Mature cystic teratoma presenting as suprarenal mass

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DESCRIPTION
A 4-year-old girl was noticed to have abdominal distension by her parents. On evaluation with ultrasound, there was a heterogeneous solid cystic lesion with a predominantly cystic component seen in the left suprarenal region. Contrast enhanced CT (CECT) of the abdomen revealed a large 11×11.5×13 cm multiloculated, hypodense cystic lesion with fat attenuation and long bone within the cystic cavity in the left suprarenal region causing compression and displacement of the left kidney (figure 1). Features were suggestive of teratoma. The endocrine workup for adrenal tumour was normal. Tumour markers which included Alpha Fetoprotein (AFP), Human Chorionic Gonadotropin (HCG) and Lactate Dehydrogenase (LDH) were within normal limits.

The child underwent explorative laparotomy and intraoperatively a large mass was seen replacing and pressing on the left kidney. Dense adhesions were present between tumour and gut loops, spleen and lateral wall. The tumour was also adherent to the aorta and inferior vena cava, but there was no invasion of these structures or any major blood vessel. Mass was well encapsulated and no infiltration of normal renal parenchyma was seen. Complete excision of the left suprarenal mass was done. Left adrenal and bilateral ovaries were normal.

Histopathological examination showed cheesy whitish pultaceous material along with hair, bone and cartilage suggesting structures derived from the different embryonic germ layers. The cyst walls were lined by stratified squamous epithelium. No immature elements were seen, findings were suggestive of mature cystic teratoma (figure 2).

No adrenal tissue or malignant component was seen. After surgery, the child had an uneventful recovery and at 12 months of follow-up, there is no abdominal distension on clinical examination and no recurrent lesion on ultrasonography. Follow-up AFP, HCG and LDH are also normal.

Teratomas are among the most common congenital tumour comprising of tissues arising from pluripotent embryonic cells. They are frequently seen in gonads. The most common site of extragonadal germ cell tumour is the sacrococcygeal region, mediastinum, head and neck, and retroperitoneum. A retroperitoneal teratoma constitutes 1%–11% of primary retroperitoneal neoplasm in children and 3%–4% of all germ cell tumours. The majority of these tumours are found in the pararenal area and are more common on the left side as in index case. There is bimodal presentation, with peak in first 6 months of life and early adulthood. They usually present as progressive abdominal distension and palpable abdominal lump.

Ultrasound of the abdomen is the first imaging modality used, CECT abdomen is the imaging modality of choice and is helpful in showing the extent of disease in retroperitoneum, and involvement of blood vessels. Although benign, teratomas can encase major blood vessels. The serum alphafetoprotein and HCG levels are good indicators


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Figure 1 (A, B) Contrast enhanced CT (CECT) abdomen in axial plane showing well-defined lesion on left suprarenal location with intralesional fatty attenuation (hollow arrow) with bony elements (solid arrow) and long bone (line arrow). (C) CECT abdomen in coronal plane shows displacement of left kidney with extrinsic effect on upper pole calyx (solid arrow).

Figure 2 (A) Cut surface of resected globular soft cystic mass, showing cheesy whitish pultaceous material along with hair (line arrow), bone and cartilage (solid arrow). (B) Histopathological examination (H&E stain) shows Cyst walls were lined by stratified squamous epithelium and shows keratin flakes, skin appendageal structures representing ectodermal components. (C) Histopathology showing glial tissue. (D) Histopathology picture showing mature cartilage with adipose tissue and nerve bundles.
for diagnosis and elevation of either marker may indicate the presence of malignant germ cell elements. However, the final diagnosis is made on pathological examination of resected specimen. Complete resection is the treatment of choice, which is recommended in all cases. Malignancy is uncommon in mature cystic teratoma and the prognosis is favourable with an 80%–100% survival reported after complete surgical excision of the tumour.

Patient’s perspective

Patient’s father: I was told by my doctor that my daughter has a mass in her abdomen, which may or may not be malignant but will require removal by surgery. The surgery went on well. I was happy to know that the tissue report did not show any cancer. Presently, my daughter is doing well and I am very happy that her case was managed very well by the team. I am on regular follow-up as advised by my doctors.

Learning points

► Retroperitoneal mature cystic teratoma usually presents within 6 months of age or in adulthood but may present in young children.
► It generally presents with abdominal distension and early diagnosis can be made by ultrasound and CT scan.
► The diagnosis is confirmed by histopathological examination of resected specimen.

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REFERENCES