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CASE REPORT

Stromal tumor presents as a large extragastrointestinal mass in the abdominal cavity

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Summary Gastrointestinal stromal tumors are nonepithelial neoplasms of the gastrointestinal tract and have been increasingly recognized in recent years. In contrast, stromal tumor outside the gastrointestinal tract is not frequently found. Here, we present a 57-year-old male patient who had abdominal fullness for several months. It was caused by a 23-cm heterogeneous tumor mass that was successfully removed from the left upper abdominal cavity. The tumor adhered tightly to adjacent organs but postoperative histopathological analysis revealed no direct connection to the stomach, liver, pancreas, spleen, or kidneys. Immunohistochemical examination of the tumor revealed proliferative spindle-shaped cells stained positive for CD117 and CD34 and negative for smooth muscle actin and S-100. The patient received regular follow up. A suspected recurrent liver metastatic lesion was noted 2 years later and radiofrequency ablation of the liver tumor was performed followed by oral imatinib treatment. No tumor recurrence was detected at 3 years after radiofrequency ablation. This case reminds us that extragastrointestinal stromal tumors should be considered in the differential diagnosis when a large heterogeneous mass is present in the abdominal cavity. The characteristics of extragastrointestinal stromal tumor are described and the literature is reviewed in this report.

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Introduction

Gastrointestinal stromal tumor (GIST) is a nonepithelial tumor in the digestive tract. The diagnosis and treatment of gastrointestinal stromal tumors have increased in recent years with the help of advanced immunohistochemical methods. However, stromal tumors developing outside the gastrointestinal tract, termed extragastrointestinal stromal tumor (EGIST), are rarer than GIST. Here, we report a patient with a large tumor that grew outside the gastrointestinal tract in the abdominal cavity, with features of stromal cells. It was diagnosed as EGIST. The characteristics of this EGIST are described and the literature is reviewed and discussed.

Case report

A 57-year-old man had a history of essential hypertension, type 2 diabetic mellitus, and chronic constipation for several years. He experienced epigastric discomfort in 2006. The sensation of the epigastric discomfort was dull and compressive, and lasted approximately 30 minutes. At that time, there was no dysphagia, odynophagia, nausea, vomiting, heartburn, hunger pain, acid regurgitation, or easy satiety. There was no overt precipitating, aggravating, or alleviating factor. He took some antacids, and rapid improvement was seen initially. However, after intermittent treatment for 9 months, the same symptoms recurred and became worse despite continued treatment. Occasionally epigastric cramps and fullness followed. His appetite was fair, but easy satiety and body weight loss of $\sim$5 kg were found. A fecal occult blood test was negative in two separate studies.

On admission, a physical examination found that the abdomen was flat without tenderness or rebounding pain but there was a palpable soft tissue mass over the epigastric region. The patient did not notice the epigastric mass at all. Plain abdominal film showed an increased soft tissue density over the upper abdomen (Fig. 1A). Whole abdominal sonography disclosed that it was a cyst-like lesion, with some blood—mucous like material inside. It was >20 cm in diameter (Fig. 1B). Upper gastrointestinal endoscopy showed an external compression sign over the posterior wall of the gastric body with normal gastric mucosa. Computed tomography of the upper abdomen revealed that there was a large mass of heterogeneous density, measuring 23 cm $\times$ 13 cm $\times$ 20 cm, with possible central necrosis and peripheral enhancement, and the spleen and stomach were compressed by the mass (Fig. 1C). The patient soon received surgical intervention. Because the tumor was involved extensively, total gastrectomy, Roux-en-Y esophagojejunostomy, splenectomy, distal subtotal pancreatectomy, cholecystectomy, and segmental colectomy with anastomosis were performed smoothly (Figs. 2A and 2B).

Grossly, there was no connecting stalk between the tumor and adjacent organs. Serial microscopic surveys showed that the stomach, pancreas, and spleen were compressed by the stromal cell tumor. There was fibrosis of the adjacent pancreatic tissue. There was no lymph node metastasis. The microscopic study of the tumor part showed spindle-shaped neoplastic cells with uniform and syncytial-appearing eosinophilic cytoplasm (Fig. 3A). The mitotic count was 12/50 high-power field (HPF). The MIB-1 test showed an increased proliferation index > 10%. Immunohistochemical study of the tumor cells showed positive for CD117 and CD34, and negative for smooth muscle actin and S-100 (Fig. 3B).

The postoperative course was smooth and uneventful. Two years later, abdominal computed tomography showed a metastatic nodule in the lateral segment of the liver. Radiofrequency ablation of the tumor was performed, followed by oral imatinib treatment. The patient remained free from tumor recurrence in the subsequent 3 years and is still under follow up.

Discussion

GIST constitutes only 1% of the total primary digestive tract cancer. The annual incidence of GIST is 15—20 cases per million population [1—4]. Due to the improvement of diagnostic immunohistochemical techniques, the numbers of GISTs have increased in recent years. GIST occurs in and throughout the whole gastrointestinal tract. The most common sites of origin are the stomach (60%), followed by

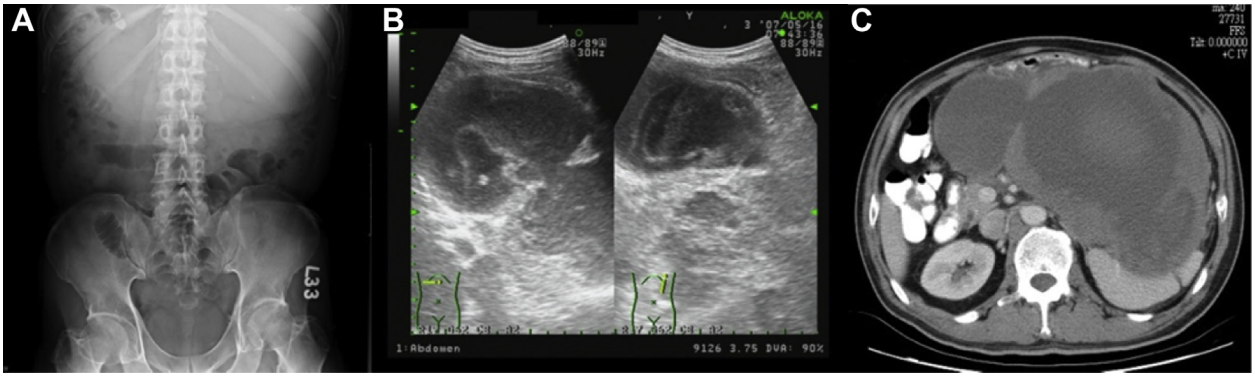

Figure 1 (A) Kidney, Ureter, Bladder (KUB) X ray found increased soft tissue density over the epigastric region with downward displacement of transverse colon gas; (B) sonographic finding of transverse and longitudinal view. There was a large cystic lesion with septation and mucous-like content in the epigastric and left upper quadrant of the abdomen. The pancreas was compressed downward by the tumor at the head and body; (C) computed tomography showed a large mass, approximately 23 cm $\times$ 13 cm $\times$ 20 cm, with low density at the central region and peripheral enhancement. The spleen and stomach were heavily compressed by the tumor mass.