

CASE REPORT

위장관 출혈과 폐쇄를 동반한 십이지장 평활근육종 1예

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Duodenal Leiomyosarcoma Presenting with Gastrointestinal Bleeding and Obstruction: A Case Report

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Duodenal leiomyosarcoma is a rare condition with a poor prognosis. Early diagnosis of duodenal leiomyosarcoma is challenging because it presents with nonspecific symptoms and endoscopic biopsies usually do not enable a definitive diagnosis. Duodenal leiomyosarcomas are diagnosed on the basis of the histopathological identification of a mesenchymal lesion composed of malignant tumor cells that on immunohistochemical examination is positive for smooth muscle actin and desmin. We report the case of a 38-year-old man who presented with gastrointestinal bleeding and obstruction who was diagnosed with duodenal leiomyosarcoma after surgical resection. (*Korean J Gastroenterol* 2020;75:94-97)

Key Words: Duodenal obstruction; Gastrointestinal hemorrhage; Leiomyosarcoma

INTRODUCTION

Gastrointestinal mesenchymal tumors are a group of non-epithelial tumors typically of mesenchymal origin, and include gastrointestinal stromal tumors (GISTs), non-GIST sarcomas, leiomyomas, schwannomas, glomus tumors, inflammatory fibroid polyps, inflammatory myofibroblastic tumors, and plexiform fibromyxomas.¹ These tumors have similar histological morphologies but different immunohistochemical features.² GISTs are the most common malignant mesenchymal neoplasms affecting the gastrointestinal tract, whereas leiomyosarcomas are rare malignant mesenchymal tumors that may occur throughout the gastrointestinal tract, though they are identified primarily

in the stomach³ and rarely in the duodenum. Because these neoplasms present with nonspecific symptoms and endoscopic biopsies do not usually result in definitive diagnoses, early diagnosis of duodenal leiomyosarcoma is challenging.^{4,5} We report the case of a 38-year-old man who presented with gastrointestinal bleeding and obstruction and was diagnosed with duodenal leiomyosarcoma following surgical resection.

CASE REPORT

A 38-year-old man without any relevant medical history visited a local medical center with complaints of nausea, vomiting, and 15 kg of weight loss. After esophagogastroduodeno-

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scopy (EGD), he was prescribed medications for duodenal ulcer. However, his symptoms persisted and he subsequently developed hematemesis, and therefore, was transferred to Pusan National University Hospital (Busan, Korea) for hemostatic therapy.

On admission, his blood pressure, pulse rate, and body temperature were 140/90 mmHg, 62 beats/min, and 36.4°C, respectively. Physical examination revealed no specific abnormalities and laboratory investigations revealed a white blood cell count of 5,140 cells/ μ L, hemoglobin 12.2 g/dL, platelet count 266,000 cells/ μ L, PT 12.3 sec (INR 1.18), AST 13 IU/L, ALT 14 IU/L, total bilirubin 0.69 mg/dL, total protein 7.2 g/dL, albumin 4.6 g/dL, BUN 9.3 mg/dL, and creatinine 0.9 mg/dL. Electrocardiography findings were normal and plain radiography revealed slight gastric distension. Contrast-enhanced abdominal CT revealed an ulcerative hypodense lesion at the

duodenal bulb (Fig. 1A), and EGD revealed an ulceroinfiltrative mass at the duodenal bulb, nearly obstructing duodenal lumen (Fig. 1B). Histopathological examination of endoscopic biopsy specimens revealed only chronic inflammation with mild eosinophilic infiltration.

No further episodes of hematemesis occurred after EGD, but obstructive symptoms worsened despite medical therapy and he was unable to ingest food. Accordingly, surgical resection was undertaken to relieve symptoms of duodenal obstruction. Exploratory laparotomy revealed significant adhesions around the second portion of the duodenum, and thus, distal gastrectomy with duodenal bulb resection and Roux-en-Y anastomosis, were performed. Macroscopic examination revealed an ill-defined, solid white mass measuring 1.6 \times 1.0 cm in muscularis propria of the duodenal bulb (Fig. 2A). Subsequent histopathological examination of the mass showed proliferating

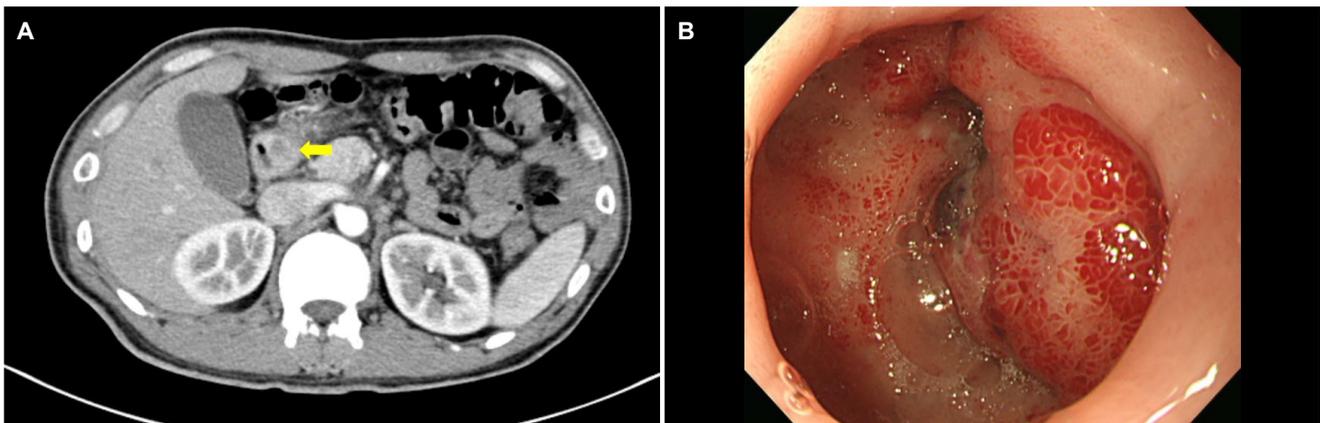


Fig. 1. (A) Contrast-enhanced abdominal computed tomography image showing an ulcerative hypodense lesion at the duodenal bulb (arrow). (B) Endoscopic image showing an ulceroinfiltrative mass at the duodenal bulb nearly obstructing duodenal lumen.

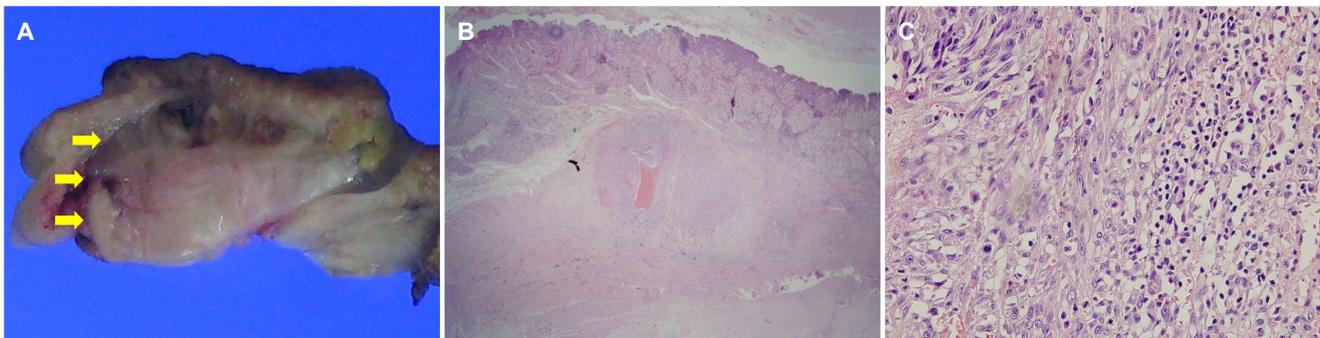


Fig. 2. (A) Macroscopic examination revealed an ill-defined, solid white mass measuring 1.6 \times 1.0 cm in size in muscularis propria of the duodenal bulb (arrows). (B) Histopathological examination of the resected specimen showed the mass was composed of proliferating spindle cells mixed with inflammatory cells within duodenal muscularis propria (H&E, \times 12.5). (C) Spindle cells showed marked pleomorphism and the mitotic count was 21/10 high-power fields (H&E, \times 400).

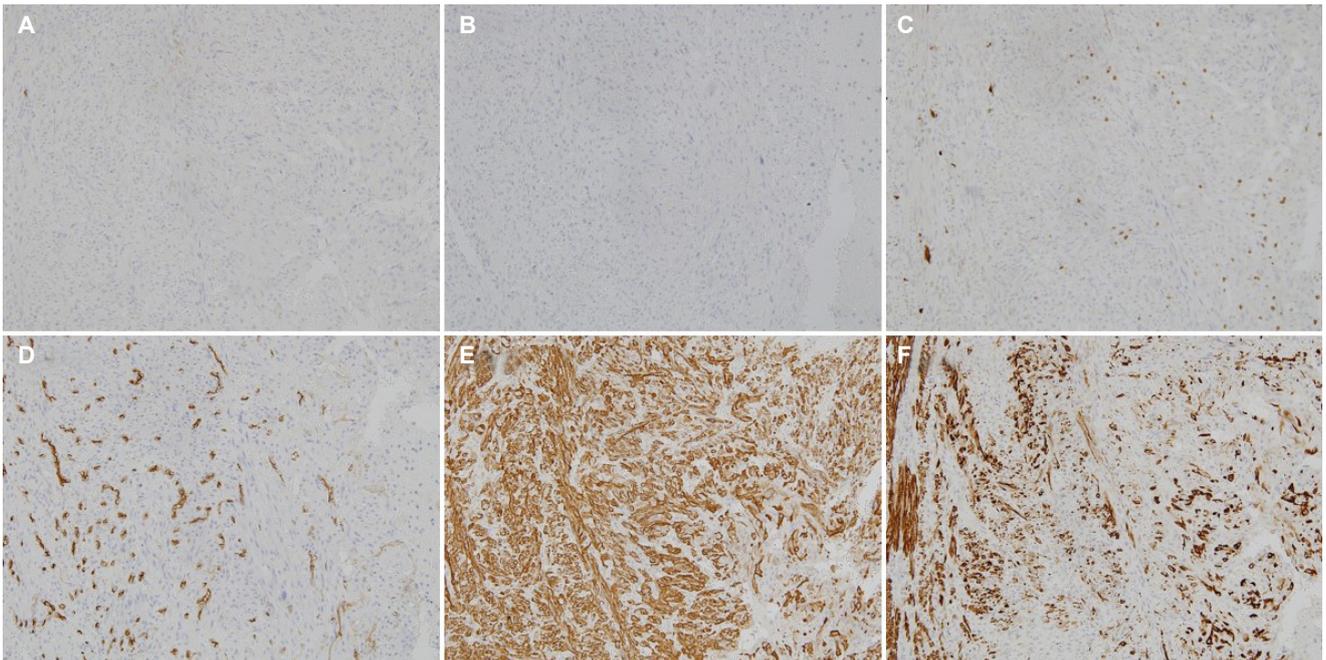


Fig. 3. Immunohistochemical examination revealed tumor cells with immunonegative for (A) c-kit, (B) DOG-1, (C) S-100, and (D) ALK-1 and (E) immunopositive for smooth muscle actin and (F) desmin (original magnification, $\times 200$).

spindle cells mixed with inflammatory cells within duodenal muscularis propria (Fig. 2B). High-power view showed spindle cells with marked pleomorphism and a mitotic count of 21/10 high-power fields (Fig. 2C). Immunohistochemical examination showed tumor cells were negative for c-kit, DOG-1, S-100, ALK-1, and CD34, but positive for smooth muscle actin and desmin (Fig. 3) and had a high Ki-67 labelling index (30%). These observations resulted in a diagnosis of leiomyosarcoma. No lymphovascular invasion or regional lymph node metastasis was observed. The patient refused recommended postoperative adjuvant chemotherapy, and no recurrence was observed at his 32-month follow-up.

DISCUSSION

Duodenal leiomyosarcomas are rare and the majority of reported cases were published prior to the introduction of immunohistochemistry for the diagnosis of GISTs.^{2,6} Immunonegativity for DOG-1, S-100 plus c-kit, and CD34 excludes neurogenic tumors, GISTs, and tumors of vascular origin, respectively,^{6,7} but it is necessary to exclude inflammatory myofibroblastic tumors in specimens showing spindle cells mixed with inflammatory cells in masses. Immunonegativity for ALK-1 excludes inflammatory myofibroblastic tumors and

immunopositivity for smooth muscle actin and desmin indicates tumors of smooth muscle origin, whereas marked nuclear atypia and frequent mitosis indicate leiomyosarcoma.⁶ Our patient was diagnosed with leiomyosarcoma based on immunohistochemical results.

Duodenal leiomyosarcomas are known to occur primarily in patients aged 50-70 years and have been reported to exhibit a slight female predominance.⁴ The second portion of the duodenum is the most common site of involvement.⁸ Our patient was a 38-year-old man with a lesion at the duodenal bulb, which is a rarely reported location. Early diagnosis of duodenal leiomyosarcoma is challenging because patients present with nonspecific symptoms and endoscopic biopsies do not usually result in definitive diagnoses,⁵ and thus, most leiomyosarcomas achieve large sizes before they are palpated or cause obstruction. Most commonly these highly vascular tumors present with bleeding, a palpable mass, or obstructive symptoms.⁵ Our patient complained of gastrointestinal bleeding, nausea, vomiting, and an inability to ingest food.

Surgery remains the treatment of choice for duodenal leiomyosarcomas, and prognosis depends on the risks of recurrence or metastasis.⁹ Reportedly, radiation therapy provides local therapeutic benefit, but does not influence long-term survival.¹⁰ Like other soft tissue sarcomas, leiomyo-

sarcomas spread to lungs and liver via hematogenous dissemination; however, lymphatic dissemination is rare.⁴ Prognosis is poor, mean survival is 50 months and the 5-year survival rate is around 50%.¹¹ The 10-year survival rate after resection is also around 50%, but only 10% without resection.¹¹ Fortunately, in our patient, the tumor was detected at a relatively early stage without distant metastasis, and no recurrence or distant metastasis had occurred after surgical resection at time of writing.

In summary, duodenal leiomyosarcoma is a rare tumor and its diagnosis is challenging. Bleeding and symptoms suggesting gastrointestinal obstruction should raise suspicion of this condition and prompt early diagnosis and treatment. Despite its low incidence, clinicians should consider leiomyosarcoma to be a rare cause of duodenal bleeding and obstruction.

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