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Abdominal mass and haematuria

PM Hewitt, WY Lau, TM Mackenzie, KF To, AKC Li

A 19-year-old woman presented with left loin pain, macroscopic haematuria and a vague abdominal mass. Renal function, full blood count and coagulation screen were normal. Plain abdominal X-rays revealed a large soft tissue mass occupying the left side of the abdomen and on intravenous urography, the kidney and ureter were displaced laterally with blood clot in the calyces. A computed tomography (CT) scan of the abdomen was done (figure 1).

**The Chinese
University of Hong
Kong, Prince of
Wales Hospital,
Shatin, NT, Hong
Kong**
**Department of
Surgery**
PM Hewitt
WY Lau
AKC Li
**Department of
Urology**
TM Mackenzie
**Department of
Anatomical and
Cellular Pathology**
KF To

Correspondence to
Professor AKC Li

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Figure 1 Abdominal CT scan

Questions

- 1 What does the abdominal CT scan show?
- 2 What are the differential diagnoses?

Answers

QUESTION 1

Axial CT scan through the kidneys shows a large multi-septated cystic lesion displacing the left kidney and splaying the renal vessels.

QUESTION 2

The differential diagnosis of retroperitoneal cystic lesions is given in the box.

At operation, a complex cyst containing serosanguinous fluid was found and complete excision was achieved together with a nephrectomy. Histological examination confirmed a benign cystic lymphangioma with intrarenal extension (figure 2). The lesion was composed of multiple thin-walled lymphatic channels lined by endothelial cells with a stroma of delicate fibrous tissue containing lymphoid aggregates. The presence of haemosiderin pigment in some areas suggested that bleeding had been chronic.

Discussion

Retroperitoneal lymphangiomas are developmental abnormalities rather than true neoplasms and are almost always benign.^{1,2} They usually present during infancy and childhood,¹ or may be asymptomatic and only found

Differential diagnosis of retroperitoneal cystic lesions

- congenital: polycystic kidney, cystic lymphangioma, enteric duplication, mesenteric cyst, teratoma
- infective: echinococcal cyst, abscess
- neoplastic: cystadenoma/adenocarcinoma, cystic nephroma, sarcoma, lymphoma (not truly cystic)
- other: pancreatic pseudocyst, ovarian cyst, haematoma/seroma

incidentally during radiological procedures, surgery or at autopsy.^{2,3}

The most common clinical manifestation is that of a slowly enlarging abdominal mass.^{2,3} However, some patients develop bowel or ureteric obstruction, or occasionally present acutely with infection, torsion, perforation or bleeding into the cyst.² Haematuria is rare⁴ and together with a palpable mass, would normally signify more ominous pathology. Although consumptive coagulopathy has been associated with lymphangiomatosis,^{2,3} this was not the case in our patient. Presumably her symptoms were of intrarenal origin or due to extension of the lesion (figure 2). The latter is thought to reflect embryological development rather than true invasion.⁵

Pre-operative diagnosis of retroperitoneal lymphangiomas is difficult, as they may be confused with other cystic conditions.² Barium studies or intravenous urography may show organ displacement,¹ while ultrasound and CT characteristically demonstrate uni- or multi-locular cysts, with septae of uniform thickness, containing fluid of water density.^{2,5} Magnetic resonance imaging can display the lesions as well as CT, although variations in signal intensities, and T-1 and T-2 values, may reflect differences in the composition of cyst fluids.^{2,3}

Complete excision of retroperitoneal lymphangiomas is the preferred treatment and is usually carried out with relative ease. Other forms of management, such as cyst-enterostomy or marsupialization, have become obsolete.² When the lesion arises from, or involves, an abdominal organ, this should be partially or completely resected with the specimen^{2,4} as there is a possibility of recurrence with incomplete removal.²

Final diagnosis

Retroperitoneal cystic lymphangioma arising from or extending into the kidney, causing haematuria.

Keywords: retroperitoneal lymphangioma, kidney, haematuria

Figure 2 The lymphangioma is composed of multiple dilated lymphatic channels with patchy lymphocyte infiltrates in their walls. Renal medullary tissue is seen on the left side of the figure. The arrow indicates a nest of renal tubules surrounded by lymphatic tissue

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