Journal of Biosciences and Medicines, 2025, 13(7), 217-225
https://www.scirp.org/journal/jbm
ISSN Online: 2327-509X
ISSN Print: 2327-5081

Misdiagnosis of Gastric Plexiform Fibromyxoma as Gastrointestinal Stromal Tumor: A Case Report

Lanquan Li, Xiaoxia Tan, Liangping Luo*

Department of Medical Imaging Center, The First Affiliated Hospital of Jinan University, Guangzhou, China
Email: *tluolp@jnu.edu.cn, 1292559456@qq.com, 2223650937@qq.com

How to cite this paper: Li, L.Q., Tan, X.X. and Luo, L.P. (2025) Misdiagnosis of Gastric Plexiform Fibromyxoma as Gastrointestinal Stromal Tumor: A Case Report. *Journal of Biosciences and Medicines*, **13**, 217-225.
https://doi.org/10.4236/jbm.2025.137016

Received: June 5, 2025
Accepted: July 14, 2025
Published: July 17, 2025

Abstract

Plexiform fibromyxoma is a rare primary mesenchymal tumor of the gastrointestinal tract, first reported by Takahashi in 2007 and officially recognized as a subtype of gastric mesenchymal tumor by the World Health Organization (WHO) (Classification of Tumors of the Digestive System, 4th ed.) in 2010. The disease is prevalent in the gastric sinus and pyloric region, and common clinical manifestations include gastrointestinal bleeding, ulceration and secondary anemia. Due to the extremely low incidence of PF and the lack of specificity in imaging, it is easily misdiagnosed as GIST, smooth muscle tumor, nerve sheath tumor, and sclerofibromatosis before surgery. Less than 200 cases of PF have been reported in the literature, and there is a lack of systematic analysis of cases with atypical clinical manifestations (e.g., acute attacks, severe anemia, etc.). We report a case of a 31-year-old female patient with acute onset of severe anemia due to bleeding from a ruptured tumor. CT of the upper abdomen suggested cystic solid occupancy in the gastric antrum, and the possibility of gastrointestinal mesenchymal tumor was considered. The patient underwent tumor resection and postoperative pathology confirmed the diagnosis of tufted fibromucinous tumor. The patient recovered well after surgery and is currently undergoing long-term follow-up. This case highlights the diagnostic challenges associated with PF, particularly when presenting atypically, and underscores the importance of multidisciplinary evaluation to avoid misdiagnosis and ensure timely management.

Keywords

Plexiform Fibromyxoma, Gastrointestinal Stromal Tumor, Benign Gastric Neoplasm, Tumor Rupture, Multidisciplinary Diagnosis, Gastric Mesenchymal Tumor

1. Introduction

Plexiform fibromucinous tumor (PF), also known as plexiform angio-mucinous-like myofibroblastoma (PAMT), is a new pathological mesenchymal tumor category advocated in the literature by Miettinen *et al*. [1] [2]. PF is a rare benign gastric mesenchymal tumor, and, to date, although it has been reported to have a histology with an infiltrative growth pattern with infiltration of the mucosa and blood vessels, there has been no over mortality or distant spread.

The age range of patients with gastric PF ranges from 5 to 81 years, and it is most common in middle-aged adults, with peak incidence between 30 and 60 years of age, and there is no significant difference in the prevalence between men and women [3]. Typical clinical manifestations of PF are gastrointestinal symptoms with a slow onset and a lack of specificity, such as abdominal pain, abdominal distension, and abdominal discomfort. According to the literature, the tumor size of plexiform fibromucinous tumor (PF) varies widely, ranging from 1.9 to 15 cm in diameter, with an average of about 5.5 cm [4]. The most commonly involved site is the gastric antrum, accounting for about 80% of cases, with the pyloric region being the most common. About 20% to 50% of cases may involve the perigastric soft tissues or duodenal bulb, and in a few cases, the esophagus, duodenum, jejunum, gallbladder, mediastinum and colon are also involved [3] [5]-[8]. Gastric mucosal erosion or ulcer formation is seen in about half of the patients, leading to the risk of gastrointestinal bleeding and secondary anemia [9] [10]. Our patient was characterized by acute onset, severe anemia (hemoglobin 38 g/L), and tumor rupture, and the clinical presentation was clearly different from the typical course of slow progression in most patients with PF [10]-[12].

In this article, we discuss the clinical and imaging features of a case of gastrointestinal sinus PF with acute onset combined with severe anemia in the context of a multidisciplinary diagnostic process to provide a differential diagnosis of atypical PF in order to avoid misdiagnosis and ensure appropriate treatment.

2. Case Report

2.1. Medical History

A 31-year-old female was admitted to the hospital with "abdominal pain and diarrhea with fever for 4 days". The abdominal pain was subxiphoid colic, accompanied by nausea and vomiting, watery stools (3 - 5 times/day), and the highest temperature was 37.8˚C. She was diagnosed as "gastrointestinal dysfunction combined with moderate anemia", and was given antidiarrheal and anti-infective treatments that were ineffective. In order to seek treatment, she was admitted to the outpatient clinic of our hospital. She was admitted to the hospital for examination: bedside DR chest radiographs showed inflammatory exudation in the lungs, and severe pneumonia was considered in conjunction with the clinical findings; her hemoglobin level was significantly reduced to 38 g/L, and she was given anti-infective treatment, blood transfusion and other treatments. Past history: history of thalassemia and iron deficiency anemia, history of blood transfusion 7

years ago. In this case, due to recurrent episodes of abdominal pain accompanied by a sharp drop in hemoglobin from a stable baseline value to 38 g/L, an enhanced CT examination of the abdomen was performed to exclude abdominal hemorrhage and to clarify the nature of the space-occupying lesion. Given the patient's underlying chronic anemia, her hemoglobin level was already below the normal range, and the significant and dramatic decrease in hemoglobin far exceeded the expected fluctuation of her underlying disease, these underlying diseases were considered as key influencing factors and potential confounders in the initial evaluation, and life-threatening etiologies, such as acute blood loss or new-onset hemolysis, needed to be ruled out as a matter of priority.

2.2. Image Performance

Our CT showed that a cystic solid occupancy was seen in the gastric antrum, with unclear border, growing outward toward the lumen, localized uneven thickening of the gastric wall, and slightly blurred surrounding fat interstitial space; the size of the lesion was about 12.3 cm × 7.3 cm × 9.9 cm, with visible segregation within it, and the segregation was unevenly thick and thin, among which the solid component was about 4.4 cm × 3.1 cm × 4.7 cm in size, and enhancement scanning enhancement of the lesion was uneven, with mild enhancement of the solid component during the arterial phase, The solid component had mild enhancement in the arterial phase and progressive enhancement in the venous and delayed phases, and no enhancement was seen in the cystic component (Figure 1). A few fluid density shadows were seen in the abdominal cavity. The main diagnosis of the image: exophytic cystic-solid occupancy in the gastric sinus, considering the possibility of gastric sinus mesenchymal tumor, abdominal fluid accumulation and peritonitis, can not be excluded from the possibility of local rupture and encapsulation of the tumor.

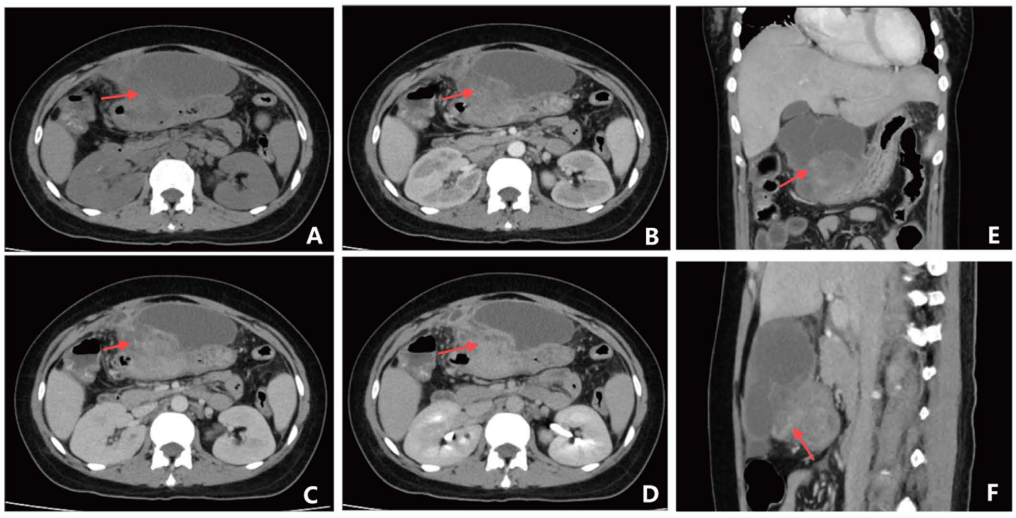

Figure 1. Scanning axial (A): exophytic cystic-solid occupancy in the gastric antrum (12.3 cm × 7.3 cm × 9.9 cm), enhancement: arterial-phase axial (B), venous-phase axial (C), delayed-phase axial (D), reconstructed coronal (E), and sagittal (F): the solid component (4.4 cm × 3.1 cm × 4.7 cm) showed inhomogeneous enhancement, with blurring of the surrounding fat interstices.

2.3. Endoscopy

The mucosa and morphology of the gastric fundus and gastric body were generally normal, and a huge spherical mass was seen in the gastric antrum, with a size of about 10 cm × 8 cm, and two depressions were seen on the surface of the mass, with thin white moss in the oral side of the depression, and a small amount of blood oozing in the anal side of the depression, and two pieces of clamped tissues were sent to the pathology (Figure 2), which did not go into the pylorus and duodenum (Pathology suggests chronic gastritis with ulcer). Because the endoscopic pathology did not clarify the nature of the tumor and imaging highly suspected a mesenchymal tumor, we ultimately decided to perform surgical exploration.

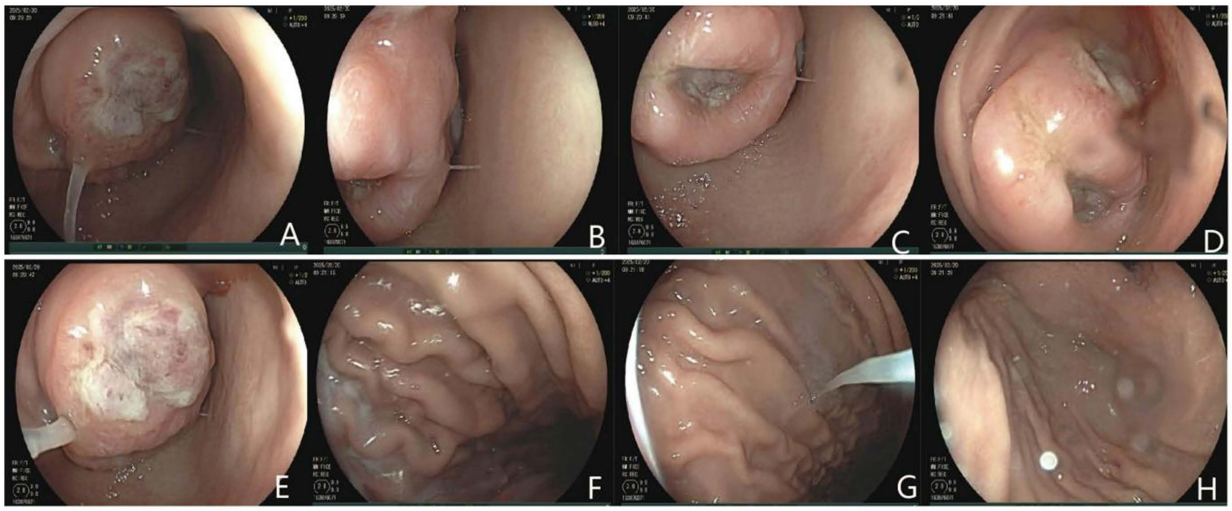

Figure 2. A large spherical mass (10 cm × 8 cm) was seen in the gastric antrum with surface ulceration with blood oozing.

2.4. Intraoperative

The patient underwent laparoscopic exploration with intermediate open gastric tumor resection + abdominal adhesion release. A longitudinal incision of about 1.0 cm was made through the umbilicus, a pneumoperitoneum needle was inserted, CO_2 was injected, the abdominal pressure was maintained at 12 mmHg, and a 10 mm 30-degree laparoscope was inserted to investigate the abdominal cavity: the mass was located in the gastric sinus, the upper abdominal cavity was filled with cystic encapsulated foci, and the tumor peritoneum was ruptured, with the size of 15 cm × 25 cm, and the pelvis was visible as a bloody fluid, and the space of the abdominal cavity was extremely small, so it was decided that it should be changed to an open surgery. A longitudinal incision of about 15 cm in length was made in the upper abdomen, and the abdomen was entered layer by layer, with a protective sleeve placed over the incision. The omentum was adherent to the upper abdominal wall, and the adhesions were carefully separated, and the tumor peritoneum was ruptured with bloody fluid, which was suctioned to stop the bleeding. Subsequently, the stomach was lifted upward and outward with a

non-invasive grasping forceps, and the ultrasonic knife was used to incise the avascular area within the gastric omental arch, severing the gastrocolic ligament to the left to the middle of the stomach (preserving 2 - 3 short gastric vessels) and to the right to the duodenal bulb. The peritoneum in front of the hepatoduodenal ligament is incised and the lesser omentum is separated to the middle of the stomach. The upper part of the duodenum was freed and closed at the pylorus with a purse-string clamp, a purse-string needle was sutured in, the proximal stomach was Kocar-clamped and severed, and a Johnson & Johnson 25-mm base nail holder was placed and the purse-string tightened. Subsequently, a small opening of about 2 cm in diameter was made in the anterior wall of the middle part of the stomach, and a tube-type anastomosis was placed to perform a gastro-duodenal anastomosis (Billoth I-type anastomosis) on the side of the greater curvature. The encompassing gastric incision margin was closed with 3-0 absorbable suture, followed by interrupted suturing of the gastro-duodenal anastomosis with absorbable suture. The surgical wound was rinsed with plenty of distilled water without active bleeding, no anastomotic leakage, no intestinal leakage, and the rinsing fluid was aspirated. One right subhepatic and one right pelvic drain were left in place. After counting the surgical instruments and accessories, each Trocar was removed under direct vision, the gas was drained, and the incisions were closed with sutures to end the surgery. When the specimen was dissected in vitro, the tumor was located in the gastric sinus, with exophytic growth, cystic and jelly-like mass on the cut surface, peripheral tumor rupture of the periphery was seen, and the tumor periphery was ruptured, with bloody fluid outflow (**Figure 3(A)**).

2.5. Pathology and Immunohistochemical Analysis

Under the microscope, the tumor showed clumped and nodular growth, with mild spindle-shaped cells distributed in a large number of mucus-like and fibromucus-like stroma, with unequal cell density, alternating cell-rich and cell-sparse areas, and nuclear schizophrenia, and abundant branching capillaries with hemorrhage and cystic degeneration in the interstitium; the tumor involved mucous layer to the outside of the plasma membrane layer, and the cut edges of the two ends did not show any special features; 1 lymph node was detected in the periosteal tissue and showed reactive changes; 4 lymph nodes were detected in the perigastric region, and no special features were seen. One lymph node was found in the omental tissue, showing reactive changes; four lymph nodes were found in the perigastric area, with no special features (**Figures 3(B)-(D)**).

SMA (+), CD99 (partially +), CD117 (−), DOG-1 (−), S-100 (−), SOX-10 (−), STAT6 (−), Desmin (−), ALK (−), EMA (-), MDM2 (−), HMB45 (−), P-CK (−), β-catenin (−), CD34 (−), Ki-67 (about 5%+, suggesting low tumor cell activity). Combined with histomorphology and immunohistochemistry, plexiform fibromucinous tumor was considered. Tumor cell proliferation was more active in some areas, and molecular testing was recommended except for low-grade malignant fibromucinous sarcoma. (**Figure 3(E)**, **Figure 3(F)**).

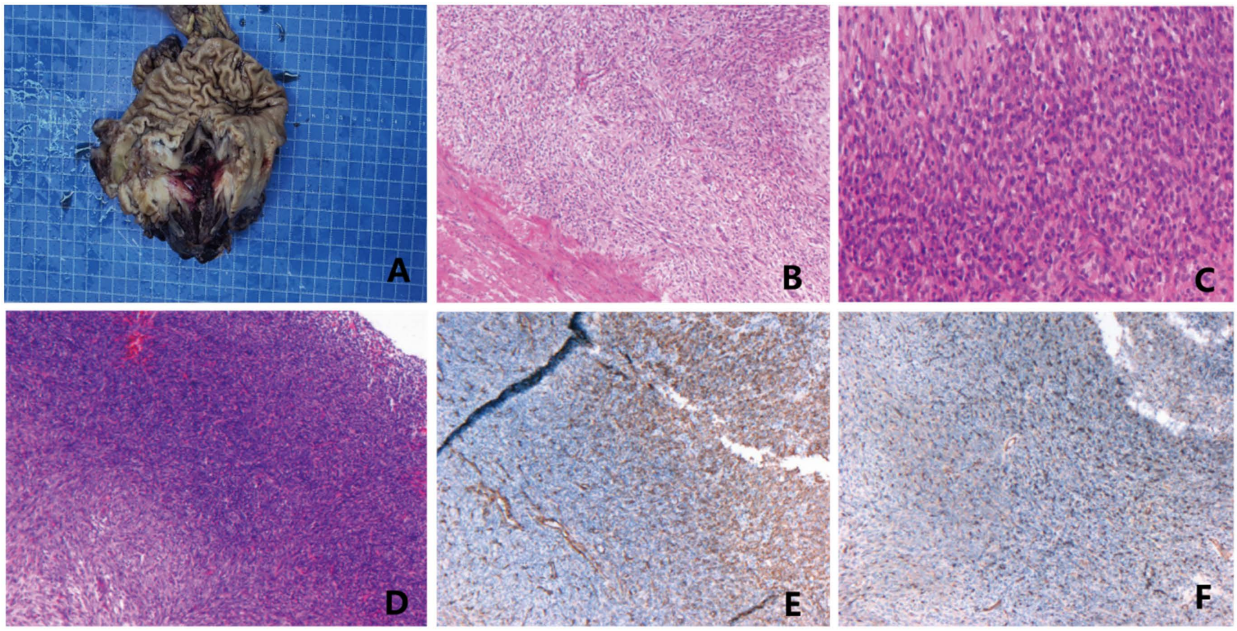

Figure 3. (A) A piece of gastric tissue measuring 13 cm × 6.5 cm × 3 cm, with a small curvature of 9 cm and a large curvature of 14 cm, and an ulcerated mass was seen; (B) The tumor consisted of spindle cells with a mild morphology distributed within a large amount of mucus-like and fibrous mucus-like stroma; (C) Areas of cellular abundance alternated with areas of cellular sparseness; (D) The tumor consisted of spindle cells with a clarified cytoplasm accompanied with a mucus-like stroma and dendritic vessels (hematoxylin/eosin staining); (E), (F) Immunohistochemical staining results of SMA (+) and CD99 (partially +).

3. Discussion

Among the ancillary investigations of gastric PF, imaging (CT/MRI) and endoscopy have been important diagnostic tools in most case reports. Endoscopy often shows submucosal tumor-like elevations, some of which are accompanied by surface ulcers, and ultrasonography suggests a multinodular mass. The imaging manifestations of gastric PF have been reported in the literature as well-defined solid, cystic, or cystic-solid masses. On CT-enhanced examination, the solid portion of the tumor is seen to be mildly enhanced in the arterial phase, with progressive enhancement in the venous and delayed phases [7]. This enhancement feature in the delayed phase is thought to be related to the tumor being rich in mucus-like stroma and vascularity, which is a typical imaging manifestation of PF [13]. Recent studies have pointed out that MRI has certain advantages in displaying tumor components. Gastric PF shows low signal on T1-weighted imaging and high signal on T2-weighted imaging, and the solid component also shows progressive enhancement after enhancement [7]. CT examination of our patient showed an exophytic cystic-solid mass in the gastric antrum, and the solid component strengthened significantly in the delayed phase. However, due to the rupture of the tumor, accompanied by pelvic and abdominal fluid, and blurred tumor boundaries, the initial diagnosis was mistaken for gastrointestinal mesenchymal stromal tumor (GIST) because of the inconsistency of the typical manifestation of "clear boundaries" in the literature.

To date, reports on the imaging features of gastric plexiform fibromucinous tumor (PF) are still limited, and most of the endoscopic and imaging presentations in the existing literature lack specificity. PF often resembles other gastric mesenchymal tumors (especially gastrointestinal mesenchymal stromal tumors, GISTs) in terms of site of onset, imaging morphology, and macroscopic and microscopic features, which makes the differential diagnosis challenging. Statistical data show that the incidence of GIST is about 150 times higher than that of PF [2] [3]. In clinical practice, when occupying lesions in the gastric antrum area are detected, doctors usually consider more common gastric mesenchymal tumors, such as GIST, firstly based on the empirical "common is reasonable" imaging thinking [14]. However, if certain subtle differences, such as enhancement patterns, are overlooked, it may lead to misdiagnosis. In this case, the tumor showed a cystic solid mass with delayed enhancement on CT, which suggested a mesenchymal origin, but due to atypical manifestations such as tumor rupture, blurred borders, and pelvic and abdominal effusion, the initial diagnosis was inclined to GIST, which was clarified by postoperative pathology [15] [16].

Definitive diagnosis of PF relies on histopathologic and immunohistochemical analysis. Microscopically, a typical tufted growth pattern, rich in mucus-like stroma with varying degrees of vascular richness, is seen. Immunohistochemically, the most common positive markers include smooth muscle actin (SMA) and waveform protein, as well as CD10, junctional proteins, calmodulin, etc., which show some degree of mixed staining characteristics [8]. Notably, GIST-specific markers such as CD117 and DOG1 are usually negative, which helps in differentiation. The immunohistochemical results of our patient showed SMA (+) and CD117 (−), contrary to the typical expression pattern of GIST, which, combined with the histologic pattern, led to a definitive diagnosis of PF. The current literature suggests that distal gastrectomy or partial gastrectomy is the mainstay of treatment for PF, and the extent of the resection may include wedge or localized resection [4]. For smaller lesions, complete endoscopic resection may also be considered [17]. Unlike GIST, no adjuvant therapy is usually required after PF, and no cases of definite recurrence or metastasis have been reported. In this case, the patient underwent laparoscopic exploration for tumor rupture with intermediate open gastric tumor resection and abdominal adhesion release, and had a good postoperative recovery. As of the 2-month postoperative follow-up, the patient had no recurrence or metastasis, and the clinical outcome was consistent with the literature.

4. Conclusion

The diagnosis of PF often relies on multidisciplinary collaboration: imaging helps to suggest the morphologic features of the tumor, histopathology provides the typical tufted growth pattern, and immunohistochemical findings-such as SMA positivity and CD117 negativity-help to exclude other mesenchymal tumors, leading to a definitive diagnosis. Although PF is a rare mesenchymal tumor of the gastrointestinal tract, its clinical awareness has gradually increased in recent years,

and studies have shown that it can occur in sites other than the gastrointestinal tract. The case reported in this study further highlights the complexity and non-specificity of the imaging manifestations of PF, emphasizing the need to be vigilant for atypical gastrointestinal sinus tumors in clinical practice and to avoid adhering to the conventional imaging diagnostic pathway. This case reminds clinicians that PF should be included in the differential diagnosis when encountering abnormal manifestations such as unclear borders with rupture and bleeding. In addition, tumor rupture, as an important feature of the acute onset of the disease in this case, may reflect the potentially aggressive biological behavior of PF, and future studies may further explore whether tumor rupture is a potential indicator of its progression or poor prognosis.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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