

Antral plexiform fibromyxoma: case report of a rare mesenchymal neoplasm

Fibromixoma plexiforme antral: reporte de caso de una neoplasia mesenquimal rara

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ABSTRACT

Plexiform fibromyxoma (PF) is a rare mesenchymal neoplasm of the stomach usually arising in the gastric antrum, and its main differential diagnosis is gastrointestinal stromal tumor. Most common symptoms are hematemesis, anemia. Immunohistochemically, positivity for smooth muscle actin (SMA) and vimentin suggests the diagnosis of PF. We report the case of a 56-year-old female patient with a 30-day history of nausea at presentation 4 years ago. Gastroscopy at that time revealed a subepithelial lesion (SEL) in the gastric antrum, measuring approximately 20 mm in diameter, with leakage of serous fluid after biopsy. Histopathology showed only an inflammatory process. Follow-up gastroscopies were performed 24, 36, and 48 months later, with surveillance biopsy at each follow-up. The last gastroscopies showed changes in lesion appearance, reduction in size, and absence of fluid leakage. Histopathology showed bland spindle cell proliferation, with a vaguely plexiform/multinodular pattern, in a fibromyxoid stroma with an arborizing capillary network without mitoses. The tumor cells were positive for SMA and negative for DOG1, CD117, CD34, S100, desmin, EMA, CD10, calponin, and beta-catenin. The choice of treatment and follow-up depends on the SEL features, but because no cases of malignancy or metastatic disease have previously been reported, the patient chose a conservative approach.

Keywords: *Fibromixoma; Gastrointestinal stromal tumors; Gastrointestinal neoplasms (source: MeSH NLM).*

RESUMEN

El fibromixoma plexiforme (FP) es una rara neoplasia mesenquimatosa del estómago que generalmente surge en el antro gástrico. Su principal diagnóstico diferencial es el tumor del estroma gastrointestinal. Los síntomas más comunes de los FP son hematemesis y anemia. Inmunohistoquímicamente, la positividad para actina del músculo liso (SMA) y vimentina sugieren el diagnóstico de FP. Presentamos el caso de una paciente de 56 años de edad que inicia su enfermedad hace 4 años con náuseas de 30 días de evolución. La primera gastroscopia reveló una lesión subepitelial (SEL) en el antro gástrico, de aproximadamente 20 mm de diámetro, con fuga de líquido seroso después de la biopsia. La histopatología mostró sólo un proceso inflamatorio. Se realizaron gastroscopias de seguimiento a los 24, 36 y 48 meses con biopsia de vigilancia en cada seguimiento. Las gastroscopias siguientes mostraron cambios en la apariencia de la lesión, reducción de tamaño y ausencia de fuga de líquido. La última histopatología mostró una proliferación blanda de células fusiformes, con un patrón vagamente plexiforme/multinodular, en un estroma fibromixóide con una red de capilares arborizantes sin mitosis. Las células tumorales fueron positivas para SMA y negativas para DOG1, CD117, CD34, S100, desmina, EMA, CD10, calponina y beta-catenina. La elección del tratamiento y el seguimiento depende de las características del SEL, sin embargo, por ser una enfermedad que no presentaba rasgos de enfermedad maligna o metastásica, el paciente eligió un mantener un enfoque conservador.

Palabras clave: *Fibromixoma; Tumores del estroma gastrointestinal; Neoplasias gastrointestinales (fuente: DeCS Bireme).*

INTRODUCTION

Plexiform fibromyxoma (PF) is a rare mesenchymal neoplasm most commonly arising in the gastric antrum, but it may also occur in the pylorus, duodenum, jejunum, colon, and biliary tract. PF affects adults of either sex

occurring over an age range of 40 to 50 years, although it may also occur in children^(1,2). Tumor size ranges from 0.3 to 17 cm⁽³⁾. PF presents as a solid, solid/cystic, or cystic mass on computed tomography (CT), magnetic resonance imaging (MRI) and Endoscopic ultrasound (EUS). Many of these lesions are misdiagnosed as gastrointestinal

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stromal tumor (GIST) because they appear as a lobulated submucosal mass, most commonly ulcerated, with a risk of gastrointestinal bleeding and consequent anemia. Most GISTs are positive for CD117, DOG1, and CD34⁽⁴⁾. Symptoms include abdominal pain and nausea, occasionally but PFs can also be found incidentally.

CASE REPORT

A 56-year-old white female patient presented 4 years ago to the Department of Digestive Endoscopy of Hospital Santa Casa de Caridade, in Bagé, southern Brazil, with a 30-day history of nausea preceded by relevant past medical history. Gastroscopy (LASEREO L590ZW, Fujifilm Co) at that time revealed a subepithelial lesion measuring approximately 20 mm in diameter, located in the posterior wall of the antrum, with leakage of serous fluid after biopsy (shown in Figure 1). Histopathology showed only a gastric inflammatory process, possibly because only superficial biopsies were performed and with the presence of *Helicobacter pylori*.

Endoscopic ultrasound (EUS) revealed a 2 cm heterogeneous hypoechoic mass with cystic components arising from the second layer of the gastric wall with no disruption of the proper muscle layer, suggested of ectopic pancreas (shown in Figure 2).

Follow-up gastroscopies performed 24, 36, and 48 months later showed a subepithelial lesion in the posterior wall of the antrum, with slight scar retraction in the center, measuring approximately 12 mm in diameter (smaller than the mass detected at index endoscopy), without leakage of serous fluid (shown in Figure 3).

Surveillance biopsy bite-on-bite with large capacity forceps was performed at each follow-up gastroscopy. Histopathology was inconclusive, except for the last follow-up, showing bland spindle cell proliferation, with a vaguely plexiform/multinodular pattern, in a fibromyxoid stroma with an arborizing capillary network and no mitoses identified (shown in Figure 4). Cellularity was generally low. Immunohistochemically, the tumor cells were positive for smooth muscle actin (SMA) and negative for DOG1,



Figure 2. Heterogeneous hypoechoic mass arising from the second layer of the gastric wall.

CD117, CD34, S100, desmin, epithelial membrane antigen (EMA), CD10, calponin, and beta-catenin (shown in Figure 4). These findings were suggestive of PF.

Because no cases of malignant transformation have previously been reported and given the features of this case, with a reduction in lesion size and no ulceration, and after discussing with the patient, a decision was made to maintain follow-up.

This case report was written in accordance with the World Medical Association Declaration of Helsinki. Informed consent was obtained from the patient.

DISCUSSION

Takahashi *et al.*⁽⁵⁾ first described it in 2007 as a plexiform angiomyxoid myofibroblastic tumor. In 2009, Miettinen *et al.*⁽⁶⁾ described 12 cases of the same tumor in the antrum and named it as PF, the terminology adopted in the 4th edition of the WHO Classification of Tumors of the Digestive System in 2010⁽¹⁾. The most common clinical features are hematemesis and anemia, and the patient may complain of abdominal pain and nausea or even be

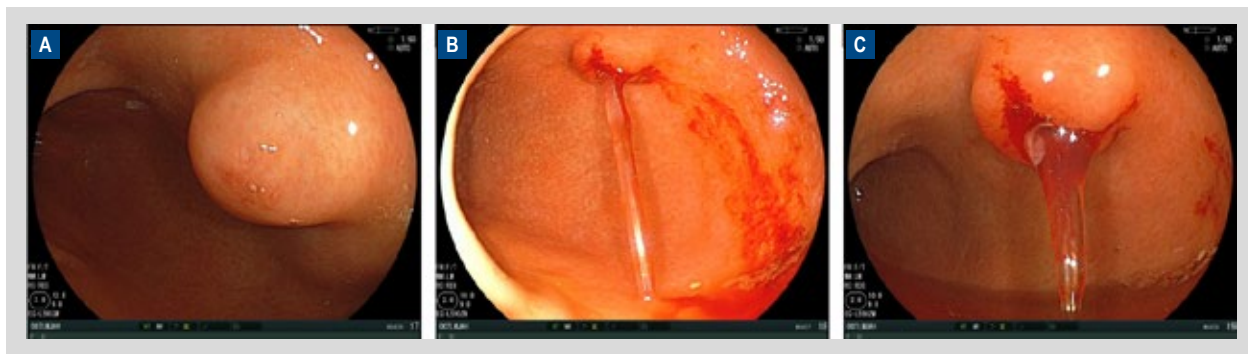


Figure 1. (A) Subepithelial lesion in the posterior wall of the antrum. (B and C) Leakage of serous fluid after biopsy.

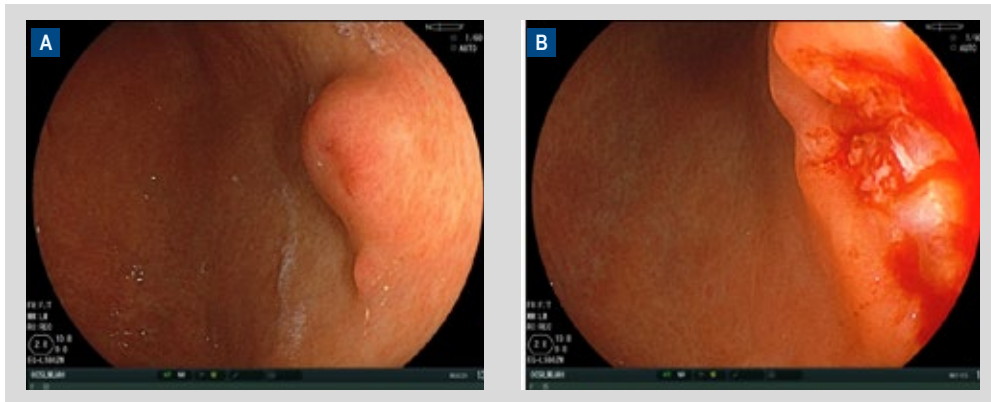


Figure 3. (A and B) Follow-up gastroscopy showing the same subepithelial lesion, with slight scar retraction in the center, smaller than the mass detected at index endoscopy, without leakage of serous fluid after biopsies.

asymptomatic. Our patient reported only nausea, showing improvement after proton pump inhibitor. Bleeding is usually associated with the presence of ulceration, with endoscopic findings suggestive of GIST, as irregular border and lobulated subepithelial mass, and occurs in about 50% of cases⁽³⁾. Histologically, PF is characterized by a plexiform growth pattern, ovoid to spindle cell proliferation, and a myxoid stroma rich in small blood vessels, without substantial cytological atypia and with low mitotic activity. Intravascular involvement has been described, suggesting a possible intravascular spread of the tumor within the gastric wall to the subserosa⁽⁶⁾.

The diagnosis is made by immunohistochemistry, with the tumor cells being positive for SMA and vimentin and negative for CD117, DOG1, CD34, S100, beta-catenin, STAT-6, and ALK. There was immunoreactivity for SMA in the case reported here, with the tumor cells being negative for all other immunostains. GIST is the most common mesenchymal tumor of the stomach and the main differential diagnosis of PF. GISTs are more aggressive than PFs and positive for CD117, CD34, and DOG1 stains, whereas schwannomas are positive for S100 and SOX10. Therefore, negative staining for CD117 and DOG1 excludes GIST, whereas negative staining for S100 excludes

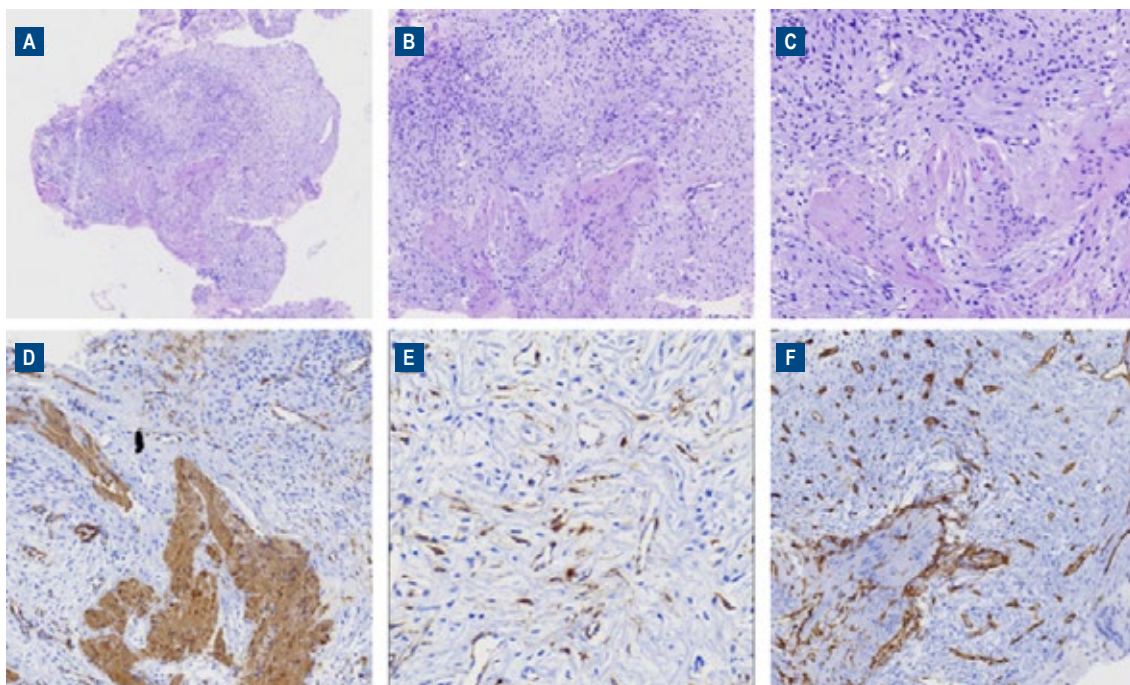


Figure 4. Histopathology and immunohistochemistry. (A, B and C) Presence of bland spindle cell proliferation, with a vaguely plexiform/multinodular pattern, in a fibromyxoid stroma showing an arborizing capillary network and absence of mitosis (hematoxylin and eosin). (D and E) The cells are positive for SMA (400x). (F) The cells are negative for CD34 (400x).

neural tumors. Vascular lesions are positive for CD34 and CD31^(4,7). EUS often shows a hypoechoic mass arising from the muscle layer. Fine-needle aspiration can either support or mislead the diagnosis. A meta-analysis evaluating EUS tissue acquisition for upper GI SELs presented a diagnostic rate of only 59.9%⁽⁸⁾. FNA was not employed due to the appearance of ectopic pancreas. CT and MRI are important noninvasive imaging modalities for diagnosis, especially of exogastric tumors, and treatment planning⁽⁹⁾. ESGE consider EUS as the best tool to characterize SELs, and it recommends EUS-guided fine-needle biopsy (EUS-FNB) or mucosal incision-assisted biopsy (MIAB) for tissue diagnosis of SELs \geq 20 mm and MIAB as first choice to SELs < 20 mm, as bite-on-bite, jumbo, snare or submucosal tunneling⁽¹⁰⁾.

In the series of 10 cases reported by Hu *et al.*⁽⁹⁾, average tumor size was 3.2 cm, 8/10 (80%) gastric lesions were located in the antrum, 7/10 (70%) patients reported abdominal pain or distension, and 1/10 (10%) patient was asymptomatic. All cases were strongly positive for vimentin and SMA⁽⁹⁾. Until the review published in 2021 by Arslan *et al.*⁽¹¹⁾, 130 cases of PF had been reported in the literature, with only 19 cases being described between 2007 and 2010. A possible explanation for the increased diagnosis of PF may be the increased awareness of this pathology. Yang *et al.*⁽¹²⁾ reported a subepithelial lesion located in the posterior wall of the upper gastric body, and EUS identified a heterogeneous hypoechoic mass arising from the muscularis propria layer, measuring 5.6 x 3.5 cm. Examination of the specimen showed a cystic portion containing mucinous fluid and a solid portion exhibiting a mucoid, gelatinous mass with extramural hemorrhage⁽¹²⁾. In the case reported here, the lesion was subepithelial with leakage of serous fluid after biopsy, and EUS identified a heterogeneous hypoechoic mass with cystic components arising from the second layer of the gastric wall. Patient returned to follow-up only 24 months after index endoscopy. It was performed with biopsy bite on bite with large capacity forceps. ESGE suggests surveillance of asymptomatic gastric SELs without definitive diagnosis, with EGD at 3-6 months, and then at 1-2-year intervals for lesions 10-20 mm in size. The choice of treatment and follow-up depends on the SEL subtype, the layer of origin, location and local expertise⁽¹⁰⁾.

Distal or partial gastrectomy is in cases of PF located in the antrum, with endoscopic submucosal dissection as an interesting less invasive alternative⁽⁹⁾.

An incidental 2 cm gastric SEL, shown to be a plexiform fibromyxoma in gastric biopsies, with a benign behavior over the course of 4 years including a dimensional down size to 12 mm.

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