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Case Report

Unusual presentation of retroperitoneal teratoma

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ABSTRACT

Mature teratoma are usually slow growing tumours often silent in nature. We report a 1-year-old dengue positive female child with a sudden increase in size of a previously non palpable abdominal lump. After investigation and exploration, it was found to be a retroperitoneal mature cystic teratoma, which did not show any features of malignant transformation, tumour rupture, haemorrhage or infection. Most retroperitoneal teratomas are benign in nature diagnosed preoperatively but rarely there can be dilemma in establishing the diagnosis. In ambiguous cases intra operative findings or histopathology reveals final confirmatory diagnosis**.**

Keywords: Mature cystic paediatric teratoma, Retroperitoneal teratoma, Acute presentation, Haemorrhage

INTRODUCTION

Mature cystic teratomas are indolent and slowly progressing tumours. Rarely they can present acutely with abscess formation peritonitis secondary to rupture. A sudden increase in size may be attributable to haemorrhage or malignant transformation. In this case report, we discuss one such rare presentation of retroperitoneal teratoma in an infant.[1,2]

CASE REPORT

A 1-year-old female child was admitted for treatment of dengue fever. Apart from the fever spikes, examination findings were unremarkable and the per abdomen findings were not striking. Incidental ultrasound of the abdomen and pelvis was suggestive of a well-defined hyperechoic lesion of size 4.7×2.8 cm indenting the splenic flexure. Over the next few days there was progressive distension of abdomen and a firm lump of size approximately 6×4 cm was felt in the left hypochondrium extending to the left lumbar region (Figure 1). All biochemical, haematological parameters and tumour markers were normal. Contrast enhanced computed tomography of the abdomen and pelvis done prior to exploration showed the presence of a large well defined cystic lesion about 10×7×4 cm in the left half of the abdomen with multiple out pouchings, few thin enhancing septations, an eccentric fat component and calcifications. There was no significant abdominal or pelvic lymphadenopathy (Figure 2a and b).

The patient underwent an emergency exploratory laparotomy. The findings included a large cystic mass which was arising from the retroperitoneum, adherent to the transverse colon and abutting the left renal vessels, left kidney, inferior vena cava, pancreas and spleen. There was no evidence of tumour rupture. The cyst was filled with serous fluid with no evidence of haemorrhage (Figure 3 a and b). The tumour was excised completely and sent for histopathological examination. Post-operative course was uneventful and the patient was discharged 6 days after the surgery. Histopathology of the excised specimen revealed the diagnosis of mature cystic teratoma with no malignant change. The patient is on regular follow up with serum alpha feto protein and ultrasound abdomen and pelvis done annually. The patient is thriving well with no evidence of recurrence.

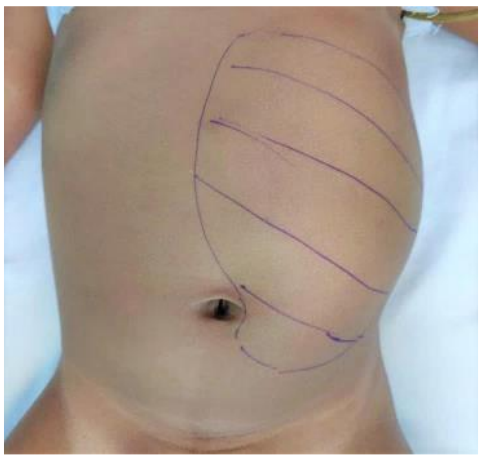

Figure 1: Clinical findings of the patient.

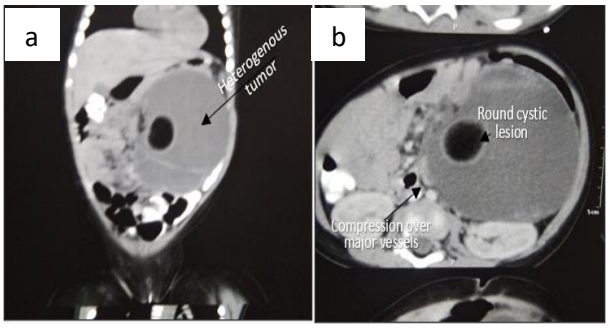

Figure 2 (a and b): Coronal and Axial section of CECT abdomen showing a large heterogeneous retroperitoneal tumor with multiple compartments within, occupying mainly the left suprarenal region, compressing over major vessels.

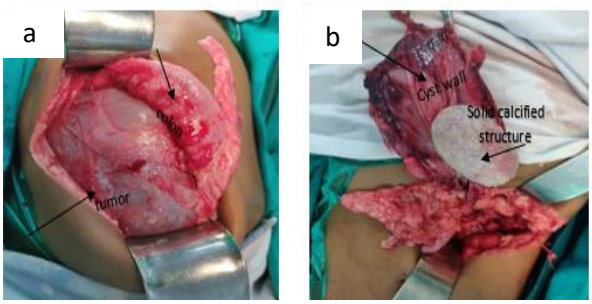

Figure 3: (a) Intraoperative image showing a large retroperitoneal cystic tumor with reflected colon; (b) complete cyst wall reflected with white, round calcified solid content.

DISCUSSION

Primary retroperitoneal teratoma is extremely rare in infancy. In children it accounts for 3.5 to 4% of all germ cell tumours and 1-11 % of primary retroperitoneal neoplasm. Teratomas of embryonic cell origin are congenital and may occur in extragonadal locations namely intra cranial, cervical, retroperitoneal, mediastinal and sacrococcygeal regions.[1,2] These are usually slow growing tumours and could be asymptomatic or present with abdominal distension or a palpable mass. Uncommonly, their acute presentation could be attributable to malignant transformation, abscess formation secondary to infection or acute peritonitis due to rupture. Neonatal teratomas possess higher incidence of malignancy in comparison to older children.[3] In our case, the patient presented with a sudden onset increase in size of a previously non palpable mass with abdominal distension which caused respiratory distress.

Even though intratumoral haemorrhage was one of the differentials, on exploration, no such finding was evident. The cyst fluid was grossly clear. There was no evidence of tumour rupture. Histopathology did not reveal a malignant focus in the excised specimen. Hence, the precise explanation for such a brisk course was not known.

Generalized inflammatory response of the well differentiated secretory epithelium within the cyst could be one of the causes. As the mature tridermic cyst contains tissues from across the body, secretions of the skin and sweat glands, respiratory epithelium, exocrine secretions of the gut mucosa or increase in pancreatic enzyme activity could be plausible explanations for increase in the cyst fluid. Within a short span of hospital stay and a background of continuing inflammatory activity (dengue positive status) our patient's symptoms progressed rapidly.

In emergency situations as in our case, the preoperative planning was precarious and was aided by the contrast enhanced CT scan of the abdomen and pelvis. Important pointers to clench its diagnosis include the visualization of adipose tissue with sebaceous or serous types of fluid.[4] These imaging studies can also display the precise location, morphology and structures abutted by the tumour, which aids in quick but precise, meticulous and complete excision of the tumour without prolongation of anaesthesia time.

The main step of treatment for retroperitoneal teratoma is complete surgical resection. The prognosis depends primarily on the adequacy of surgical resection. The most important aspect would be to dissect the tumour from the major vessels which are invariably stretched over the lesion. Detailed gross as well as microscopic examination of the excised specimen is the gold standard for the diagnosis. Prognosis is excellent after complete surgical excision with an overall 5-year survival rate of nearly 100 %.[5]

CONCLUSION

A sudden onset of symptoms in a teratoma could be due to increased secretory activity of the well differentiated teratoma or a generalized inflammatory response of the lining of the tumour. A high index of suspicion and early

diagnosis is critical for timely management of such unusual presentations.

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