Surgical management of giant retroperitoneal lymphangioma in a child

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DESCRIPTION

A 3-year-old boy was noticed by parents to have right flank distension. On evaluation with ultrasonography, there was a large multiseptate collection with internal echoes occupying the right half of retroperitoneum. CT of the abdomen revealed a 10×12×13 cm well-defined, thin-walled multiloculated cystic lesion with enhancing septae displacing the right kidney and extending from the under surface of the liver above to the pelvic brim below (figure 1). The boy underwent laparotomy and intraoperatively a 12×12×10 cm globular cystic mass was found, arising from the retroperitoneum on the right side displacing the bowel and right kidney and ureter medially and having extension into the posterior abdominal wall and paravertebral muscles (figure 2). The cystic mass was completely excised and its histopathological examination revealed a fibrocollagenous cyst wall along with flattened epithelium at places with moderate-to-dense lymphoplasmacytic inflammatory infiltrate and areas of haemorrhage. The cystic cavity was filled with a proteinaceous and haemorrhagic material. These features were suggestive of a lymphangioma. After surgery, the child had an uneventful recovery and at 18 months of follow-up, there is no abdominal distension on clinical examination and no recurrent lesion on ultrasonography.

A lymphangioma is a benign proliferation of lymphatic tissue believed to develop from the early sequestration of lymphatic channels that fail to establish connections with normal draining lymphatics. Retroperitoneal lymphatic malformations are extremely rare and comprise less than 1% of the abdominal lymphatic malformations.¹ Three histological types of lymphangiomas are described: cystic, capillary and cavernous.² Retroperitoneal lymphangioma is commonly of cystic type.³ Most cystic lymphangiomas in the body are present in the first 2 years of life, however lymphangiomas of the retroperitoneum are usually diagnosed in older children or adults.³ It can present as a soft, slowly growing and painless mass or it may be detected incidentally during the evaluation of an unrelated complaint.

Ultrasound is a sensitive and often the initial tool for evaluation of cystic abdominal masses, particularly for identifying septations and floating debris.³ On CT, cystic lymphangiomas are well-defined, homogeneous, usually multicystic and may have mild enhancement of the septa or the wall after intravenous contrast administration.³ They may not be limited to a particular abdominal compartment and may displace intra-abdominal organs and

Figure 1 CT abdomen in (A) non-contrast axial, (B) contrast-enhanced axial and (C) coronal planes showing retroperitoneal multiloculated cystic lesion.

vessels. An important feature on imaging that differentiates these lesions from other pathologies is the insinuating nature crossing multiple compartments, as was in this case. They may also form unilocular or multilocular cystic masses and can encroach on vital structures. Pancreatic pseudocyst and retroperitoneal cystic teratoma must be kept in mind as a differential diagnosis.⁴

However, the final diagnosis is made on pathological examination which shows large macroscopic or microscopic cysts without any capsule and lined by endothelial cells. The cysts are filled with

Figure 2 Intraoperative photograph shows globular cystic retroperitoneal mass.

Patient's perspective

Patient's father: I was told by my doctor that my son has a fluid-filled swelling in his belly which did not look like cancer but required removal by surgery. The surgery went on well I was happy to know that the tissue report did not show any cancer. Presently, my son is doing well and I am very happy that his case was managed very well.

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Learning points

- ▶ Giant retroperitoneal cystic lymphangioma is rare in young children and it usually presents with abdominal distension without any other symptoms.
- ▶ Complete resection is the only curative treatment.
- ▶ The diagnosis is confirmed by histopathological examination of resected specimen.

chylous, serous, haemorrhagic or mixed fluid. The connective tissue is composed of collagen with and without lymphocytes.

Complete resection as the treatment of choice is recommended in all cases including asymptomatic cases due to risk of future complications like haemorrhage, infection, malignant transformation or compression of vital structures.⁵ Conservative methods, including aspiration, cyst enterostomy and peritoneal marsupialisation, are now rarely considered due to the high rate of recurrence.⁶ Surgically unresectable lesions can be treated with intracystic injection of a sclerosing agent, preferably OK-432.⁷

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