Unusual combination of adenocarcinoma in the sigmoid colon and giant cystic retroperitoneal lymphangioma

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We present here a rare case of a retroperitoneal cystic lymphangioma (RCL) combined with adenocarcinoma of the sigmoid colon, lithiasis of the urinary bladder and a large renal cyst. Although RCL is a benign lesion, it may cause clinically important morbidity owing to its large size. It can compress and infiltrate vital structures or present with such complications as intracystic hemorrhage, cyst rupture, volvulus or infection.1–3

Case report

A 78-year-old man complained of dull abdominal pain present for 2 months. Physical examination and blood test results were unremarkable. An abdominal ultrasound demonstrated a cystic mass in the left pararenal space, a renal cyst on the right side and lithiasis of the urinary bladder. A computed tomography (CT) scan revealed a giant mass in the left retroperitoneal space, extending from the tail of the pancreas to the pelvis, with tiny mural calcifications, representing a possible mesenchymal tumour or a liposarcoma (Fig. 1). The mass displaced posteriorly the left renal and splenic arteries. Colonoscopy demonstrated a lesion on the wall of the sigmoid colon; a biopsy specimen revealed that it was an adenocarcinoma.

The patient underwent laparotomy, followed by complete resection of the retroperitoneal mass (Fig. 2). It contained a “milky” fluid. Microscopic examination of the cystic wall demonstrated fibrosis, inflammatory cell infiltrate and the presence of endothelial-lined vascular channels. We drained the cystic mass in the right pararenal space and performed a sigmoidectomy with an end-to-end anastomosis to resect the sigmoid colon adenocarcinoma, followed by removal of 2 stones through a cystotomy. The patient’s postoperative period was uncomplicated.

Discussion

Retroperitoneal cysts are rare benign abdominal lesions. There are no definitive diagnostic tests, and the diagnosis is often difficult to make. Usually, the patient’s history will exclude only traumatic and parasitic cysts.4,5 The lesion in our patient was developmental and lymphatic.

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The prevalence of cystic lymphangioma is about 1 in 6000 babies born, and localization to the retroperitoneum is less than 5%. Initial presentation in adulthood is uncommon, but RCL may present incidentally in later life, typically enlarging slowly and remaining asymptomatic for a long time. If the RCL is large, it may present with symptoms of pressure on adjacent organs, as in our patient.

Differential diagnoses that should be considered for cystic lesions in the retroperitoneum include duplication cysts, pancreatic pseudocysts, ovarian cysts, hematomas, sarcomas, teratomas and abscesses. Computed tomography is ideal for assessing retroperitoneal tumours because it provides discrete sectional images of the organs and retroperitoneal compartments and gives other important information: lesion location, size and shape; the presence and thickness of a wall; the presence of septa, calcifications or fat; and involvement of adjacent structures.4

Surgical excision is the accepted treatment for RCL, and total excision of the lesion is often possible. In some lymphangiomas, a stem with a small base can be identified macroscopically.2 It is important that this also be excised to prevent local recurrence. If the stem of the cyst and the feeding lymphatic vessels are not ligated, chylous ascites will occur.5

The benign nature of the RCL warrants a conservative surgical approach such as aspiration; however, to avoid recurrence, infection, rupture or bleeding, total surgical resection is recommended. This report highlights the importance of a complete evaluation when a patient presents with abdominal distension and of considering RCL in the differential diagnosis.

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References