JAMA Surgery Clinical Challenge

Retroperitoneal Mass

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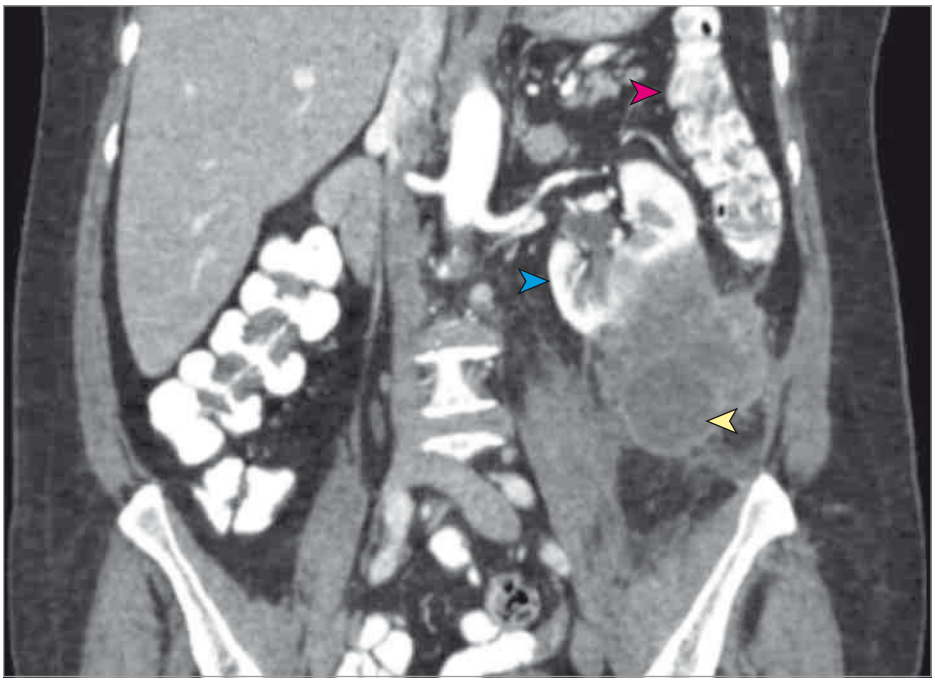

Figure 1. Coronal computed tomographic image showing a retroperitoneal mass with a cystic component and involvement of adjacent kidney and mesocolon. Red arrowhead indicates descending colon; blue arrowhead, left kidney invaded by mass; and yellow arrowhead, retroperitoneal mass with cystic component.

A 57-year-old woman with non–insulin-dependent diabetes mellitus and no other medical history had worsening left-sided back pain over 4 months and no other symptoms. Physical examination findings were significant for mild left flank pain, but there was no costovertebral angle tenderness. Laboratory results were within normal limits. Axial imaging demonstrated an exophytic, retroperitoneal mass with solid and cystic components involving the left kidney, posterior abdominal wall, and mesocolon (**Figure 1**). The mass measured 20 cm in its greatest dimension. Findings on colonoscopy and mammography were negative. Positron emission tomography–computed tomography did not reveal any other abnormality.

Quiz at jamasurgery.com

WHAT IS YOUR DIAGNOSIS?

A. Lymphoma

B. Malignant transformation of an enterogenous cyst

C. Retroperitoneal sarcoma

D. Primary neoplasm arising from the kidney

Diagnosis

B. Malignant transformation of an enterogenous cyst

Discussion

The patient presented with a retroperitoneal mass. The differential diagnosis included primary neoplasm arising from a retroperitoneal visceral structure (ie, pancreas, adrenal glands, kidneys), retroperitoneal sarcoma, lymphoma, or metastatic lesion. Lower on the differential diagnosis were cystic teratomas and other congenital masses of the retroperitoneum. Given that mammography, magnetic resonance imaging, and positron emission tomography–computed tomography did not reveal additional abnormalities, a metastatic lesion was excluded. Similarly, colonoscopy findings were also negative.

With a working diagnosis of retroperitoneal sarcoma, the patient underwent a percutaneous biopsy of the mass.[1] Findings were consistent with adenocarcinoma of unknown primary, making sarcoma and lymphoma less likely. The patient underwent an uneventful resection of the retroperitoneal mass en bloc with left nephrectomy, left colectomy, and resection of the involved posterior abdominal wall musculature, consistent with oncologic principles.[2,3] The posterior musculofascial defect was reconstructed with biologic mesh implantation. Pathological analysis of the specimen revealed a moderately differentiated adenocarcinoma extending from the retroperitoneal area into the lower pole of the left kidney and the wall of the attached portion of colon (**Figure 2**). Embedded in the mass was a fibrous enterogenous cyst with a dysplastic lining with focal mucinous differentiation. Adenocarcinoma was noted to be emanating from the cyst wall. Lymph nodes and margins were negative for carcinoma. Immunohistochemistry was positive for CK-7 and focally positive for CK-20.

Enterogenous cysts, also known as *alimentary tract duplication cysts*, are exceedingly rare with a reported incidence of 1 in 4500 patients by autopsy series.[4] They are by definition congenital as they are derived from cells sequestered from the primitive foregut, midgut, or hindgut and share a blood supply with the adjacent bowel. They can be simple or multiple cysts commonly found on the mesenteric aspect of the intestine. Symptomatic duplications are commonly diagnosed during infancy or early childhood, whereas asymptomatic duplications are more commonly incidental findings on routine imaging studies. Complete excision is the optimal treatment in both scenarios.[5] To our knowledge, this is the fourth case in the literature of a retroperitoneal duplication cyst with adenocarcinomatous changes.[5-7] Prognosis is excellent, even in malignant transformation, provided that margins are negative. As most adenocarcinomas recur locally within the first 3 years, annual surveillance is indicated for up to 5 years.

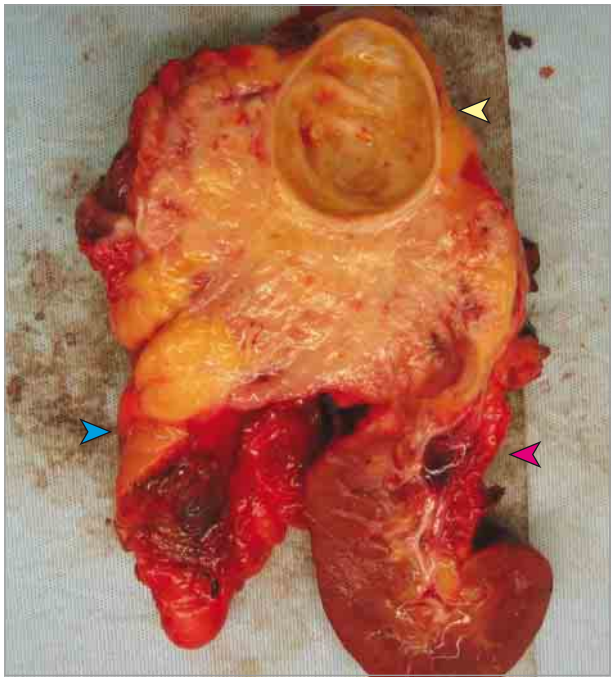

Figure 2. Gross pathology specimen of a retroperitoneal tumor showing an embedded fibrous cyst and involvement of adjacent kidney and mesocolon. Yellow arrowhead indicates retroperitoneal cyst; blue arrowhead, colon mesentery; and red arrowhead, left kidney invaded by adenocarcinoma.

ARTICLE INFORMATION

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