Retroperitoneal teratoma in 4 months old girl: Radiology and pathology correlation.

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Abstract

Retroperitoneal teratoma comprises 3.5–4% of all germ cell tumors in children and 1–11% of primary retroperitoneal neoplasms. The imaging findings are distinctive. The present report is a case of a 4-month-old girl in whom a retroperitoneal teratoma was found to have classical appearance on imaging. The tumor was resected and histopathological examination confirmed the diagnosis of immature, high grade, teratoma.

Key words: Retroperitoneum, teratoma, radiology and pathology correlation.

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Introduction

Germ cell tumors are congenital tumors containing derivatives of all the three germ layers. They are frequently seen in gonads. [1] Their occurrence in extragonadal sites are not unknown. [1,2,3] The involvement of extragonadal sites in decreasing order of frequency are mediastinum, sacrococcygeal region, retroperitoneum, and pineal gland. [2,3,4] Extragonadal primary teratomas are usually encountered in infants and children [1,2,3,4]. The present report is a case of a 4-month-old girl in whom a retroperitoneal teratoma was found to have classical appearance on imaging. The tumor was resected and histopathological examination confirmed the diagnosis of immature, high grade, teratoma.

Case report

A 4-month-old girl presented to the emergency department at our institute with poor feeding for 2 days duration and abdominal distension since 2 months ago. There was no history of fever, weight loss, bowel or urinary complaints. No past medical or surgical history were documented. On examination, the abdomen was distended. A large firm mass was palpable in the center of the abdomen extending into the left hypochondrium measuring about 10 x 12 cm in maximum dimensions. No tenderness on palpation or overlying skin changes. The rest of general examination was unremarkable. Routine blood tests and

urinalysis were all within normal limits. The serum levels of alpha-fetoprotein (AFP) was raised measuring 2317.



Figure 1. Frontal abdominal radiograph showed large midline abdominal opacity associated with internal areas internal areas of calcification. Mass effect is also noted marked by displacement of the bowel loops peripherally.

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A plain abdominal radiograph was performed at that time in frontal projection and showed a well defined round radio-dense mass seen projecting at the center of the abdomen and measuring about 9 x 12 cm in maximum dimensions associated with internal areas of calcification. Mass effect is also evident marked by displacement of the bowel loops peripherally [Figure 1].

Ultrasound examination of the abdomen was then requested and demonstrates a large heterogeneous and complex mass located within the middle aspect of the abdomen associated with an areas of cystic components. Doppler interrogated images were also obtained and reveled minimal internal vascularity [Fig. 2].

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Figure 2: Three selected ultrasound images of the upper abdomen demonstrate a large heterogeneous and complex mass located within the middle aspect of the abdomen associated with an areas of cystic components. Doppler interrogated image reveled minimal internal vascularity.

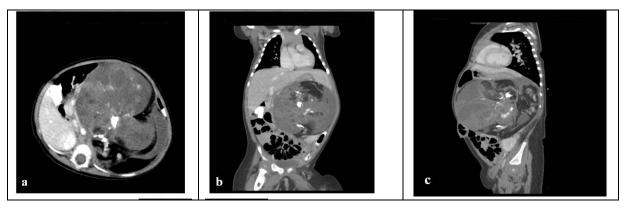


Figure 3: 3 Selected axial, coronal, and sagittal CT scan images of the abdomen and pelvis demonstrates a large retroperitoneal complex and heterogeneous mass occupying predominantly the left suprarenal region. internal areas of calcification, fatty tissue, and cystic contents are also seen.

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Computed tomography (CT) scan of the abdomen and pelvis was performed for further evaluation of the previously described radiograph and ultrasound abnormalities after administration of intra venous (IV) contrast medium. Coronal and sagittal reformatted images were also obtained. The CT scan demonstrates a large retroperitoneal complex and heterogeneous mass occupying predominantly the left suprarenal region. Its measures about 11.7 x 10.4 x 8.8 cm in anterior-posterior, width, and carino-

caudal maximum dimensions respectively. Internal areas of calcification, fatty tissue, and cystic contents are also seen. The mass displaced the aorta and IVC to the contralateral right side and the left kidney caudally to the left iliac fossa [figure 3]. There was no significant enlarged intra abdominal or pelvic lymph nodes. The rest of abdominal and pelvic organs are within normal limits.

Exploratory transverse laparotomy was performed for this

girl and revealed a large complex solid and cystic mass. No invasion to the aorta or inferior vena cava was reported. No involvement of the ovaries were documented. The entire tumor was then excised and send to the pathology department for further analysis. The solid component of the mass contain tooth formation, hemorrhagic areas and fatty content [Fig. 4].

Microscopically, the mass demonstrates some areas of multiple immature neural element consistent with immature, high grade teratoma. The remaining part of the tumor consists of multiple mature elements which include cartilage keratinized epithelium and tooth formation grossly [Figure 5a,b,c].

The final diagnosis was made to be immature high grade teratoma of the retroperitonium. The patient was discharge from the hospital in good and stable condition. Acetamenophin (70 mg orally every 6 hours) was adminstered to the patient for one week. Serial follow up for AFP reveled significant reduction measuring 96, 27, and 11 after about 5 months duration.



Figure 4: Gross image of the resected tumor showed a large complex cystic and solid mass measuring about 15 x 11 x 9 cm and weighting 592 grams. The solid areas contained tooth formation, hemorrhage, fatty tissue, and mucinous components.

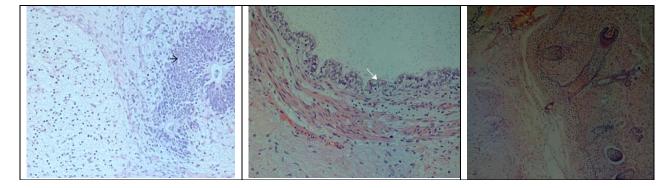


Figure 5 a,b, and c: 3 selected histology images were obtained. (a) high grade immature neuroepithelium is seen (arrow). (b) normal respiratory epithelium (arrow). (c) benign squamous epithelium with hair follicle and hair shaft (arrow).

Discussion

Germ cell tumors contain the derivatives of all the three germ layers. ^[1,2,3,4] The migratory capacity of germ cells may account for the anatomic variety seen with these tumors and this explains the occurrence of teratoma in the gonads and the midline structures. ^[1,2,3,4] Retroperitoneal teratoma comprises 3.5–4% of all germ cell tumors in children and 1–11% of primary retroperitoneal neoplasms [5,6]. Patients usually present with abdominal distension or a palpable mass like our case. Occasionally, the tumor is present antenatally and diagnosed at birth, these neonatal teratomas have a higher incidence of malignancy than those in older children [7].

Ultrasound of the abdomen is usually the first imaging

modality employed in the evaluation of any pediatric abdominal mass. However, in retroperitoneal teratoma, xray may demonstrate calcification as in our case or formed bony components such as teeth and phalanges (which are pathogenomic). Schey and Vesley have recommended only a plain abdominal X-ray and excision of tumor if the characteristic calcification is demonstrated [8]. Lack and Travis have also reported that the presence of bones or teeth on an x-ray was the most helpful in establishing a diagnosis [9]. CT-scan is useful to delineate the extent of the disease in retroperitoneum and its relationship to major vessels. However, CT-scan can overestimate the degree of tumor adherence to adjacent structures than actually seen on exploration [10]. Therefore, CT-scan findings should not prevent surgical exploration of the tumor and even bilateral lesions are amenable to complete removal. Hayasaka and Yamada have reported internal homogeneity, fat density, cyst formation, and calcification to be important predictors of a benign retroperitoneal tumor on CT [11]. Some authors have even advocated angiography, inferior venacavography, and needle biopsy for the accurate diagnosis of these tumors [12]. Among haematological investigations, serum alpha feto-protein level is a good indicator for diagnosis and assessing the recurrence of tumor.

Complete excision of the tumor offers the best chance of cure [10]. Malignancy is uncommon in retroperitoneal teratoma hence nonmutilating excision is possible and should be attempted even in lesions involving both sides of abdomen [13]. The most important aspect of the excision is to dissect the tumor from renal and other major vessels, which are invariable stretched out over the lesion.

Prognosis is generally good and curative if the tumor is completely removed. Lastly the single most important factor in prognosis in Retroperitoneal teratoma is complete removal which must be tried in every case of retroperitoneal teratoma irrespective if its size.

Conclusion

Retroperitoneal teratoma is a uncommon entity which has distinctive imaging findings. Majority of the lesions are benign. Plain radiographs findings of calcifications, bone, or teeth are pathognomonic. Ultrasonography and CT are useful tools for further evaluation of the extent of the lesion. The lesions are mostly amenable for curative surgical excision. Prognosis is generally good and recurrence can be monitored with tumor marker like AFP.

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Conflict of interest: Non

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