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Mesenteric cystic lymphangioma with myxoid degeneration: unusual CT and MR manifestations

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Abstract. We report an unusual case of mesenteric cystic lymphangioma presenting as a large multilocular mass with a well-enhanced solid component and a central cleft, which were were pathologically correlated to the prominent stromal myxoid degeneration interspersed with abundant capillaries and the central fibrosis, respectively. The findings of computed tomography and magnetic resonance imaging are illustrated. Recognition of the multilocular configuration of the enhanced stroma may help to make the correct preoperative diagnosis.

Abdominal cystic lymphangiomas (ACL) are uncommon benign tumors with an incidence of 1 in 20000 pediatric admissions [1]. ACL most commonly involve the mesentery, but also occur in the omentum and retroperitoneum. These lesions usually appear as well-defined unilocular or multilocular thin wall cysts without or with septations [2–5]. We describe the unusual computed tomographic (CT) and magnetic resonance (MR) findings of a case of mesenteric cystic lymphangioma with pathologic correlation.

Case report

A 7-year-old boy received right inguinal herniorrhaphy at another hospital 10 days before being referred to our hospital owing to abdominal distension and intermittent abdominal pain. On physical examination, distended abdomen with diffuse tenderness and muscle spasm was found. The laboratory findings were unremarkable except for mild eosinophilia. Plain film of the abdomen revealed a large mass displacing the bowel gas to the right lower quadrant and pelvis. Abdominal ultrasound (US) performed by the pediatrician revealed a huge solid tumor with heteroechogenicity occupying the whole abdomen.

Pre-enhanced CT scan demonstrated a 25-cm homogeneous low density mass (15 HU) with delicate septa outlining a central cleft. Enhanced CT scan showed that there was inhomogeneous enhancement (40–70 HU) throughout the tumor except in the central cleft (Fig. 1). MR imaging (1.5-T Signa System, General Electric Medical Systems, Milwaukee, Wis.) showed that the mass was homogeneously hypointense on T1-weighted images (T1WI). T2-weighted images (T2WI) revealed a large hyperintense mass with a hypointense central cleft. Gd-DTPA enhanced T1WI revealed a predominate well-enhanced solid component which was organized in a multilocular pattern (Fig. 2).

At surgery, a 20 × 25 × 25-cm huge reddish mesenteric mass occupying nearly the whole abdomen as found. Total excision of the tumor along with 60 cm of jejunal loops was done. On sectioning, the tumor was multiloculated with a central scar, but a large portion of the mass was filled up by a reddish solid substance. Microscopically, endothelium-lined cystic lymphatic spaces, prominent stromal myxoid degeneration with abundant capillaries, and central fibrosis were shown (Fig. 3). The final diagnosis was mesenteric cystic lymphangioma with prominent myxoid degeneration. The patient recovered uneventfully and there was no evidence of recurrence 3 months after surgery.

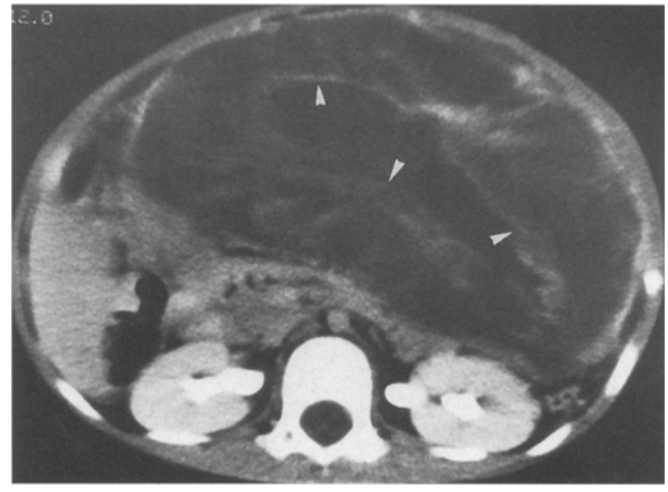

Fig. 1. Axial enhanced CT shows faint inhomogeneous enhancement of the abdominal tumor peripheral to the hypodense central cleft *(arrowheads)*

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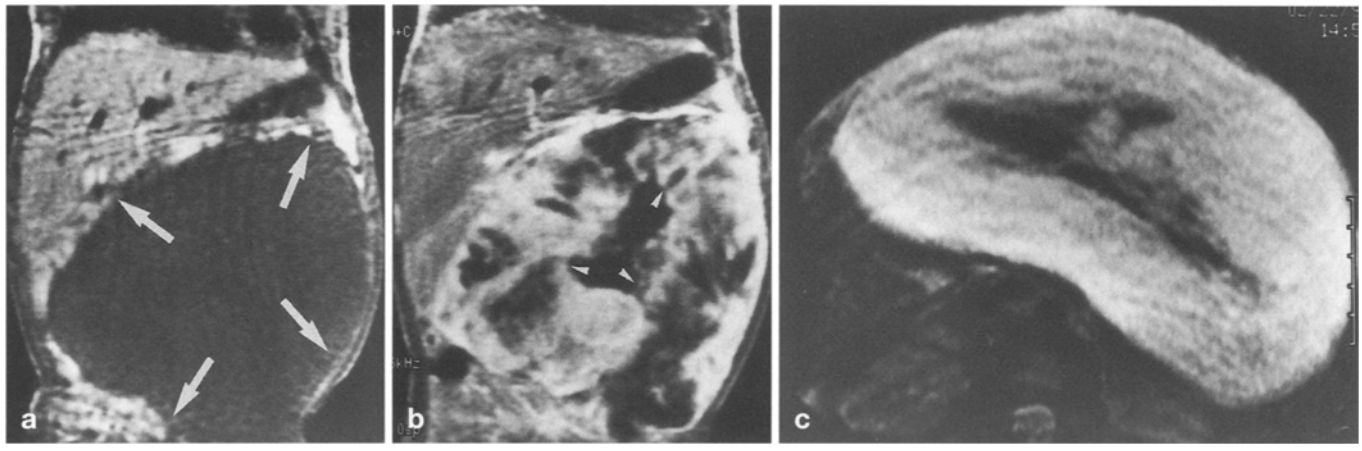

Fig. 2a Coronal T1-weighted image [spin-echo (SE) TR 500/TE 10] shows a huge abdominal tumor with homogeneous low signal intensity *(arrows)*. **b** Gd-DTPA enhanced coronal T1-weighted image (SE 466/11) reveals predominate well-enhanced soft tissue arranged in a multilocular pattern and the hypointense central cleft *(arrowheads)*. **c** T2-weighted image (SE 2500/90) shows the hypointense central cleft while the rest of the tumor has become hyperintense

Discussion

Lymphangiomas are believed to be developmental anomalies with failure of developing lymphatic tissues to establish normal communication with regional lymphatic drainage, resulting in dilatation of the abnormal channels. Histologically, lymphangiomas are classified as simple, cavernous and cystic types, depending on the size of the lymphatic spaces [2]. About 95 % of lymphangiomas occur in neck and axillary regions. The remaining 5 % may occur throughout the body, mostly in the abdominal mesentery [2–5].

About 25 % of ACL are identified in the first decade of life [2–4]. Abdominal pain and distension are the most common presentations. The abdominal pain may be due to bleeding, infection, torsion or compression [1]. Palpation of ACL may be difficult as it is soft and may even mimic ascites [5]. Furthermore, as in our case, abdominal pain and muscle spasm hampered detailed physical examination. US and CT facilitate early diagnosis of the ACL and early treatment is advocated to avoid severe complications [1].

The imaging features of ACL have been well described. On US, ACL are typically single or multiple anechoic cysts without or with septations [3]. On CT, they usually present as well-defined homogeneous unilocular or multilocular low density lesions without or with slight wall enhancement. The attenuation value of the cysts is usually consistent with fluid density [2, 4]. Occasionally, a low CT attenuation value (– 15 HU) due to the chylous content may be identified [4, 6]. On MR imaging, the lymphangiomas are usually hypointense on T1WI and hyperintense on T2WI, with focal linear inhomogeneities corresponding to the fibrous septa [7, 8]. In our case, a solid abdominal tumor was initially diagnosed by US. While ACL with hemorrhage or infection may mimic solid tumors or necrotic metastasis sonographically [2–4, 8], CT can reveal the cystic nature of

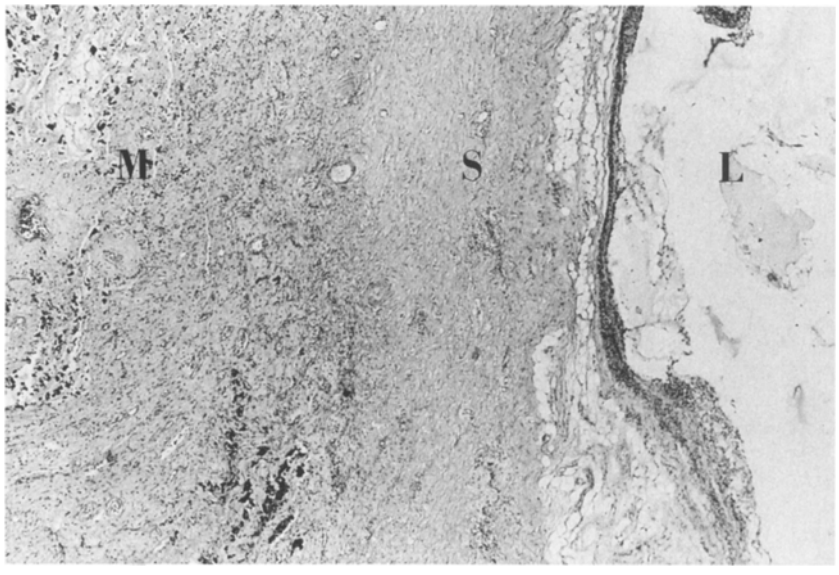

Fig. 3. Histologic section shows a cystic lymphangioma with dilated lymphatic space *(L)* surrounded by fibrous fatty septa *(S)* and marked stromal myxoid degeneration with abundant capillaries *(M)* (hematoxylin and eosin; original magnification × 21)

most of the lesions. However, the enhanced CT examination of our patient revealed inhomogeneous enhancement (40–70 HU) throughout the tumor, except in the central cleft, indicating that the mass was predominately solid rather than cystic. Gd-DTPA-enhanced T1WI demonstrated the enhanced solid component and its multilocular architecture more clearly. On the other hand, the central cleft remained hypointense on T1WI and T2WI. The unusual well-enhanced solid part of the tumor corresponded to the area of prominent stromal myxoid degeneration interspersed with abundant capillaries, whereas the central cleft represented the central area of fibrosis. On T2WI both the myxomatous stroma and the dilated lymphatic spaces became hyperintense. To our knowledge, ACL with these unusual CT and MR manifestations have not been previously documented. Munechika et al. suggested that primary malignant abdominal tumors like liposarcomas, leiomyosarcomas, fibrosarcomas and malignant teratomas can be distinguished from ACL by the presence of some solid component [2]. Our case demonstrated that ACL with hypervascular myxoid degeneration might occasionally masquerade as a solid mass on CT and MR imaging. Nevertheless, the stromal myxoid degeneration occurred in between the fibrous septa and, thus, the multilocular pattern was still preserved. The authors think that this feature is an important clue to the correct CT and MR diagnosis.

In summary, the CT and MR characteristics of ACL are influenced by the stromal architecture, vascularity and content of the cystic spaces. Besides hemorrhage and inflammation, our case illustrates that the stromal myxoid degeneration can also alter the CT and MR appearance of ACL. ACL should be included in the differential diagnosis of solid abdominal masses, especially when a multilocular stromal pattern could be identified.

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