

Retroperitoneal Mature Cystic Teratoma in a 3-month-old Infant: A Case Report and Literature Review

Abstract

Retroperitoneal teratoma (RPT) is an exceedingly rare neoplasm in infancy. We came across a 3-month-old infant with large RPT in the left suprarenal area. The tumor was completely excised, and the patient was successfully discharged. Histopathological examination suggested the diagnosis of mature cystic teratoma.

Keywords: *Alpha-fetoprotein, extragonadal, germ cell tumors, retroperitoneal teratoma*

Introduction

Germ cell tumors (GCTs) are congenital neoplasms comprising of derivatives from the three germ layers. Conventionally, they are divided into gonadal and extragonadal types. Most common sites of GCT are gonadal locations; ovarian or testicular.^[1,2] Although extragonadal GCT is more frequently encountered in children, the majority are located in the sacrococcygeal region.^[3] They are also seen in the mediastinum, especially the anterior mediastinum, head and neck, particularly in the pineal gland. Retroperitoneal teratomas (RPTs) are uncommon neoplasms accounting for 3.5%–4% of GCT in children.^[1,2,4]

Case Report

A 3-month-old male infant presented to emergency department with progressive abdominal distention and poor feeding for 1 month. He was born by normal vaginal delivery at home as the 3rd child of the family, and antenatal ultrasonography was not done. Family history and postnatal period are unremarkable. At admission, he was weighing 4 kg and having mild pallor without icterus, cyanosis, or lymphadenopathy. Abdominal examination revealed a large mass of 10-cm diameter predominantly on the left upper quadrant extending to the right side. There was no local rise of temperature or skin changes over the mass. Mass was firm, nontender, and retroperitoneal in nature. Blood

investigations revealed hemoglobin 9% g and normal routine biochemical tests. Serum beta-human chorionic gonadotropin was normal (0.44 mIU/ml), but serum alpha-fetoprotein (AFP) level was raised, measuring 253.3 ng/dl. Plain abdominal X-ray demonstrated a large, well-defined round radiodense mass predominantly in the left side of abdomen and extending to the right side. The bowel gas shadows were pushed to the periphery [Figure 1]. Abdominal ultrasonography suggested a heterogeneous solid cystic lesion with predominantly cystic component seen in epigastrium and left hypochondrium region. Computed tomography (CT) scan of abdomen showed a retroperitoneal, multiloculated, cystic lesion in the left hypochondrium and epigastrium with enhancing walls, septa, calcification, and fatty tissue within it [Figure 2]. There was no intra-abdominal lymphadenopathy, and other organs were normal. Features were suggestive of RPT in the left hypochondrium and epigastrium.

Laparotomy was done with transverse supraumbilical incision. A large, firm, retroperitoneal mass of 10-cm diameter with smooth outline predominately in the right suprarenal area extending beyond the midline found [Figure 3a]. The tumor was adherent to spleen, stomach, and compressing left kidney from above. The tail of pancreas was flattened and stretched over the mass. The tumor was also adherent to the aorta and inferior

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vena cava, but there was no invasion of these structures or any major blood vessel. There were no significant lymph nodes, and other intra-abdominal organs were normal. The tumor was excised completely [Figure 3b] and sent for histopathological study. Postoperative period was uneventful, and the infant was discharged after 7 days on breastfeeds. Gross pathological examination showed variegated, solid-cystic areas on cut section. Microsections from the cyst wall suggested epidermal lining cells with dermal appendages, and the solid areas revealed mature adipocytes and mature neural tissue with glial cells. No evidence of undifferentiated elements was detected. The microscopic diagnosis was consistent with mature cystic teratoma [Figure 4]. The baby is under follow-up for the last 3 months with normal AFP, and there are no features of recurrence. He is growing and gaining weight according to his age.

Discussion

RPTs constitute 1%–11% of primary retroperitoneal neoplasms in children.^[1,2,4] The most common site of congenital extragonadal teratoma is sacrococcygeal region.^[3,5] Although Morgagni first described a case of RPT in 1761, very few cases have been reported in literature.^[6] Most of these tumors are benign, but

malignancy may be encountered, especially when they are diagnosed antenatally or in the neonatal period.^[1,2,5] Majority of these tumors are detected in the pararenal area and are more common on the left side.^[4,7] They usually present as progressive abdominal distension and palpable abdominal lump. There may be accompanied pain abdomen, vomiting, poor feeding, or constipation. Abdominal radiography may suggest features of calcification, teeth, and bony structures. CT scan is very helpful in showing the extent of disease in retroperitoneum, tumors occupying both sides of the retroperitoneum, and involvement of blood vessels. Although benign in nature, teratomas can encase major blood vessels.^[3] Sometimes CT scan overevaluates the tumor adherence to adjacent structures than practically seen at laparotomy.^[7,8] A preoperative diagnosis of RPT was made in the index case. Calcification was also detected in the CT scan of our patient, which is seen in about 50% of cases of RPT.^[4]

Malignancy is very uncommon in RPTs, and nonmutilating excision is possible.^[1,7,8] The most important part of our dissection was to separate the tumor from the pancreas, kidney, and major blood vessels. Complete surgical



Figure 1: Plain X-ray abdomen showing large abdominal opacity with displacement of bowel loops to the periphery

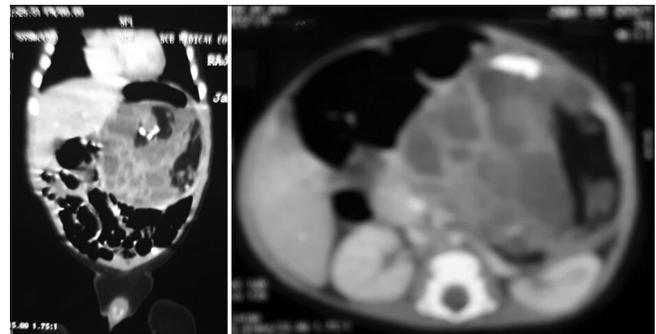


Figure 2: Computed tomography scan of abdomen showing large heterogeneous, multiloculated, retroperitoneal mass predominantly on left suprarenal area and extending to opposite side. There are cystic spaces, fatty tissue, and internal calcification

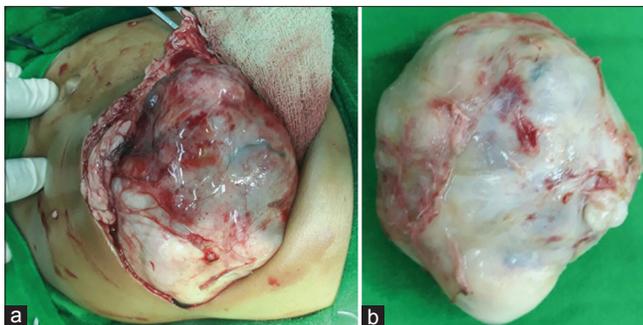


Figure 3: (a) Intraoperative photograph of the retroperitoneal tumor. (b) Gross picture of the resected retroperitoneal teratoma

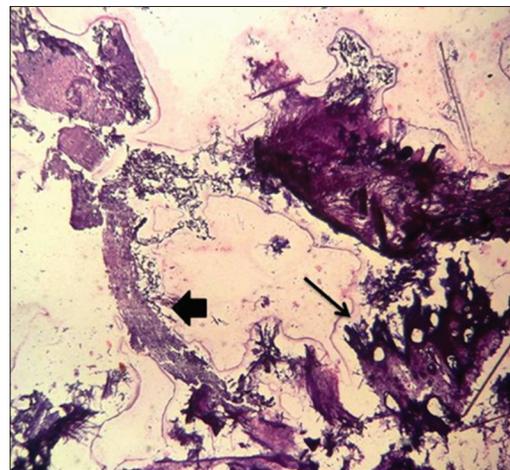


Figure 4: Photomicrograph of teratoma shows epidermal tissue with dermal appendages (long arrow) and mature neural tissue (stout arrow) (H and E, ×100)

excision provides the best chance of cure and is the most important prognostic factor.^[2,8] However, it is difficult when the tumor encases the major vessels. Postoperatively, serum AFP can serve as a reliable tumor marker for monitoring the recurrence.^[1,8] Histopathologically, the index case was diagnosed as mature cystic teratoma. The immature extragonadal teratomas need multimodality therapy. Adjuvant chemotherapy should also be given in case of intraoperative tumor spillage and incomplete removal.^[5]

In conclusion, RPT at the age of 3 months is very unusual. Preoperatively, the diagnosis can be made because of distinctive imaging features. Early diagnosis and complete resection can cure benign mature teratomas in retroperitoneum and prevent recurrence.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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