CASE REPORT

Retroperitoneal teratoma simulating giant adrenal myelolipoma: a diagnostic puzzle

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SUMMARY

Teratoma is a germ-cell tumour that is common in para-axial regions. Retroperitoneal teratoma is rare and comprise only about 1% of all teratomas. We present a 21-year-old female who presented with a lump in the abdomen. After radiological investigations, she was diagnosed as case of adrenal myelolipoma which on excision turned out histologically to be retroperitoneal teratoma. Our case highlights the limitation of radiological investigations in preoperative diagnosis of teratoma.

BACKGROUND

Teratoma is a germ-cell tumour derived from totipotent cells and made of the cell types of more than one germ-cell layer. Retroperitoneal teratoma is rare and comprises about 1% of all teratomas.¹ Teratomas are common in children, but their prevalence in adults is rare. Adrenal myelolipoma, on the other hand, is a rare, benign, metabolically silent lesion arising in all the age groups. We report a unique case of giant retroperitoneal teratoma in a young pregnant female masquerading as adrenal myelolipoma.

CASE PRESENTATION

A 21-year-old, 6 weeks pregnant female presented with heaviness in the right flank for last 2 years and a lump in upper abdomen for 6 months. There was no abdominal pain, fever, haematuria, urinary symptoms, menstrual or bowel complaints. The patient was thin built. Blood pressure and pulse rate were 114/68 mm Hg and 82/min, respectively. On examination, a large, firm, non-tender lump was palpable occupying almost whole of the abdomen (right hypochondrium, epigastrium, right lumbar, umbilical, right iliac fossa), more on the right side (figure 1).

INVESTIGATIONS

Ultrasonography and contrast-enhanced CT (CECT) abdomen showed a giant hypodense cystic mass lesion (25×15.3×22.9 cm) in right hypochondrium, epigastrium and crossing the midline to left lumbar region with fatty attenuations with calcifications. The mass lesion was displacing the right kidney in right iliac fossa, liver being pushed upwards, pancreas posterosuperiorly, bowel loops laterally and in pelvis (figure 2). Right adrenal gland was not seen separately. Rest of the organs were normal with no evidence of lymphadenopathy

and ascites. In view of its location and fat content on CT scan, possibility of adrenal myelolipoma was kept. Laboratory studies depicted normal myelogram, blood chemistry and urinalysis. The biochemical markers used to identify functioning adrenal tumour, for example, plasma and urinary metanephrines, urinary 17-ketosteroids and overnight dexamethasone suppression tests, were negative.

DIFFERENTIAL DIAGNOSIS

Right-sided giant adrenal myelolipoma was the first possibility. Retroperitoneal mass (lipoma, angiomyolipoma, liposarcoma) can be considered in differential diagnosis.

TREATMENT

Considering the thin built of the patient and huge mass, patient was planned to undergo medical termination of pregnancy (MTP) after informed consent. After MTP, she underwent excision of retroperitoneal mass through subcostal incision (intra-peritoneal, size ~12 cm). Intraoperatively, right renal vein was stretched, elongated and anteriorly placed over the mass (figure 3A). Right kidney was displaced downward to right iliac fossa (figure 3B). The resected mass measured 33×27×15 cm and weighed 5 kg. Grossly, the mass was whitish-yellow in appearance, cystic with smooth surface and bosselated at places (figure 3C). Postoperative course was uneventful.

Sections from the retroperitoneal mass showed all the three components of germ cell layer, that is, ectoderm, mesoderm and endoderm (figure 4). Ectoderm composed of stratified squamous epithelial lining, keratin flakes and glial component. Mesoderm composed of cartilage, blood vessels, adipose tissue, bone marrow, lymphoid collection and foamy macrophages. Endoderm composed of transitional epithelial lining, glandular lining, columnar lining of cells and mucinous salivary gland. Histopathology was consistent with dermoid cyst/mature cystic teratoma. As the abdominal CT and chest X-ray images were negative for primary tumour in the ovaries or mediastinum, diagnosis of a primary retroperitoneal mature teratoma was confirmed.

OUTCOME AND FOLLOW-UP

At 3-month follow-up, patient is completely asymptomatic.

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Figure 1 Clinical photograph of the patient showing large lump more towards right side of abdomen.

DISCUSSION

The lipomatous lesions of the adrenal gland include myelolipoma, lipoma, angiomyolipoma, liposarcoma. Myelolipoma is the second most common adrenocortical tumour and the most common adrenal lipomatous tumour.² Myelolipoma is a rare, metabolically silent benign adrenal lesions consisting of mature fatty tissues and bone marrow elements. Adrenal myelolipoma was the presumptive diagnosis in our case based on the patient age, symptoms, suprarenal location of tumour, radiological characteristics with fat within the tumour and suspected benign nature of disease.

Teratoma is a germ-cell tumour derived from totipotent cells and made of cell types of more than one germ-cell layer. Teratomas are located in paraxial regions, common sites being gonads followed by extragonadal sites such as intracranial, cervical, mediastinal, retroperitoneal and sacrococcygeal sites. Primary retroperitoneal teratomas are exceedingly uncommon accounting for only 1% of all primary teratomas and can be mistaken for adrenal lesions.¹ Histopathological examination is confirmatory for final diagnosis.

The incidence of the retroperitoneal mature cystic teratoma peak in the first 6 months of life and in early adulthood.³ As in our case, based on radiology, the right-sided adrenal gland was inseparable from the mass, giving an appearance that the tumour

Figure 2 Contrast-enhanced CT abdomen (longitudinal sections) showing giant hypodense cystic mass lesion in right hypochondrium, epigastrium and crossing the midline to left lumbar region with fatty attenuations with calcifications (A). The mass lesion is seen displacing the right kidney in right iliac fossa, liver being pushed upwards, pancreas posterosuperiorly, bowel loops laterally and in pelvis (B).

Figure 3 (A) Intraoperative photograph showing elongated right renal vein (marked by black arrow) and is seen crossing the mass anteriorly. (B) Picture showing the mass after dissection with right kidney pushed in iliac fossa (marked by black arrow). (C) Photograph showing excised specimen.

might have arisen from the adrenal gland. Such an unusual radiological finding may cause difficulty in diagnosis.

Adrenal/retroperitoneal teratoma poses a strong diagnostic challenge to radiology. Plain X-ray of abdomen can show calcification or diffuse mass. Ultrasonography shows fluid collection and calcification. They are better appreciated on CT/MRI. Radiological findings such as calcification and predominant fat content often make a diagnosis of teratoma difficult to distinguish from lipomatous tumours of the adrenal gland such as myelolipoma or angiomyolipoma. Compared with these other adrenal tumours, teratomas tend to exhibit a more heterogeneous appearance with a mixture of fluid, fat and sebum forming a fat-fluid level.

After diagnostic dilemma, management decision depends on the nature of the mass, the need for complete resection, measures to prevent recurrence, neoadjuvant or adjuvant treatment and minimally invasive versus open surgery. Even with technical advancement in laparoscopic surgery, such large tumours (33×27×15 cm) may require open surgical resection and inevitable poor cosmetic outcome and delayed postoperative recovery. Earlier, adrenal tumour size >5–6 cm was considered a contraindication for laparoscopic approach, but recent studies have shown that laparoscopic approach can be done for larger size adrenal tumours (~15 cm).⁴ Patients undergoing laparoscopic removal have longer operative time but had shorter hospital stays and faster recovery.⁵

In our case, although a presumptive diagnosis of adrenal myelolipoma was made, but the intraoperative finding of normal

Figure 4 Histopathological slide showing section of (A) cartilage, (B) bone, (C) melanocytes and (D) transitional epithelium.

Learning points

- ▶ Primary benign retroperitoneal teratoma is a rare tumour, especially in the adult population.
- ▶ Adrenal myelolipoma masquerades as retroperitoneal teratoma on radiological imaging.
- ▶ Giant retroperitoneal tumour are difficult to manage by minimally invasive surgery; hence, open surgery still is a viable tool.
- ▶ Serum tumour markers such as alpha-fetoprotein, -human chorionic gonadotropin should be done in case of suspicion of teratoma.

adrenal gland over the mass and considering histopathological examination report, a final diagnosis of primary retroperitoneal teratoma was made.

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