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Symptomatic Giant Mesenteric Cystic Lymphangioma in Adulthood

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CASE REPORT

Cystic lymphangiomas are congenital malformations of the lymphatic system representing 5% of benign tumors in infants. Intra-abdominal localizations are rarely observed (1/20,000 in children),[1] and the evidence in adulthood is rare and generally because of slow growth.

We present a 40-year-old man with a 5-month history of abdominal discomfort associated with nocturia and pollakiuria. Clinical examination of the patient revealed a palpable mass in the suprapubic quadrant of the abdomen. The ultrasound examination revealed an hypoechoic septated lesion extended from the hypogastrium to cranially measuring up to 20 cm diameter. The contrast-enhanced computed tomography (CT) scan confirmed a mesenteric cyst with parahydric content (Figure 1). A 3D CT showed evidence of bowel dislocation (Figure 2).

We opted for surgical treatment, and an extended midline laparotomy was required to explore the abdomen. The decision not to perform a diagnostic laparoscopy was mainly because of the massive abdominal distension and the mass measures. The neoplasia was excised entirely with wide mesenteric margin preserving vessels and bowel. A pathologic evaluation revealed a multiloculated cystic lesion, with milky-like content, described as a benign cystic lymphangioma showing positive staining for CD31 (marker of endothelial cells) and D2-40 (marker of a glycoprotein found on the lymphatic endothelium) (Figure 3).

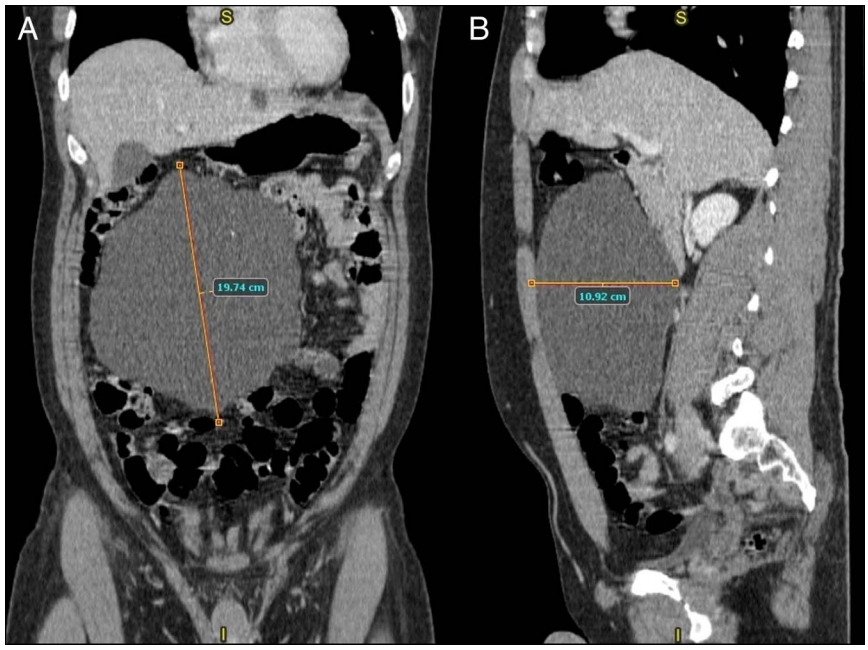

Figure 1. Computed tomography (A) coronal view and (B) sagittal view showing the mesenteric cystic lymphangioma.

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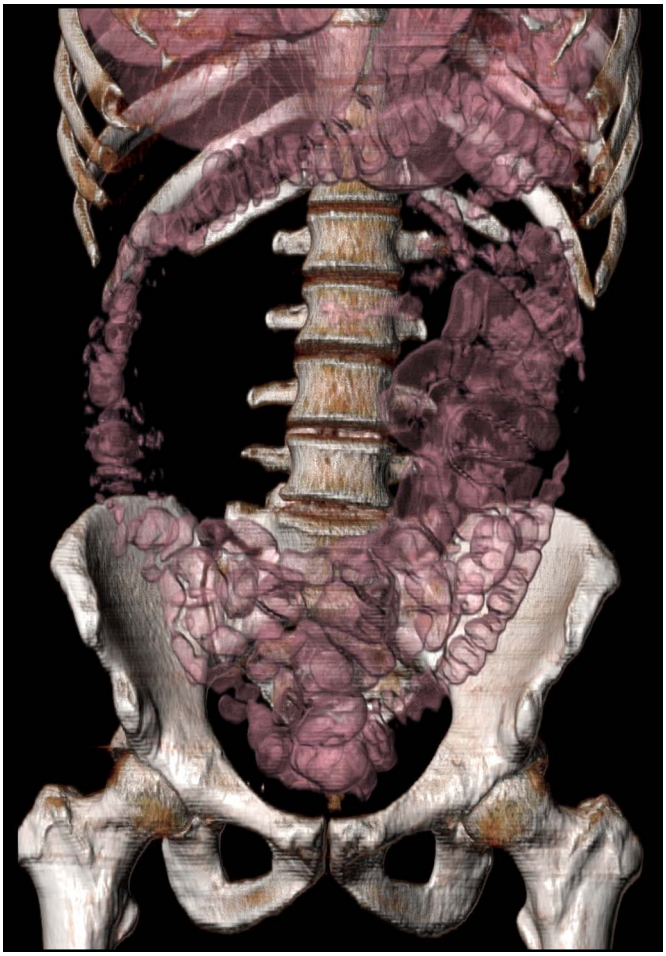

Figure 2. Computed tomography in 3D reconstruction showing the dislocation of the small and large bowel.

Abdominal cystic lymphangiomas are rare congenital benign tumors due to lymph sequestration and the formation of cysts in lymphatic channels of retroperitoneum.[2] The pathogenesis is not clear, and different theories are proposed: infiltrating neoplasm, regionalized lymphatic stasis because of congenital blockage of lymphatic drainage, lymph node degeneration, obstruction secondary to inflammation, or fibrosis of existing lymphatic channels.[3] Clinical manifestations are variable, but 50% of cases present as asymptomatic abdominal masses. When symptomatic, they may present nonspecific but painful abdominal symptoms or they can sometimes be evidenced as acute abdomen because of complications such as obstruction, volvulus, bleeding, and intrabdominal visceral compression.[4] Abdominal radiographs may identify complications or bowel displacement, and abdominal ultrasound may help revealing sharply defined multiloculated cysts with water-like content divided by septations and is the modality of choice for follow-up in children,[2] whereas a CT scan may help in providing the exact location, size and organs, and vessels involvement.[2] A certain diagnosis is then given by the pathological examination. The treatment of choice is the total surgical resection while content aspiration, and incomplete resection is associated with a high recurrence rate (10%).[5] Laparoscopic approach results are feasible in multiple centers but partial intraoperative content aspiration is often necessary for larger lesions.[4]

Although abdominal cystic lymphangiomas are considered benign, they may become locally invasive, involving vessels and bowel and could hide a malignant mucinous tumor.[4] Although recurrences are possible in the above cases, the usual prognosis is excellent, not requiring any long-term follow-up. In conclusion, abdominal cystic lymphangioma is a rare adulthood neoplasm that may uncommonly be associated with atypical abdominal symptoms and take to life-threatening complications, so we suggest an immediate surgical treatment with complete cyst resection.

DISCLOSURES

Author contributions: M. Rossini and A. Annicchiarico wrote the manuscript. M. Rossini, A. Annicchiarico, and F. De Giorgi reviewed

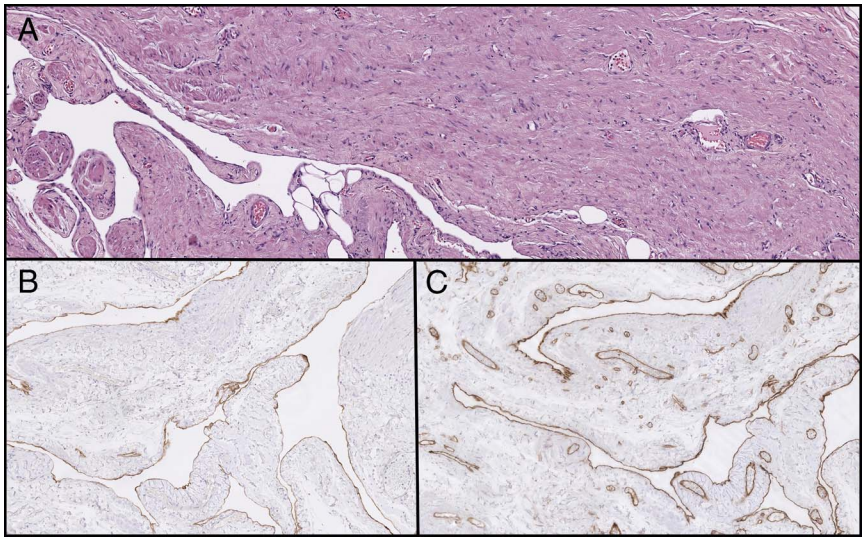

Figure 3. (A) Hematoxylin and eosin stained sections of lymphangioma, (B) D2-40 reacts with a glycoprotein that is only found on the lymphatic endothelium, and (C) immunohistochemical staining with CD31 evidence endothelial cells normally seen in vascular tumors.

the literature. P. Del Rio and L. Viani revised the manuscript for intellectual content. A. Annicchiarico is the article guarantor.

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