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Inflammatory Myofibroblastic Tumor Treated with Laparoscopic Proximal Gastrectomy and Double-Tract Anastomosis

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Inflammatory myofibroblastic tumors (IMTs) of the stomach are extremely rare in adults, and their oncologic prognosis is not well understood. We present a 28-year-old man with a proximal gastric IMT. The patient visited the emergency department of Yeouido St. Mary's Hospital with syncope and hematemesis. Hemoglobin levels were <5.5 g/dl. Gastric fibroscopy showed a protruding mass 4×4 cm in size, with central ulceration on the posterior wall of the fundus and diffuse wall thickening throughout the cardia and anterior wall of the upper body. Endoscopic biopsy revealed indeterminate spindle cells, along with inflammation. Given the risk of rebleeding, an operation was performed despite the uncertain diagnosis. Because the mass was circumferential, laparoscopic proximal gastrectomy and double-tract anastomosis were performed to ensure a safe resection margin. The pathological diagnosis was consistent with an IMT originating from the stomach, although the tumor was negative for anaplastic lymphoma kinase gene mutation.

Key Words: Stomach neoplasms; Laparoscopy; Gastrectomy

Introduction

Inflammatory myofibroblastic tumors (IMTs) are rare neoplasms that occur preferentially in children and young adults.[1] IMTs most commonly occur in the lungs, mesenteries, omentum, and retroperitoneum.[1,2] IMTs involving the stomach are extremely rare. These distinctive tumors are composed of myofibroblastic cells accompanied by an inflammatory infiltrate of plasma cells, lymphocytes, and eosinophils.[3] IMTs present with nonspecific clinical symptoms, and their preoperative diagnosis can be difficult. IMTs in the gastrointestinal tract can present with bleeding, obstruction, anemia, or intussusception, depending on the site.[4]

Herein, we present a case of an IMT that involved nearly the entire proximal stomach. The patient underwent laparoscopic proximal gastrectomy with double-tract anastomosis.

Case Report

A 28-year-old man presented with syncope and hematemesis. His symptoms were not associated with abdominal pain, dyspepsia, fever, or weight loss. The patient had no notable medical or operational history except an admission for bronchial asthma during childhood. Physical examination of vital signs revealed hypotension with tachycardia, but no other abnormalities. Hemoglobin was <5.5 g/dl, and hematocrit was 16.5%. Emergency gastric fibroscopy detected a protruding mass 4×4 cm in size, with central ulceration at the posterior wall of the fundus and two slightly protruding submucosal lesions with central erosion on the lesser curvature side of the upper stomach body (Fig. 1). A biopsy of the fundic protruding lesion revealed no diagnostic information except that it was a spin-

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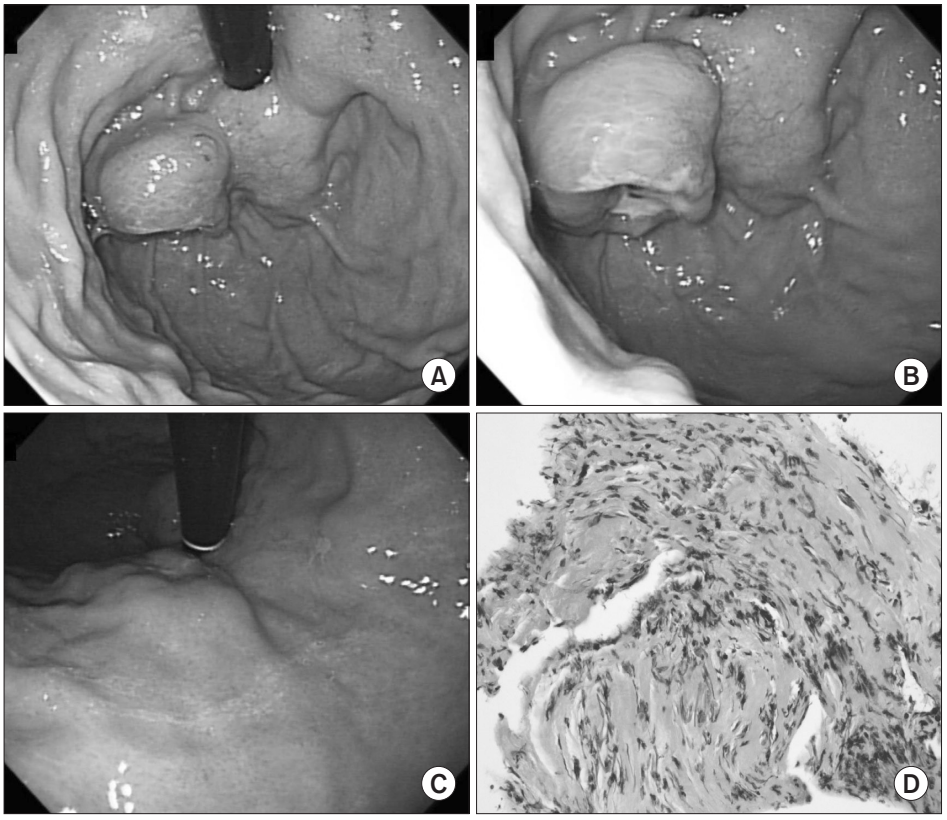

Fig. 1. Preoperative endoscopic and biopsy findings. (A, B) Protruding mass 4×4 cm in size with central ulceration. (C) Two submucosal tumors on the lesser curvature side of the cardia and upper body. (D) Histological examination of the fundic mass revealed nonspecific spindle cells and inflammatory cells (H&E, ×100).

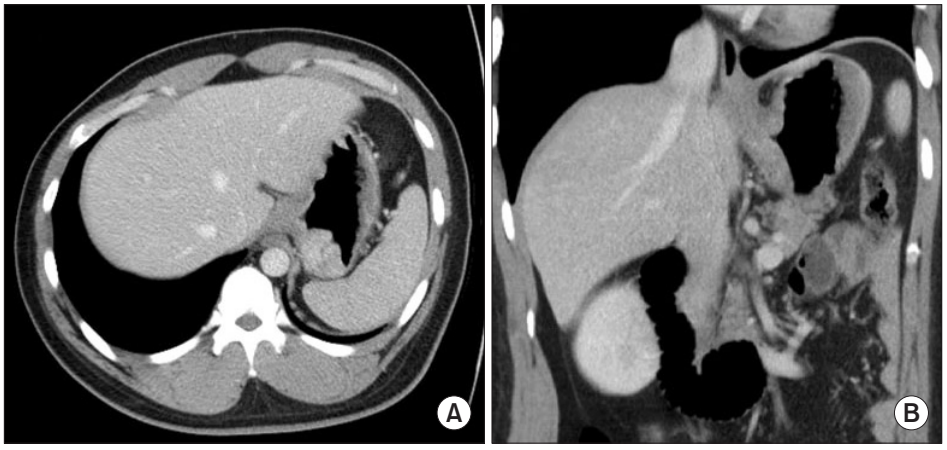

Fig. 2. Computed tomography scan shows fundic mass with diffusely thickened gastric wall on the lesser curvature side of the upper body and cardia. (A) Axial image. (B) Coronal image.

dle cell-type lesion (tests for CD34, CD117, actin, and S-100 were all negative). Computed tomography showed a 3.6-cm enhancing ulcerofungating mass with a 4-cm eccentric wall thickening and an enhancing mucosal layer on the lesser curvature side of the upper stomach body (Fig. 2).

Although the diagnosis was uncertain, an operation was planned due to the risk of rebleeding. Because the fundic mass and the thickened portion of the lesser curvature formed one continuous, massive lesion, laparoscopic proximal gastrectomy was performed to resect the entire area involved. The stomach was reconstructed through double-tract anastomosis. The cut surface of the resected specimen revealed two lesions and involvement of the muscle layer (Fig. 3). Histological examination showed that diffuse fibrotic lesions had invaded the submucosal and subserosal layers. The tumor was mainly composed of spindle-shaped cells in a hyalinized stroma with lymphoplasmacytic infiltration compatible with a diagnosis of IMT. Although rearrangement of the anaplastic lymphoma kinase (*ALK*) gene was not found, the tumor was focally positive for actin, and other immunohistochemical results (CD34, CD117, and S-100 tests were all negative) excluded gastrointestinal

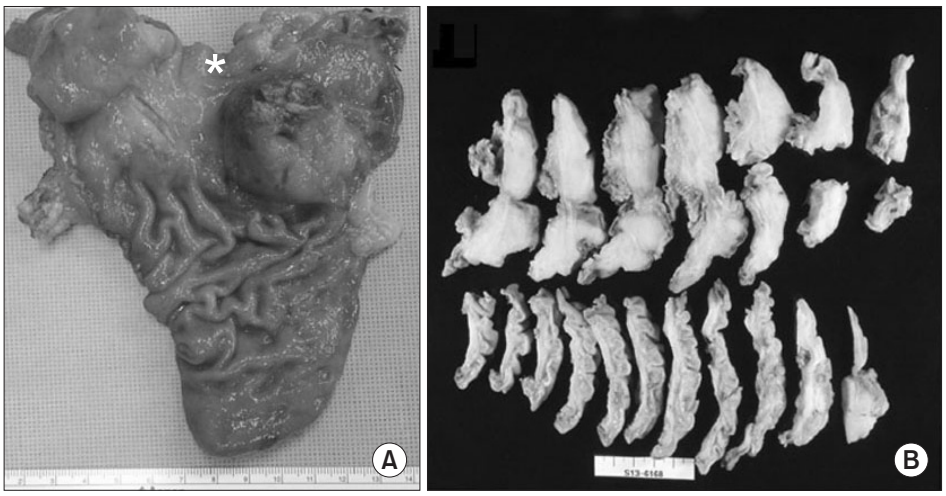

Fig. 3. (A) Opened specimen shows protruding mass on posterior wall of fundus and diffuse wall thickening of cardia and high body (*esophageal mucosa). (B) Cut section shows large mass with infiltration into the muscle layer.

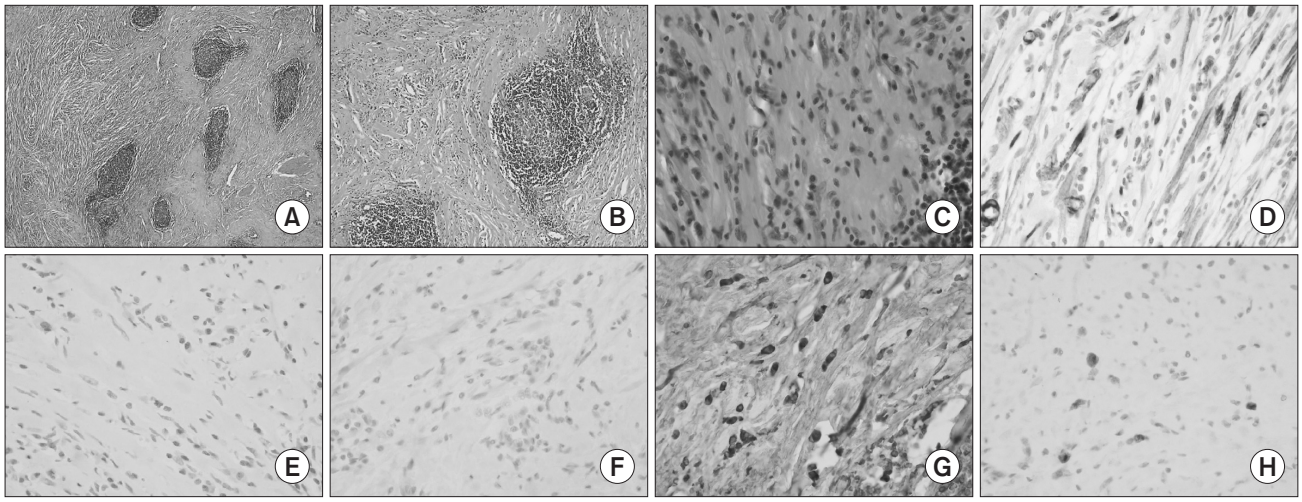

Fig. 4. Histological findings show spindle-shaped tumor cells under a myxoid stroma with lymphoplasmacytic infiltration among the tumor cells. (A) H&E, ×40. (B) H&E, ×100. (C) H&E, ×400. (D) Actin (+), ×400. (E) CD117 (−), ×400. (F) Anaplastic lymphoma kinase mutation (−), ×400. (G) IgG (+), ×400. (H) IgG4 (+), ×400; IgG4/IgG ratio <0.4. IgG = immunoglobulin G.

stromal tumor, leiomyoma, and neurogenic tumor. In addition, immunoglobulin G4 (IgG4)-related sclerosing disease could be ruled out because IgG4/IgG ratio was less than 0.4 (Fig. 4). Finally, the tumor was diagnosed as an IMT. The patient showed an uneventful postoperative course, and no local recurrence or distant metastasis was found during the 18-month follow-up.

Discussion

IMT was first described in 1937 as a primary lung tumor and has since been reported in various sites, including the mesenteries, the retroperitoneum, and the gastrointestinal tract.[4] It mainly occurs in young adults and children, and the lung is the most frequent location. Various terms have been used to describe this entity, such as 'inflammatory pseudotumor,' 'pseudosarcomatous myofibroblastic proliferation,' and 'plasma cell granuloma.' IMT is now considered a true tumor, not a pseudotumor.

Gastric IMT is a very rare condition. According to a recent review, primary gastric IMTs have been reported in only 17 adult patients.[5]

The pathogenesis of IMT is unclear. Allergic, immunologic, and infectious reactions have been postulated.[6] Approximately half of IMTs harbor a clonal cytogenetic aberration that activates the *ALK* tyrosine kinase receptor gene on the 2p locus of chromosome 23.[7] Rearrangement of the *ALK* gene is another postulated pathogenesis.

Manifestations of gastric IMT may include abdominal pain, hematemesis, melena, a palpable mass, anemia, and high fever.[6,8]

Hemoperitoneum has rarely been reported as an initial finding in patients with gastric IMT.[9]

Because gastric IMTs localize in the submucosal layer, endoscopy cannot distinguish them from other submucosal tumors. Endoscopic biopsy frequently reveals a normal gastric mucosa. Thus, surgical resection with an adequate margin is the most reliable diagnostic and treatment method. Abdominal computed tomography is helpful for determining the necessary extent of resection.[6] In the present case, a proximal gastrectomy was performed because the gastric IMT involved nearly the entire proximal stomach. The histological appearance of gastric IMTs is characterized by proliferation of spindle-shaped myofibroblastic cells and lymphocytic infiltration among tumor cells. Differential diagnoses for IMT include inflammatory fibrinoid polyp, gastrointestinal stromal tumor, leiomyoma, and follicular dendritic cell sarcoma.[3]

ALK expression distinguishes IMT from its differential diagnoses. However, not all gastric IMTs are positive for *ALK* rearrangement; only about 50% of IMTs in children and young adults show such rearrangement.[1] In addition, IMTs test positive for actin with most immunohistochemical tests.[3] Immunohistochemical tests for CD117, CD34, and S-100 can help distinguish IMT from differential diagnoses. The typical characteristics of an IMT were observed in the present case (myofibroblastic proliferation, lymphoplasmacytic infiltration distributed among the tumor cells). However, these findings are not pathognomonic. Although our case failed to show the *ALK* mutation, the diagnosis was made based on morphological characteristics, the focally positive actin test, and negative results for CD34, CD117, and S-100 staining.

IMT has intermediate biological potential, frequently recurring but rarely metastasizing.[10,11] Among 17 reported gastric IMT cases in adults, only one case recurred, as a peritoneal dissemination following total gastrectomy, distal pancreatectomy, splenectomy, transverse colectomy, and lymph node dissection, which were necessitated by the large size of the tumor.

Gastric IMT is still a rare tumor and has an undetermined prognosis. Although hematogenous metastasis has not been reported, local recurrence can occur. Resection with an adequate margin is the only treatment option, and regular follow-up is mandatory.

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