoneum is a rare site for extragonadal teratomas. Less than 4% occurs in children and 90% its benign. May mimic as mesenteric cysts in presentation. Ultrasound and CT scan plays important role in diagnosis. Here we are presenting a case of 10 years male who had large painless abdominal lump and on radiological evaluation diagnosed as large mature teratoma . Exploratory laparotomy was done tumor was found encircled with IVC. Complete resection of the tumor was achieved and after histopathological examination suggested mature teratoma. Patient was discharged after uneventful recovery. Sometimes large size and anatomical location and surrounding vital structures produce challenge for complete surgical resection and good prognosis.*

INTRODUCTION

Primary benign extragonadal teratomas (dermoid tumors) are rare non seminomatous tumors derived from more than one of the three germ cell layer (ectoderm , mesoderm, endoderm) [1]

Historically, teratomas world attributed to demons, sexual misconduct and abnormal fertilization. As with teratogenic, the name derives from the Greek word teras, meaning 'monster'.More commonly dermoid tumors originates from midline structures (paraxial) and most common sites are testes and ovaries. Rarely they also found at extragonadal sites such as intracranial, cervical, mediastinal, retroperitoneal, and sacrococcygeal regions [2]Retro- peritoneum is rare site of dermoid tumors (3.5 to 4% of all germ cell tumors in children)[3] more commonly dermoid tumors having female predominance but in some studies it is found 2:1 , male : female ratio.[4]

1%–2.5% of germ cell tumors originate in an extragonadal location. These tumors are believed to arise from aberrant primordial germ cell rests that are due to (a) faulty migration of germ cells from the yolk sac or endo- derm to the urogenital ridge or (b) germ cells dis- tributed physiologically to liver, bone marrow, and brain [5].Due to its enormous size and close approximation with the intraabdominal vital structures, these tumours pose a challenge to the surgeon, at the same time giving a good result after complete excision.

We are presenting a case of a 10 years male child in which large retroperitoneal dermoid tumor diagnosed and confirmed by CT scan and surgically excised completely Even challenged byencircled IVC.

CASE REPORT

A 10 years old male child presented with a slowly growing painless lump at right hypochondrium since one year. He had dull aching intermittent pain since 6 months. There were no associated symptoms in spite of the size of the lump. Clinical examination revealed a large mass of variegated (soft and firm Consistency occupying the entire right side of the abdomen. =

Lump was moving with respiration and dull on percussion. There was no history of bowel and bladder disturbances. His general physical examination revealed as an afebrile, active child with no jaundice, pallor, cyanosis, clubbing and lymphadenopathy. Abdominal examination revealed a soft, non-tender, with a soft palpable mass over the right hypochondrium. No organomegaly. Bowel sounds were normal. Rest of the physical examination was unremarkable. Laboratory investigations are within normal limits.

Plain X ray abdomen showed calcification over L-1, L-2 vertebrae.

On ultrasound abdomen revealed a mixed echoic complex well defined mass in the abdomen with solid and cystic areas with septations and lobulations. right kidney was clearly seen not involved with the tumor (Figure 1). In view of calcification and US findings diagnosis of Dermoid tumor was made. Serum alpha fetoprotein level was found within normal limit. CT scan abdomen revealed a large 18 cm x10 cm heterogeneous mass with cystic , solid , calcific and fatty components (Figure-2). It was anterior to right kidney ,duodenum and pancreas abutting bowel loops. Most noticeable finding was its location which was right para aortic and IVC was in close proximity of the tumor. Exploratory laparotomy was planned and a large multilobulated mass was present . IVC was encircling the tumor and multiple branching were draining the tumor. In fact tumor was in between IVC and aorta .IVC was dissected carefully and all the branches ligated. Tumor was freed from all around and taken out.

On gross examination tumor was of size 20 cm x12cm x 8 cm. on cut section cystic portion of tumor was filled with the sebum, proteinaceous fluid and greasy material along with the hair within it (Figure 3). Areas of calcification compatible with bone were also noted. Histological examina-

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tion there was sheets of squamous epithelium, hairs and bony elements (Figure 4). Features suggestive of mature cystic teratoma benign in nature. Post operative recovery was uneventful needed one unit whole blood transfusion.

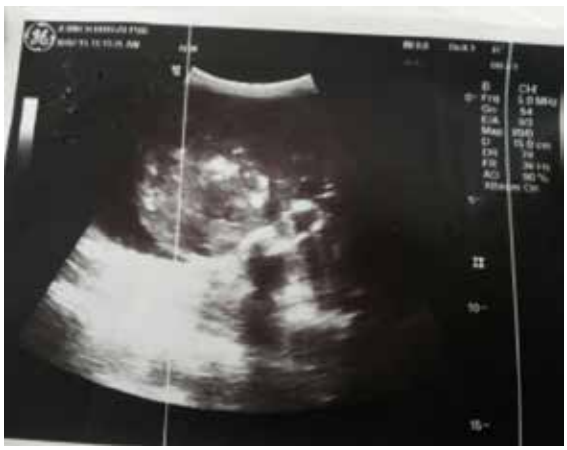

Figure 1- Ultrasound shows well defined mass in the abdomen with solid and cystic areas with septations and lobulations.

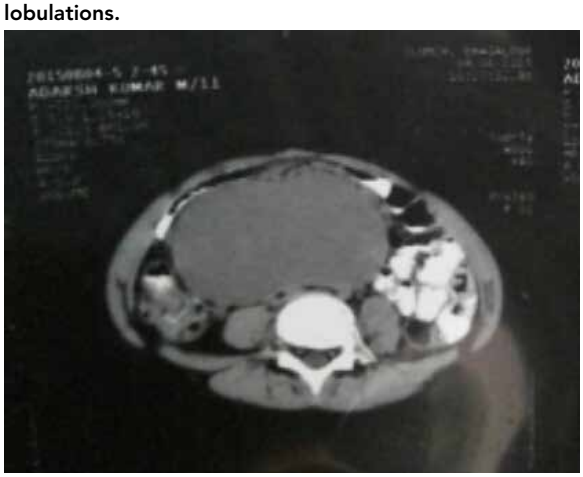

Figure-2-CT scan abdomen revealed a large 18 cm x10 cm heterogeneous mass with cystic, solid, calcific and fatty components.

Figure 3- Cut section shows cystic portion of tumor was filled with the sebum, proteinaceous fluid and greasy material along with the hair.

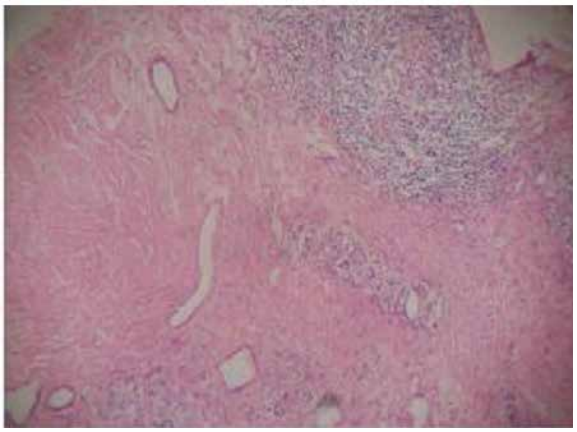

Figure 4- Histology sheets of squamous epithelium, hairs and bony elements.

DISCUSSION

Teratomas are the tumors that arise from the pleuripotent embryonic cells[6] of the germinal layers and will have elements of Ectoderm, Mesoderm and Endoderm.[7, 8]Teratomas generally ariss from uncontrolled proliferation of pleuripotentcells , germ cells and embryonal cells. Teratomas arising from germ cell sources can be congenital or acquired and are usually found in gonads (testes and ovaries). But teratomas of embryonic cell sources are always congenital and are usually found in extragonadal locations, such as intracranial, cervical, retroperitoneal, mediastinal, and sacrococcygeal sites [9, 10]Teratomas can occur anywhere in the body or in any organs but mostly found in the paraxial and midline locations.[7,8] Retroperitoneal teratomas are the least and gonadal teratomas are highest in their occurrence; other sites being sacrococcigeal, near pineal body, near thyroid and anterior mediastinum.[7] Only 3.5 to 4% of all teratomas occur in the children. Among retroperitoneal teratomas, over one-half occur in the pediatric age group, and about 90% are benign.[6] Malignancy is uncommon in retroperitoneal teratomas except tumor which presents antenatally and diagnosed at birth. The chance of malignant transformation in extra gonadal teratomas occurring in infancy and childhood is 10% whereas 26% in the adults.[7,11]Teratoma can be benign or malignant, and benign teratoma can be either mature or immature. Mature teratoma (dermoid cyst) contains well- differentiated tissues from at least two germ cell layers. Ectodermal layers are seen in all, mesodermal layers in 90% of lesions, and endodermal layers in the majority of lesions. Mature teratomas are predominantly cystic. Calcification (toothlike or well defined) and fat can be seen in 56% and 93% of cases, respectively [12] Patients with teratomas are usually asymptomatic due to enough retroperitoneal space[13] and may become symptomatic due compression on surrounding structures7 like colon, kidney, pancreas stomach or the vessels. Incidence of retroperitoneal teratomas is more on left and twice in female than male.[7]

In about 53 to 62% of teratomas, calcification is seen at the rim of the tumor.[6] Calcification, bone formation, and teeth on Ultra sound or X-ray, are manifestations of benign teratoma.[6,14] However calcification cannot be considered indication of benign tumor since 12.5% of calcified tumors are malignant.[6,11] Patient can be evaluated by X-ray of the abdomen and contrast CT. Some authors recommend angiography, inferior venacavography and needle biopsy for accurate diagnosis. Alfa-feto protein (AFP) can be used as

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biomarker for diagnosis as well as for recurrence. Main aim of evaluation of the benign teratoma is to diagnose malignant change which is reported in about 0.25 to 0.8% cases.[12] Prognosis is good if the cyst wall is not penetrated. Macroscopically teratomas are of two types: - A) Cystic usually benign and B) Solid teratomas are generally malignant and formed of various tissues like fibrous, fatty, bones and cartilage and also may have immature embryonic tissue. Complete surgical excision of the tumor tissue is most important factor for cure of the patient.Tissue adherence which can be observed in both benign and malignant form of teratomas, requires extended surgery for removal of adhered organ for the completeness of surgery and good prognosis.

Table 1: Differential diagnoses of retroperitoneal cysts.

Female	Male
Lymphangioma	Lymphangioma
Cystic teratoma	Cystic teratoma
Cystic haematoma	Cystic haematoma
Cystic mesothelioma	Cystic mesothelioma
Bronchogenic cyst	Bronchogenic cyst
Epidermoid cyst	Epidermoid cyst
Tailgut cyst	Tailgut cyst
Mesenteric cyst	Mesenteric cyst
Pseudocyst (non-pancreatic)	Pseudocyst (non-pancreatic)
Pseudomyxoma peritonei	Pseudomyxoma peritonei
Urinoma	Urinoma
Lymphocele	Lymphocele
Endosalpingiosis	
Mullerian cyst	
Vulval cyst	
Parovaian cyst	
Vaginal cyst	
Paraurethral cyst	
Mucinous cystadenoma	

Several imaging modalities elucidate different characteristics of a teratoma. For example, ultrasound can identify the cystic, solid or complex components of the tumor · The cystic portion may be further differentiated into sebum, non-fat fluid and structures resembling fetal parts. However, ultrasound has its limitations as Davidson et al found that ultrasound poorly identified fat and calcifications, which are suggestive of teratoma.

CT has several advantages over ultrasound. First, it gives more specific information on the fat, proteinaceous fluid and calcification components of the teratoma through Hounsfield units, which allows quantitative comparison of substances of different radiodensities. The presence of fatty portions of the tumor in the horizontal interface with dependent fluid, which probably represents sebum, is virtually pathognomonic of a teratoma[11]

Magnetic resonance imaging (MRI) and angiography offer other benefits. MRI is superior to both ultrasound and CT in defining the anatomical relationship of the teratoma with adjacent organs and local tumor spread. MRI can also distinguish fluid, fat, calcium and soft tissue elements, as well as predict resectability and evaluate recurrence

In our case clinical diagnosis was mesenteric cyst but on radiograph showed calcification and on ultrasound cystic and solid component given clue about teratoma and after CT scan it became confirmed.

CONCLUSION

Retroperitoneal teratomas are uncommon tumors in children and majority of the lesions are benign. X-ray findings of calcification/ bone/ teeth are pathognomonic. Ultrasonography and CT scan are useful to delineate the extent of the tumor. Despite extensive local spread, the lesions are amenable to curative surgical excision.

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