Giant retroperitoneal cystic lymphangioma in a seven-months-old girl

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CASE REPORT

A seven-month-old girl presented with complaints of gradually increasing abdominal lump for four to five months. Clinical examination revealed grossly distended abdomen with an ill-defined nontender, cystic lump, 30 × 15 cm, involving the entire abdomen more on the left side with a positive transillumination test and fluid thrill [Figure 1]. Other systemic examinations and routine laboratory investigations were within the normal limits.

The plain film of the abdomen showed haziness with no evidence of calcification. Ultrasonography of the abdomen revealed a 29 × 14 cm, thin-walled, multiloculated, anechoic cystic mass with septations in the retroperitoneal area occupying almost all the quadrants of the abdomen [Figure 2]. Left kidney and intestines were displaced to the right side of the abdomen and spleen was displaced anteriorly and medially. The CT scan of the abdomen confirmed the findings of ultrasonography [Figure 3].

On surgical exploration, a large retroperitoneal, multiloculated, cystic [Figures 4 and 5] lesion was found; this cyst was adherent to the left dome of the diaphragm, splenic mesentry, left kidney and splenorenal area. It was aspirated and completely excised. Histopathology confirmed the diagnosis of retroperitoneal cystic lymphangioma. Postoperative recovery was uneventful. A follow-up ultrasonography of abdomen performed after a month of operation showed no cysts or recurrence.

Figure 1: Clinical photograph of a patient showing distended abdomen with positive transillumination

Figure 2: Ultrasonography of the abdomen and pelvis showing multiloculated retroperitoneal cyst

Figure 3: CT scan of the abdomen and pelvis showing multiloculated retroperitoneal cyst pushing left kidney, intestines, etc. toward the right side
Retroperitoneal lymphangioma is a rare congenital malformation arising from the retroperitoneal lymphatics. The mechanism for the formation of lymphangioma is the early developmental sequestration of lymphatic vessels that fail to establish connection with normal draining vessels at approximately 14 to 20 weeks of intrauterine life.\(^1\,^2\)

Lymphangiomas have a marked predilection for the head and neck regions. More than 95% are situated in the head, neck, axilla and extremities. Other sites including the abdomen are observed only in 5% of the cases. Retroperitoneal lymphangiomas are rarer than abdominal lymphangiomas of the mesenteric origin and 90% of it is usually present as palpable soft abdominal mass by the second year of life.\(^3\)

Although lymphangiomas are benign lesions, they can compress and infiltrate vital structures or can exist with complications such as intracystic haemorrhage, cyst rupture or infections.\(^3,^4\)

Lymphangiomas are histologically classified into the following: capillary (lymphangioma simplex), cavernous and cystic lymphangioma (cystic hygroma) with retroperitoneal lymphangioma being almost always cystic; these retroperitoneal cysts may be unilocular or multilocular and contain serous or chylous fluid.\(^3,^5\)

Differential diagnosis includes multiloculated ascitis, mesenteric cysts, enteric duplication cysts, mesothelial cysts, nonpancreatic pseudocysts, etc.

Complete surgical enucleation remains the therapy of choice for retroperitoneal lymphangioma to date.\(^2\,^4\) Laparoscopic excision and image-guided percutaneous catheter drainage followed by sclerotherapy of the retroperitoneal lymphangioma has also been reported in children.\(^6,^7\) Retroperitoneal lymphangiomas have more chances of recurrence; hence, a regular postoperative follow-up with repeated sonography is suggested.

REFERENCES


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