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Case Report

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# Plexiform Fibromyxoma Is A Rare Benign Gastric Tumor: A Case Report

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**Abbreviations:**

PF: Plexiform Fibromyxoma**;** PAMT: Plexiform Angiomyxoid Myofibroblastic Tumor**;** EMR: Endoscopic Mucosal Resection**;** IMT: inflammatory myofibroblastoma; CT: Computed Tomography; GIST: Gastrointestinal Stromal Tumor; ALK: Anaplastic Lymphoma Kinase

## 1. Abstract

**1.1. Background**: Plexiform fibromyxoma is a rare mesenchymal tumor of the stomach, found almost exclusively in the antrumor pylorus regions.

**1.2. Case Presentation**: We incidentally found a protruding mucosal lesion in an asymptomatic 38-year-old man upon gastroscopy. The lesion was located in the upper gastric body, with mucosal plica disruption. An abdominal computed tomography scan showed that the lesion broke through the serous layer of the stomach and was adhered to the spleen. The patient underwent a complete resection (R0) by laparoscopy. Histology showed multiple intramural and subserosal nodules with characteristic plexiform growth featuring bland spindle cells situated in an abundant myxoid stroma with low mitotic activity. Immunohistochemistry was negative for CD117, Dog1, CD34, and S100and positive for Vimentinand SMA. CD10 staining was partially positive. The tumor was benign, and thus far, neither recurrence nor metastasis has been reported.

**1.3. Conclusions**: Clinicians should be aware of this rare tumor to avoid misdiagnosis. In the differential diagnosis of plexiform fibromyxoma, it is important to exclude the more common gastrointestinal stromal tumors, gastric carcinoma, neuronal and vascular tumors, inflammatory fibroid polyps, abdominal desmoid-type fibromatosis, solitary fibrous tumors, and smooth muscle tumors.

**1.4. Background:** Plexiform fibromyxoma(PF) is a rare type of mesenchymaltumor. It was first reported in 2007 by [1] as a plexiform angiomyxoidmyofibroblastic tumor (PAMT), but has since been classified as a gastrointestinal mesenchymal tumor by the 2010 World Health Organization Classification of Digestive System Neoplasms [2] and is now termed plexiform fibromyxoma. Most cases of PF occur in the gastric antrumor pylorus regionsand rarelyoccur in the duodenum [3]. We report a case of PF in theupper gastric body that was incidentally discovered as a mucosal protruding lesion with mucosal plica disruption on gastroscopy. The lesion has not recurred during 6 months of follow-up.

## 2. Case Presentation

A routine gastroscopy (health examination) was performed on an asymptomatic 38-year-old man, during which a protruding mucosal lesion with plica disruption was incidentally found in the upper gastric body. The lesion surface was covered with mucus and was difficult to wash. The surface of the lesion consisted of green blood vessels (Figure 1). The initial biopsy was diagnosed as chronicinflammation. Upon endoscopic mucosal resection(EMR), we found that it was difficult to elevate the lesion via submucosal injection. Computedtomography (CT) of the abdomen showed a gastric lesion that went through the gastric serosa and attached to the spleen (Figure 2). The patient underwent a complete resection (R0) by laparoscopy. The size of the resected tumorwas approximately 3.0 × 2.5cm. The lesion surrounded the splenic artery and the short gastric artery (Figure 3). The biopsy was suspicious forsubmucousal inflammatory myofibroblastoma (IMT) by hematoxylin and eosin and immunohistochemical staining (Figure 4a). Postoperative pathological examination confirmed the presence of proliferative spindle cells from the lamina propria to the serosa layers of the stomach (Figure 4b). Upon immunohistochemistry, the spindle cells were found to be negative

for CD117, Dog1, CD34, and S100and positive for Vimentin and SMA (Figure4c-4f). Immunohistochemistry revealed partial positivity for CD10.The Ki67 labeling index was approximately 1%, and no vascular invasion was observed. The results supported the diagnosis of gastric plexiform fibromyxoma.

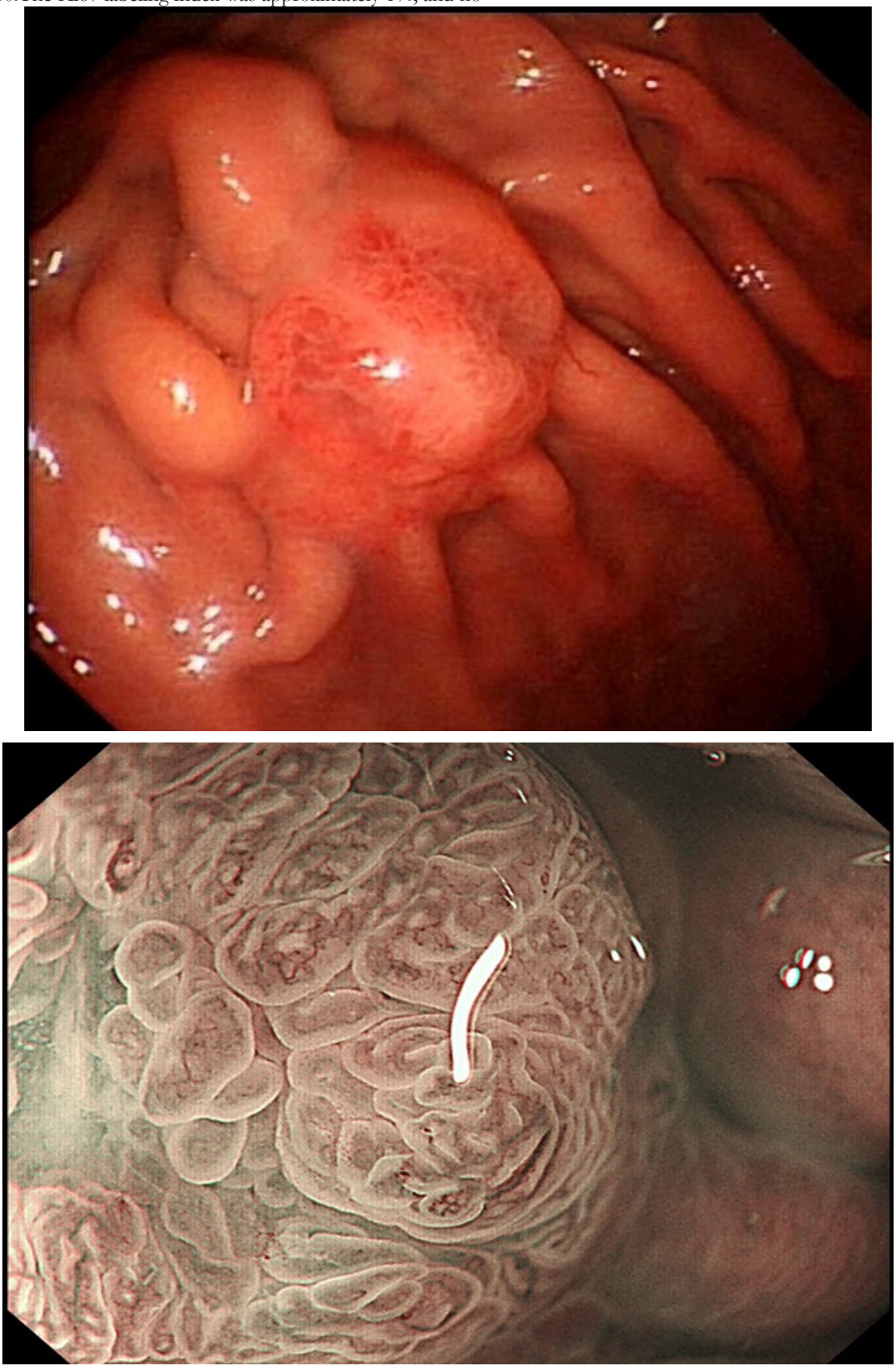


**Figure1** a: White light endoscopic (WLE): Protuberant lesion, covered with mucus and disruption of peripheral plica. b: Magnifying-narrow band imaging (ME-NBI): abnormal microvasculature, green blood vessels on the surface of the lesion, light blue crest (LBC)-positive.

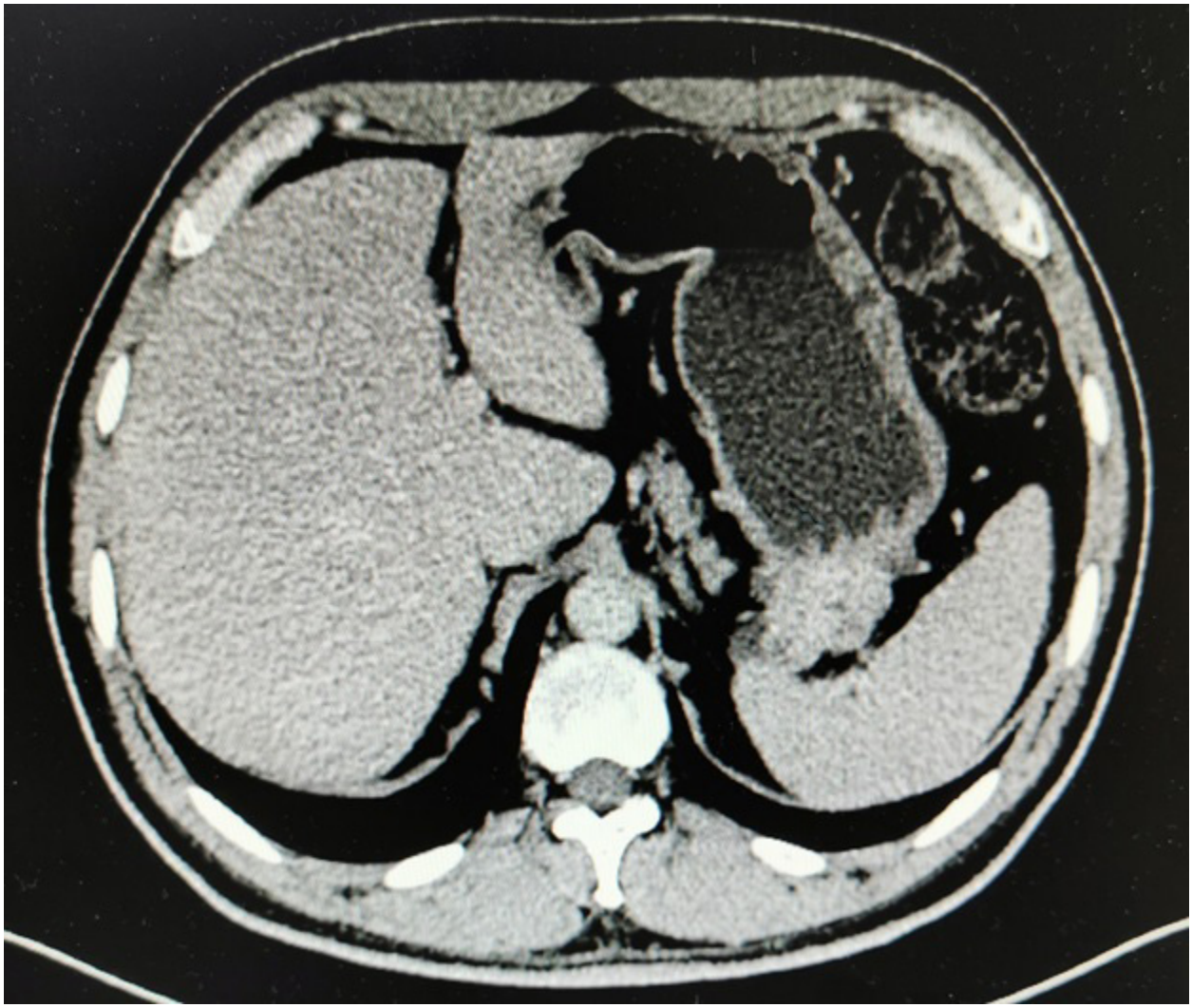
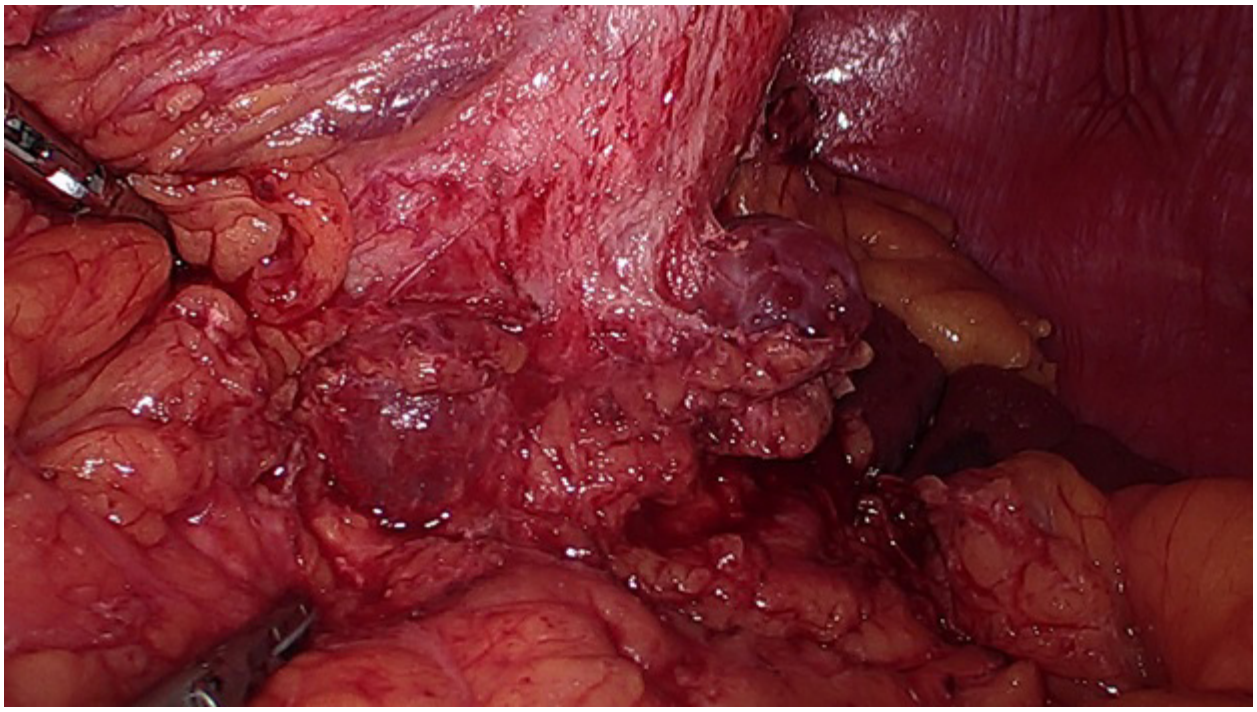


**Figure 2:** The abdominal computed tomography (CT) revealed an indistinct boundary between the gastric lesion and the spleen.



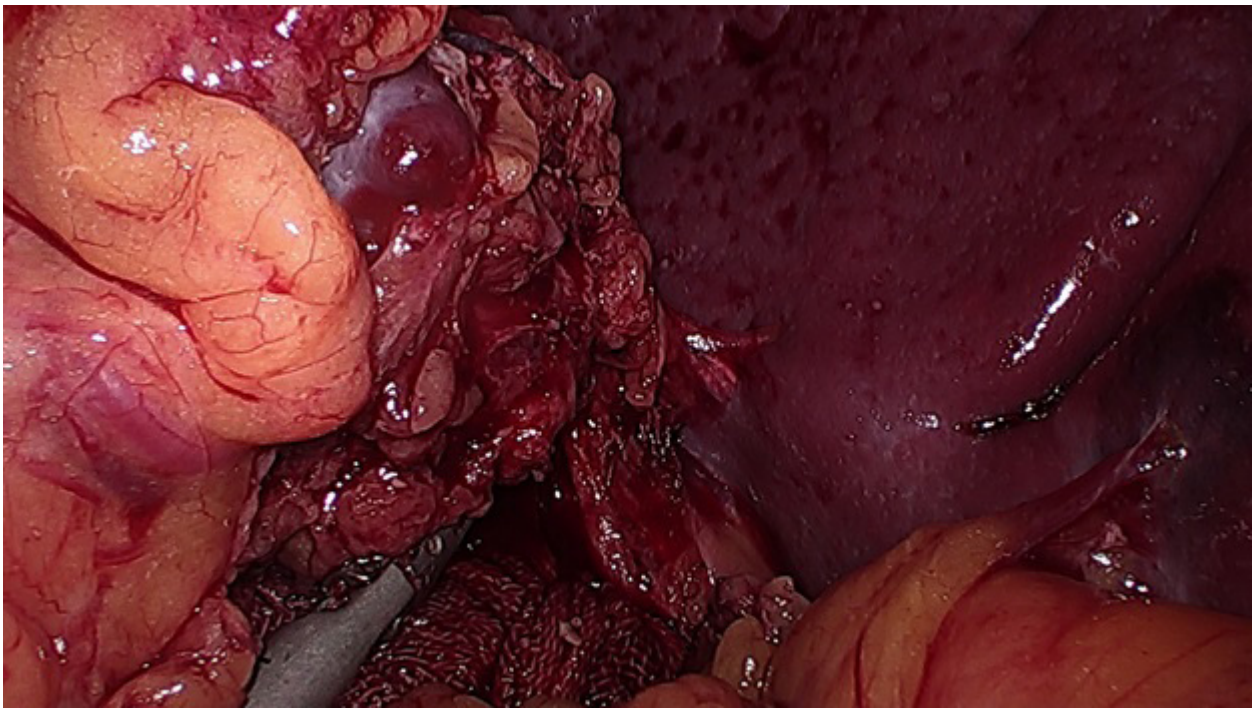
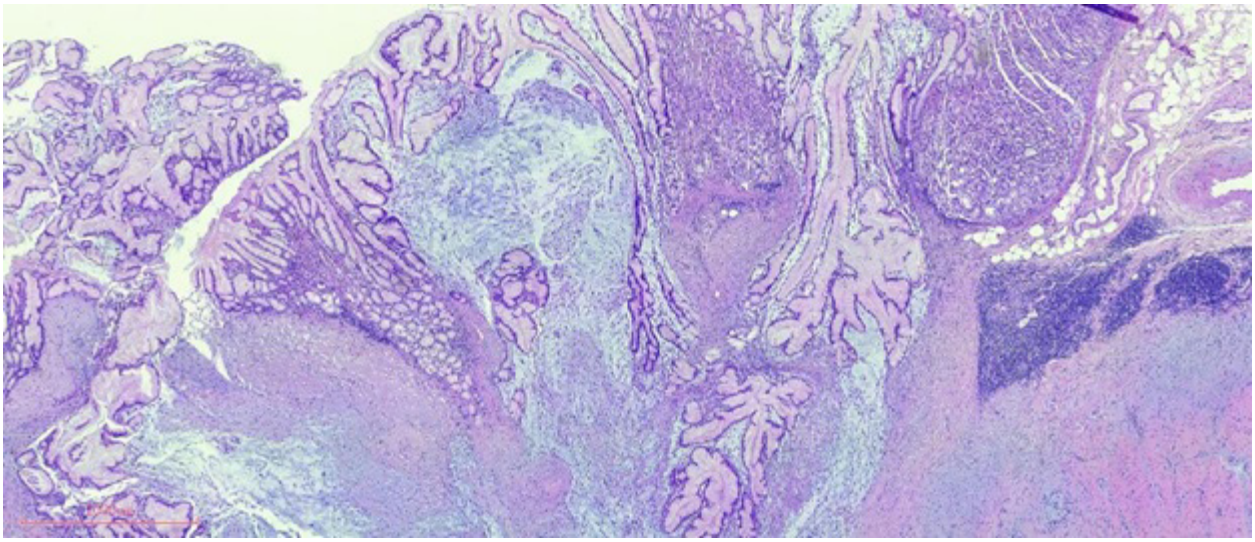
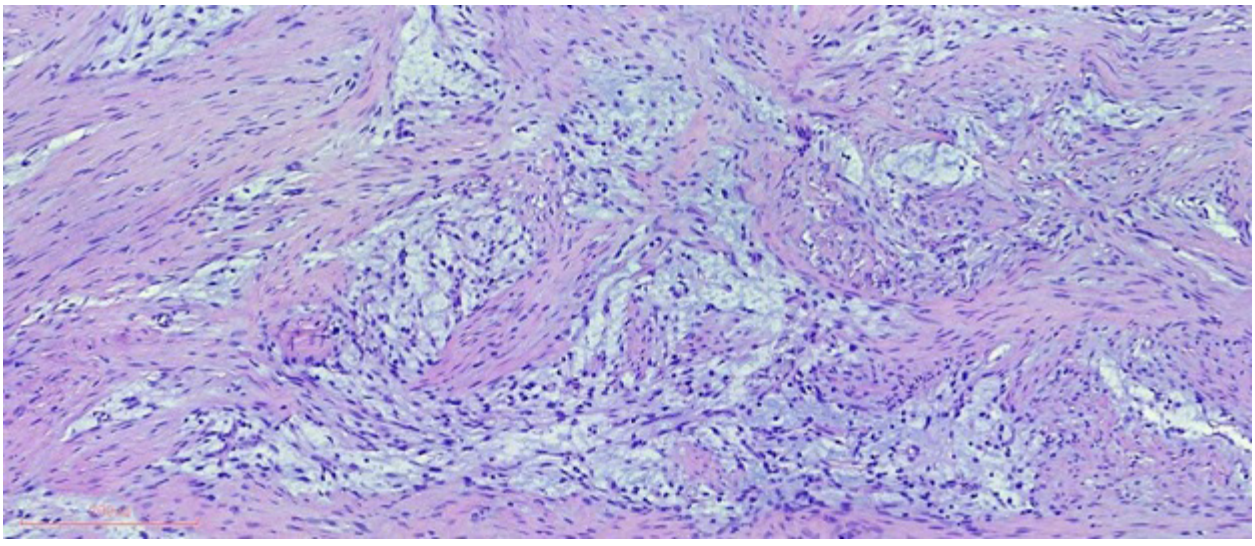
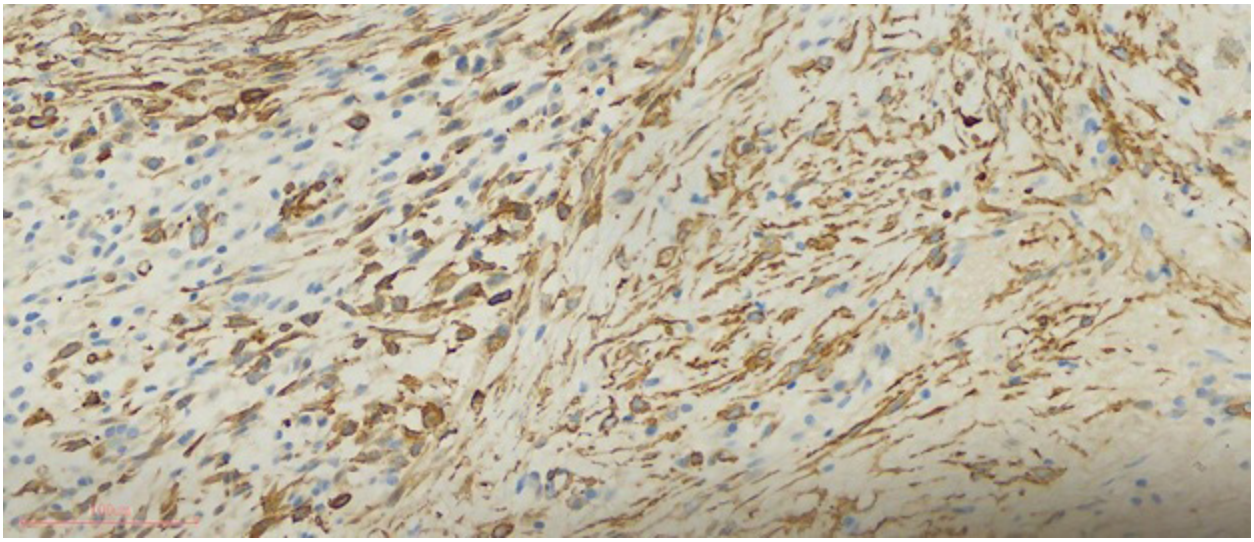
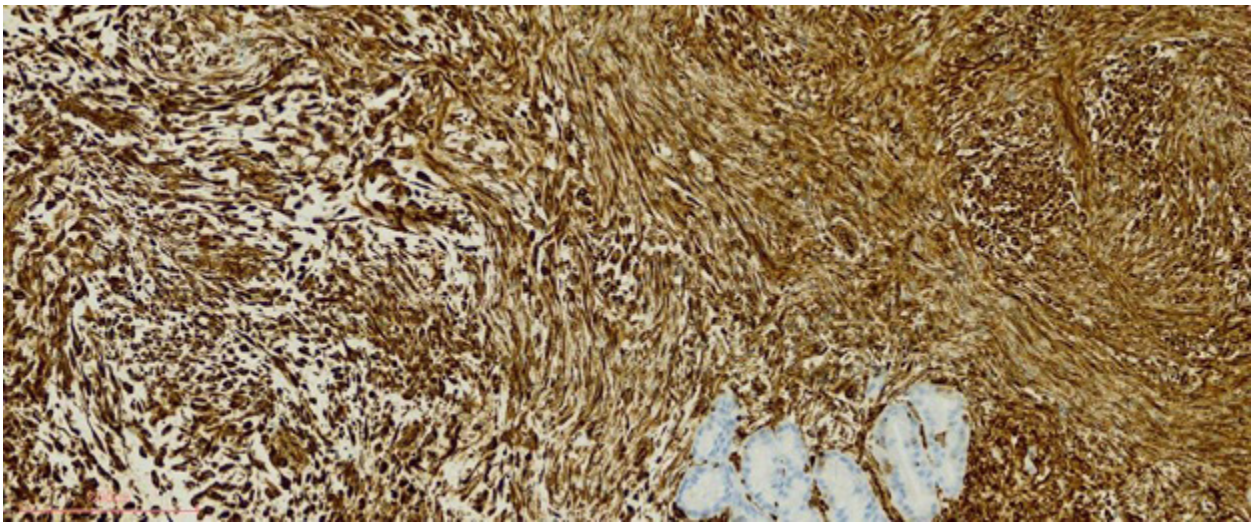


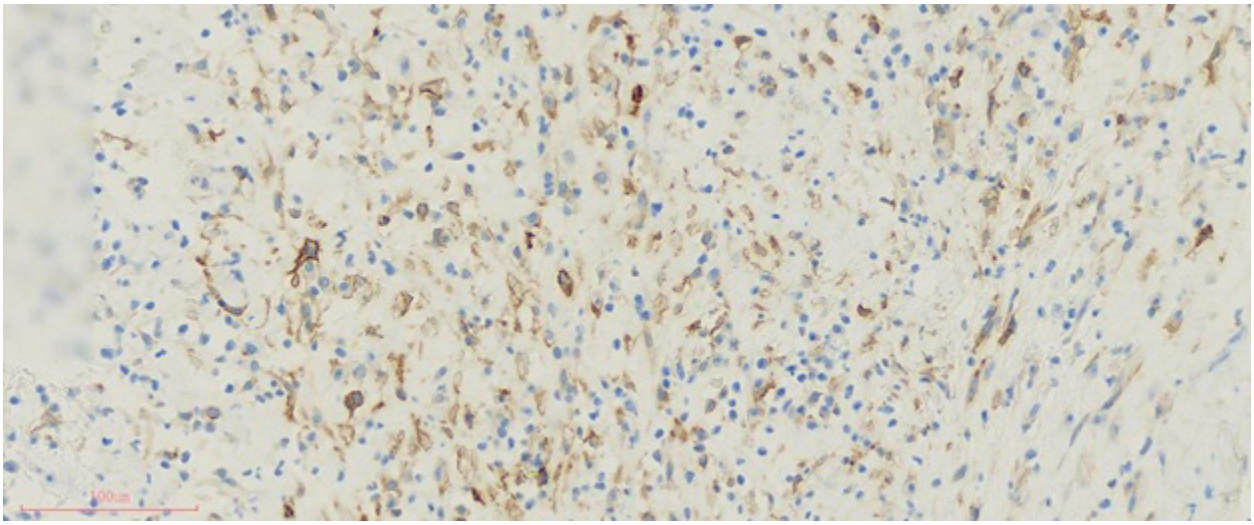
**Figure 3:** a:The splenic artery is surrounded by the lesion.b: The short gastric artery is surrounded by the lesion.











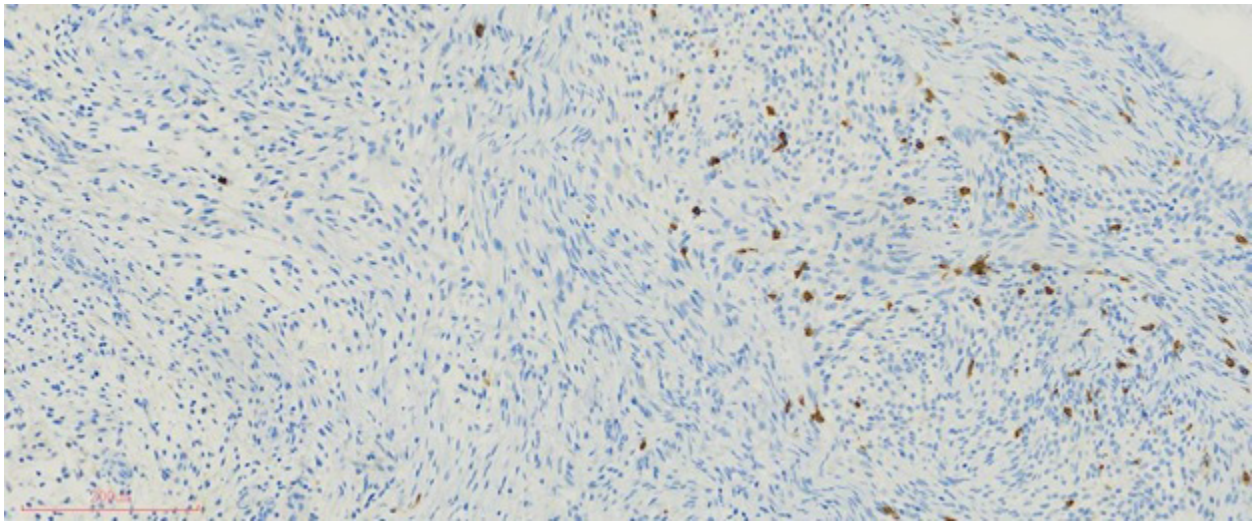


**Figure 4** a: Proliferation of bland myofibroblastic cells(He×20). b: Spindle-shaped bland tumor cells were separated by an abundant intercellular myxoid or fibromyxoid matrix(HE×100). c-e: Immunohistochemical analysis showing positive staining of SMA (×200), Vim (×100), CD10(×200). f: Immunohistochemical analysis showing negative staining of CD117(×100). Abbreviations: HE: hematoxylin and eosin; SMA: smooth muscle actin; Vim: Vimentin.

## 3. Discussion and Conclusions

PF occurs in a broad age range of patients (5-81 years, mean age 43 years), with no difference in the incidence rate between men and women [4-6]. The tumor is most often found in the gastric antrum, followed, in decreasing order, by the gastric body, gastric fundus, duodenum, jejunum, gallbladder, and the mediastinum [4,7]. PF are typically pink or reddish, glistening tumors, elastic in texture, and covered with ulcerative, erosive, or smooth mucosa [6]. They mostly originate from the submucosa and muscular is propriaand can extend from the mucosa to the serosa, causing ulcersor perforation. In a report summarizing 76 cases, the tumor surface was found to be ulcerated in 50 cases (65.8%), no nulcerated in 22 (28.9%), and eroded mucosa in 4 (5.3%) [8,9]. In this case, the lesion appeared to be an adenomatous protuberance with mucosal plica disruption and green vessels with villous white zones on the surface; it looked like gastric cancer. The pathological result of the first biopsy was chronic inflammation. The disruption of the folds around the mucosal bulge suggested that the lesion was not limited to the mucosal layer. Enhanced CT showed progressive heterogeneous enhancement of the lesion and revealed thatit was closely related to the spleen. EMR resection was performed to further clarify the pathological diagnosis, and the endoscopic findings were consistent with the CT findings. A diagnosis of PF depends on the pathological and immunohistochemical examinations, and cannot be achieved by clinical examinations alone [10]. PF has a characteristic plexiform pattern. Immunohistochemically, PFis almost always positive for vimentin and SMA, and variably positive for desminand CD10.The most important differential diagnosis of PF is gastrointestinal stromal tumor (GIST) and IMT.A plexiform pattern is highly unusual for GIST. Approximately 95% of GISTs are positive for CD117, 60% to 70% are positive for CD34, 30% to 40% are positive for smooth muscle actin, 5% are positive for S100 protein, and 1% to 2% are positive for desmin. There is evidence that protein kinase C-theta antibody may also be useful to differentiate GISTs from GIST mimickers [11]. Gastric IMT is a rare tumor. IMTs are generally actin positive and may also show staining for desmin and cytokeratin. Immunohistochemical staining of anaplastic lymphoma kinase (ALK) [12] can contribute to the identification of IMT and PF. Approximately 50% of IMTcases are positive for ALK (13).

The main treatment of PF is surgical removal, while medical treatment serves an assistant role for symptomatic management. Tumor resection is considered to be effective [14]. To date, no cases with local recurrence or distal metastasis after resection have been reported, except for abdominal dilatation and vascular invasion. In this case, the tumor surrounded the short gastric and splenic arteries. PF is a rare mesenchymal tumor with increasing clinical attention. At present, PF is reported to occur in the stomach, duodenum, and small intestine. The diagnosis of PF relies on a pathological examination, and it should be distinguished from other gastrointestinal mesenchymal tumors. PF is benign, and local recurrence or distant metastasis has yet to be reported [15].

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