

Retroperitoneal teratoma in child A case report and literature review

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Abstract

Of all primary retroperitoneal teratomas, less than four percent occur in children and 90% are benign. we report a case of mature retroperitoneal teratoma in a 4 months year old girl. She presented with right side, slow growing, abdominal mass since 1 month; Ultrasound of abdomen found Large right suprarenal mass, of multiloculated cysts, containing stalls of variable size and thick septa. Computed tomography (CT) assessed the vascular and visceral anatomical relationships.

Patient underwent surgery, the tumor was separated from all adherent tissue with complete resection. Without vascular and organs injuries. Histopathological examination of tumor was suggestive of mature teratoma. Post operative recovery was favorable and patient was discharged from the department. The oncologic outcome of retroperitoneal teratoma is positive especially with complete resection.

Keywords: teratome, retroperitoneal, resection, prognosis

Introduction

Retroperitoneal teratomas are uncommon lesions in children. The majority are benign but complete excision is necessary for cure [1].

Malignancy occurs in ~15% of cases. Of the benign teratomas, 25% contain immature elements [2].

Around 70% of these tumors are known to be diagnosed in infants and 70% are found to occur on the left side of the abdomen [2].

these tumors can grow to be quite large before signs or symptoms are detected.

Because of the location and massive size, major vessel anatomy may be distorted. Vascular injuries are well-recognized surgical complications with urgent repair [2].

Cas report

Girl aged 4 months old years, without significant past medical history, presented with, abdominal mass since 1 month. At physical examination, she had no abdominal tenderness. there was a mass measuring (13 cm × 10 cm), firm, on the right lumbar and iliac region, normal overlying skin, not fixed to the underlying muscles; the surface was smooth. Patient had a very good general condition. No family history was reported.

Ultrasound abdomen found Large right suprarenal mass, with multiloculated cysts, containing variable size and thick septa.

Computed tomography (CT) scan of abdomen revealed retroperitoneal tissue process heterodense, containing multiple calcifications, multiloculated zones, and enhanced septa, mesuraing 130 * 100 * 99 mm.

It exerts a mass effect on the ipsilateral kidney with a border of separation. This mass comes into contact with the aortic caval axis. It encompasses the celiac trunk, the superior mesenteric artery and the right renal artery whose remain permeable. (Figure 1)

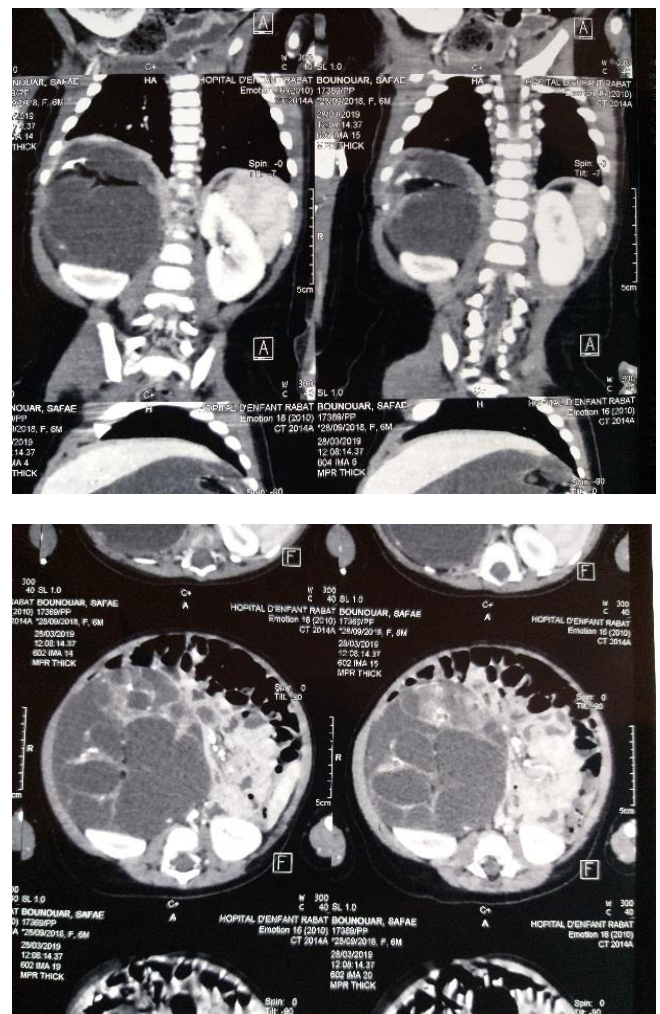


Fig 1: Retroperitoneal tissue process heterodense, containing multiple calcifications, multiloculated zones, and enhanced septa, mesuraing 130 * 100 * 99 mm.

Haematological investigations and serum alpha-fetoprotein assay (AFP) were done to obtain preoperative values, were normals.

The intraoperative findings revealed a large retroperitoneal tumor, with solid and cystic components, occupying mainly the right and central abdomen.

The mass was densely adherent to the adjacent structures, It extended upwards, so as to push the liver,

This mass did not have any adhesions with the pancreas and digestive tract medially.

There was no infiltration of aorta and inferior vena cava.

Right kidney was deformed and intimately attached to the tumor (figure 2).



Fig 2: Right kidney was deformed, and intimately attached to the tumor.

The renal vessels are typically found stretched over the tumor and can be carefully dissected out.

Following the separation of the mass from all adjacent tissues, the mass was completely excised. (Figure 3 and 4)

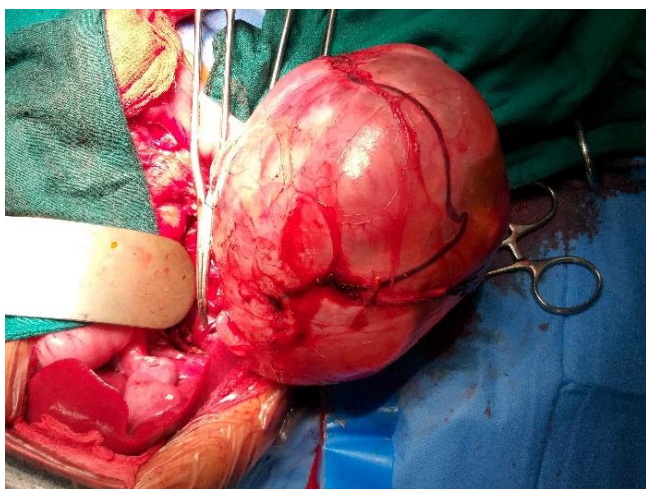


Fig 3: Separation of the mass from adjacent structures.



Fig 4: the tumot was compleley exised.

The mass was sent for histopathology examination, showed feature of mature teratoma.

The post-operative time was uneventful, without complications, and the child was discharged on the 10 th postoperative day on full oral feed.

Discussion

1. Introduction

Retroperitoneal tumors form an important group of neoplasms in infancy and childhood. The two most common retroperitoneal tumors in this age group are neuroblastoma and Wilms' tumor; teratomas are third in order of frequency [3].

accounting for 2–5% of pediatric tumors [4]. and 1–11% of primary retroperitoneal neoplasms [5].

Retroperitoneal teratomas occur outside the pelvis, often in a suprarenal location and more often on the left side than on the right [3].

Due to its enormous size and close approximation with the intraabdominal structures, these tumours pose a challenge to the surgeon, at the same time giving a good result after complete excision [6].

Most of these tumors are benign, but malignancy may be encountered, especially when they are diagnosed antenatally or in the neonatal period.

Majority of these tumors are detected in the pararenal area and are more common on the left side [7].

2. Clinic

Retroperitoneal teratoma is rare and difficult to early diagnose in infancy because of nonspecific signs and symptoms. On time antenatal examination and child health care is necessary in order to get early diagnosis and treatment [2].

These tumors can grow to be quite large before signs or symptoms are detected [2].

They usually present as progressive abdominal distension and palpable abdominal lump. There may be accompanied pain abdomen, vomiting, poor feeding, or constipation [7].

Recommendation on examination in the nearby Maternal and Child Health centers for pregnant woman and young child is necessary. Early detection and diagnosis of teratoma

is the most efficient way to improve the prognosis and reduce medical expenses^[5].

3. Paraclinic

Ultrasound abdomen is usually the first imaging modality employed in the evaluation of any paediatric abdominal mass. However, in RPT, x-ray may demonstrate calcification or formed bony components such as teeth and phalanges (which are pathogenomic)^[6].

Preoperative evaluation of the vascular anatomy is crucial for surgical Planning^[8].

The major retroperitoneal arteries, veins, and organs distorted by the retroperitoneal teratoma were identified through review of original CT/MRI images^[8].

CT-scan is useful to delineate the extent of the disease in retroperitoneum and its relationship to major vessels. However, CT-scan can over estimate the degree of tumor adherence to adjacent structures than actually seen on exploration.

Therefore, CT-scan findings should not prevent surgical exploration of the tumor and even bilateral lesions are amenable to complete removal^[5].

Although benign in nature, teratomas can encase major blood vessels^[7].

authors have even advocated angiography, inferior venacavography, and needle biopsy for the accurate diagnosis of these tumours, which was not done in this case. Among haematological investigations, serum alpha fetoprotein level is a good indicator for diagnosis and assessing the recurrence of tumour^[6].

4. Histopathologic

The presence of germ cell components as well as immature or undifferentiated tissue is the major pathological evidence for malignancy^[4].

The mechanism underlying malignant transformation of mature teratoma remains controversial.

The malignancy rate of teratoma is 7.6% in pediatric patients under 1 year of age, 63.2% in those aged 1–2 years, and remains unchanged in those aged more than 2 years. Unlike children, adults with retroperitoneal teratoma may experience a long-term malignant process^[4].

traditionally Ohno and Kanematsu (1998) propose that all mature teratomas are of malignant potential and should be considered as pre-cancerous tumors.

Tiny areas with malignant lesions should be carefully examined, which may help predict a higher risk of relapse. When adjacent structures are identified in completely differentiated teratoma^[4].

5. Therapeutic intervention

Retroperitoneal teratomas are mostly benign in nature, and complete surgical resection is the mainstay of treatment^[8].

The relatively large tumor size and intimate relationship between tumors and adjacent vasculature and organs may increase surgical difficulty and contribute to increased perioperative complications^[2].

Infancy, lower body weight, and tumors crossing the midline were associated with an increased risk of perioperative complications. Additionally, patients with teratomas that originated from the left retroperitoneum were at marginally significant increased risk of perioperative complications^[8].

Surgical removal of retroperitoneal teratoma is typically

challenging^[8].

Literature presents a contradictory report on the extent of adhesions and hence the ease of dissection in these tumors. Some reports describe teratomas as unattached retroperitoneal tumors, making dissection easy.

More reports, however, describe teratomas as tumors adherent to the adjoining viscera making dissection extremely difficult. Significant adhesions to the stomach, liver, CBD, pancreas, gall bladder, spleen, mesocolon, and diaphragm have been described. Resection of the tumor could result in injury to these structures adding to the morbidity^[9].

6. Extension reports

a. Vessels

As in other series, injuries to any major vessels, including the inferior vena cava, renal vein, portal vein, and superior mesenteric artery occurred in our series.

Even resection of the inferior vena cava has been reported^[8].

The IVC may be absent, occluded, or anteriorly displaced. Accidental ligation of the IVC has been described. Small caval tears may be repaired. It has been suggested that if IVC resection is required, it may be reconstructed using a vascular prosthesis. If resection of the tumor requires the removal of the IVC or if there is uncontrolled hemorrhage from the same, the IVC should be resected as children usually develop adequate collaterals without event^[9].

b. Kidneys

most important aspect of the excision is to remember the close relationship of these tumors with the kidneys. The renal vessels are invariably stretched out. over the lesion, with care however, they can be separated from the teratoma^[10].

Persistence of hypertension in some patients could be attributed to vascular damage to the renal vessels. While the kidneys may be significantly displaced, there is no report of nephrectomy being required during tumor excision.

If inadvertently any major renal is injured, can be repaired^[6].

Nevertheless, it is advisable that consent for nephrectomy be taken before surgical resection^[9].

c. Tumor rupture

In all probability, rupture of a thin-walled cyst and spillage does not adversely affect the long-term outcome. Capsule rupture, staged dissection, and marsupialization before resection have been described^[9].

In large cystic tumors with dense adhesions to surrounding vital structures, avoiding cyst rupture or positive tumor margins may not always be possible. A positive tumor margin does not correlate with recurrence and will not by itself make a case for considering chemotherapy^[9].

7. Prognosis

Prognosis is generally good and curative if the tumor is completely removed^[7].

Most important prognostic factor is complete removal, however as it is possible that histologically mature tumor may take a malignant clinical course. Careful follow up is necessary in these patients^[11].

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 of its size [5].

However, it is difficult when the tumor encases the major vessels [7].

Malignancy is uncommon in retroperitoneal teratomas except endodermal sinus tumors and hence non-mutilating excision is possible and should be attempted even in lesions involving both sides of the abdomen [10].

Lack and Travis have reported guarded prognosis for unresectable lesions, immature teratomas and endodermal sinus tumors. Aggressive chemotherapy has been advocated for the latter lesions, which have a propensity for metastasis [10].

In view of their size, vascular distortion and intimate relation to surrounding structures, retroperitoneal teratomas are not easy to resect, being associated with significant morbidity. Beyond the perioperative period, these tumors have an excellent prognosis [9].

Conclusion

Retroperitoneal teratoma is rare and difficult to early diagnose in infancy because of non specific signs and symptoms.

An acceptable surgical risk underlines the value of en bloc resection of the lesions.

Even extensive local spread and intimate relation to surrounding and stretching adjacent structures, the lesions are amenable to curative surgical excision in the hands of the experienced surgeon.

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