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Extraordinary large primary retroperitoneal cystic teratoma: An extremely rare neoplasm

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Abstract
Teratomas are the germ cell tumors which comprises of tissues from all the three germ cell layers. Primary retroperitoneal teratoma is a relatively rare tumor in adults. The primary retroperitoneal teratoma constitutes 6–11% of the retroperitoneal tumors. Sixty percent of the retroperitoneal teratomas occur in children less than 15 years.
It occurs more commonly in females than males. The order of frequency for teratoma is ovary, testis, mediastinum and the retroperitoneum at last. Early diagnosis and surgery are the mainstay of treatment. We report a rare case of large primary retroperitoneal teratoma. This is the largest primary retroperitoneal teratoma reported in the literature to our knowledge.

Case report

A 19-year-old male presented with progressively increasing mass and pain in the left side of the abdomen. On physical examination there was a large lobular, firm mass occupying the whole left side of the abdomen reaching up to the right side of the abdomen. On evaluation, the complete blood count, renal function and liver function tests were within normal range. Ultrasonography of the abdomen showed a normal right kidney and large cystic tumor on the left side obscuring the left kidney. On contrast enhanced CT (CECT) scan of the abdomen, there was a large non-enhancing mass of size 36 cm × 25 cm × 16 cm occupying the left pararenal space with anterior and medial displacement of the left kidney (Fig. 1). Both kidneys were normal in size and appearance and the mass was also displacing the right kidney. In the upper part, the mass was anterior to the aorta but in the lower part the mass was lying posterior to the aorta and inferior vena cava. There was calcification on the tumor surface. Tumor markers like AFP, βHCG, LDH were within normal reference range. The patient was planned for surgical excision of the tumor. Total excision of the tumor was performed through a midline incision. Although there was dense adhesion of the tumor to the left kidney, the tumor could be separated leaving the left kidney

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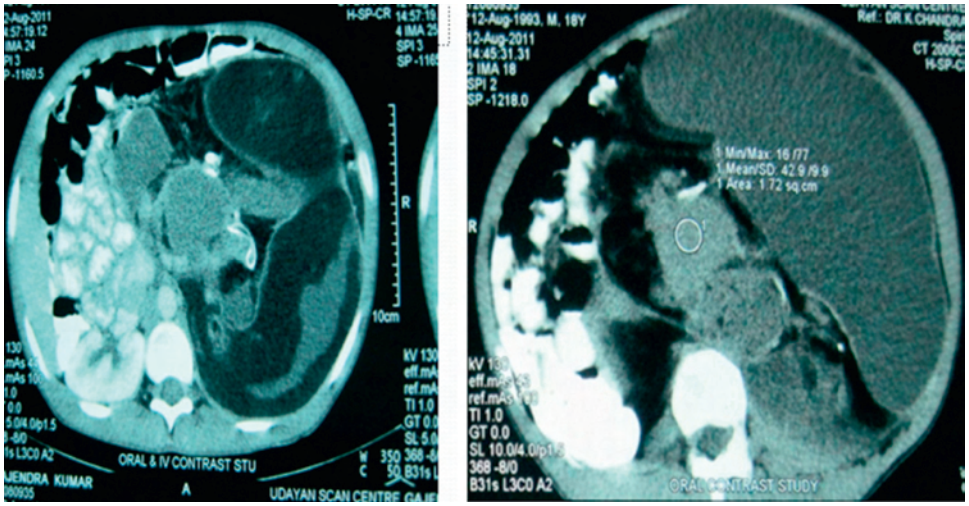

Figure 1 CECT scan of the abdomen showing non-enhancing mass with the presence of calcification.

preserved. The tumor was measured at 42 cm × 29 cm × 17 cm in size and weighted 4200 g (Fig. 2). The cystic portion of the tumor was filled with gray-white greasy sebum, teeth and hairs.

Microscopic examination of the tumor showed a cystic mass with presence of hair, adipose tissue, squamous epithelium, respiratory epithelium, cartilage and chronic inflammatory cell infiltrate (Fig. 3). There were no immature components identified microscopically. There was no evidence of malignancy in these components. The tumor was diagnosed as a mature cystic teratoma.

Discussion

Teratomas are congenital tumors comprising of tissues, arising from pluripotent embryonic cells [1]. The common sites of occurrences for teratomas in decreasing order of frequency are ovary, testis, mediastinum and retroperitoneum [4]. Retroperitoneal teratomas are extremely rare tumors in adults [2]. It occurs commonly at an earlier age with a female predominance.

Retroperitoneal teratomas often remain asymptomatic. It may present as a large abdominal mass with pain abdomen. Radiological manifestations of teratomas are presence of calcification, teeth and

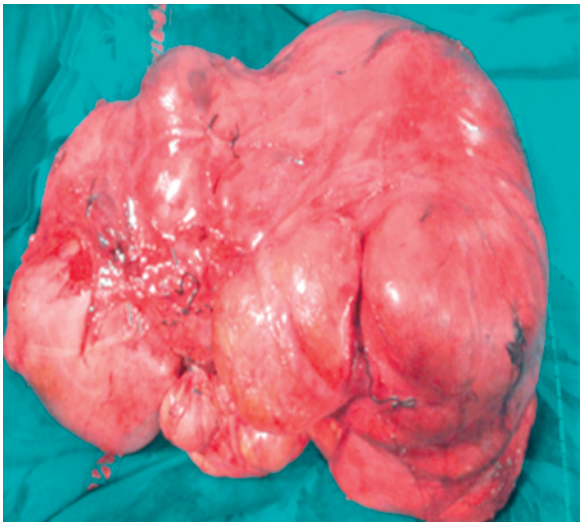

Figure 2 Macroscopic appearance of retroperitoneal tumor.

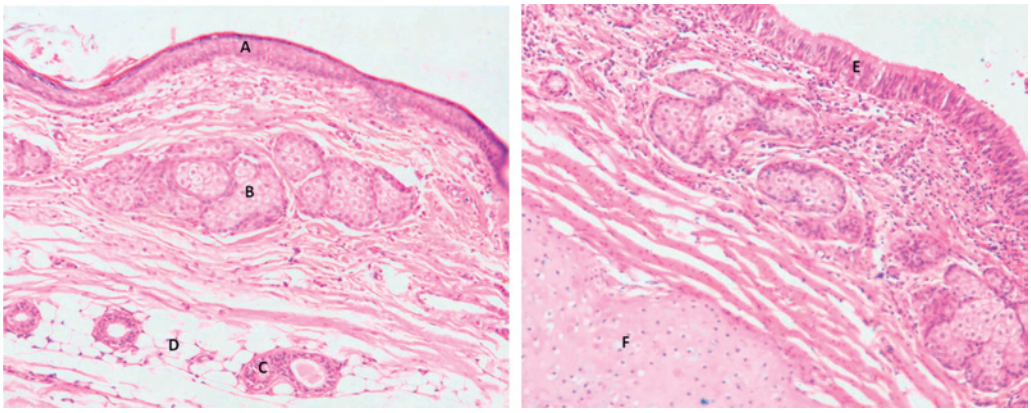

Figure 3 Microscopic examination (H&E section) showing the various components of germ cell layers such as (A) squamous epithelium, (B) sebaceous gland, (C) sweat gland, (D) adipose tissue, (E) respiratory tract epithelium and (F) cartilage.

fat [5]. Calcification is seen in 50–60% cases and is often helpful in making diagnosis preoperatively [6]. Diagnosis can be possible with the help of plain radiography and CECT/MRI abdomen. Management is surgical excision. Chemotherapy and radiotherapy have little role in the management of retroperitoneal teratoma.

Macroscopically, teratomas are classified into two types: (1) cystic and (2) solid. Cystic teratomas are generally benign in nature and solid teratomas are malignant. Malignancy rates are higher in adults (14–26%) than in children (6–7%) [3,6,7]. Since definitive diagnosis is not always possible preoperatively, complete excision is the best treatment for these patients.

Conclusion

Any young patient with a large tumor in the retroperitoneum with the presence of calcification, fat and teeth on radiography should raise the suspicion of teratoma. Since it does not respond to chemotherapy and radiotherapy, surgery is the mainstay of treatment. Close follow-up is required, as histologically mature teratoma may turn malignant.

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