



RESEARCH ARTICLE

PLEXIFORM FIBROMYXOMA: A RARE MESENCHYMAL GASTRIC TUMOR

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ABSTRACT

Background: Plexiform fibromyxoma, also known as a plexiform angiomyxoid myofibroblastic tumor, is a relatively new pathological entity described for the first time in 2007. This tumor is a very rare mesenchymal non-gastrointestinal stromal tumor (non-GIST) with fewer than 100 cases.

Case Summary: A 26-year-old woman presented with epigastric pain. Upper gastrointestinal endoscopy and endoscopic ultrasound revealed a gastric well-circumscribed, submucosal mass. A gastrointestinal stromal tumor (GIST) was suspected.

The patient underwent an atypical gastrectomy. The gross specimen showed a submucosal well limited glossy white tumor. Microscopic study showed a hypocellular proliferation composed of bland spindle cells set in a fibromyxoid matrix with a rich arborizing thin-walled capillary network. Spindle cells were organized in a fascicular pattern. Immunohistochemical studies revealed high reactivity for Smooth Muscle Actin (SMA), focal reactivity for Desmin with no staining for c-KIT, DOG1. The diagnosis of gastric plexiform fibromyxoma was then made.

The patient had a favorable prognosis without relapse or metastasis during the 2-year follow-up after the surgery.

Conclusion: Plexiform fibromyxoma is a very rare mesenchymal tumor with numerous similitudes to gastrointestinal stromal tumor (GIST). Immunohistochemistry is the basis of the differential diagnosis.

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INTRODUCTION

Plexiform Fibromyxoma (PF), also known as a Plexiform Angiomyxoid Myofibroblastic Tumor (PAMT), is a relatively new entity. This tumor is a very rare mesenchymal non-gastrointestinal stromal tumor (non-GIST). PF presents numerous similitudes to gastrointestinal stromal tumor (GIST). Immunohistochemistry is the basis of the differential diagnosis. Here, we report a case of PF of the stomach in a 26-year-old woman.

Case Presentation

A 26-year-old woman with a history of chronic constipation presented with epigastric pain. Upper gastrointestinal endoscopy revealed a well-circumscribed, submucosal mass located in the antrofundic junction. The mucosa covering the mass presented an erosion. Biopsy was superficial and did not interest the submucosa.

Further study with endoscopic ultrasound showed a 2.5cm necrosed tumor developed within the gastric muscularis propria. A gastrointestinal stromal tumor (GIST) was suspected.

Thoraco-abdomino-pelvic CT did not show signs of metastases. The patient underwent an atypical gastrectomy. She had no complications during the postoperative period.

The gross specimen showed a submucosal mass measuring 3.5 x 2 x 1cm. Cut section revealed a well limited glossy white tumor (Figure 1).

Microscopic study showed a well-circumscribed tumor developed within the submucosa and the muscularis propria. This proliferation was hypocellular composed of bland spindle cells set in a fibromyxoid matrix with a rich arborizing thin-walled capillary network. This matrix was Alcian blue positive. Spindle cells were organized in a fascicular pattern and had eosinophilic cytoplasm and inconspicuous nucleoli without atypia nor mitosis (Figure2). There was no lymph node metastasis.

Immunohistochemical studies revealed high reactivity for Smooth Muscle Actin (SMA), focal reactivity for CD10 and Desmin with no staining for c-KIT, DOG1, CD34, S100 protein and HMB45 (Figure 3).

The diagnosis of gastric plexiform fibromyxoma was made. The patient had a favorable prognosis without relapse or metastasis during the 2-year follow-up after the surgery.

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Figure 1 Gross specimen showing a submucosal mass and cut section revealing a well limited glossy white tumor

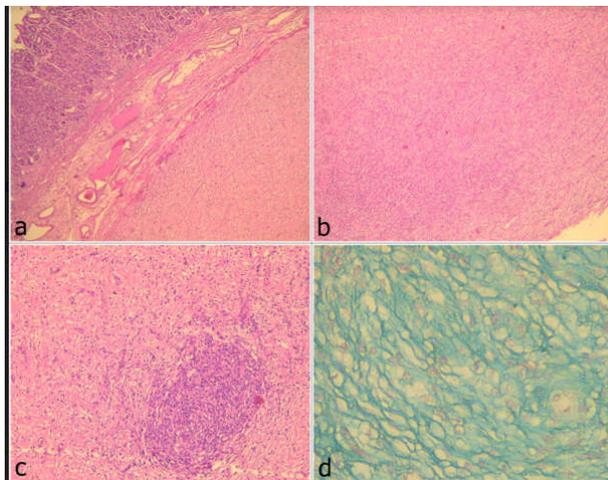


Figure 2 Microscopic study showed a well-circumscribed tumor developed within the submucosa and the muscularis propria (a). The tumor was hypocellular composed of bland spindle cells set in a fibromyxoid matrix with a rich arborizing thin-walled capillary network. Spindle cells were organized in a fascicular pattern (b-d). (a,b: Hematoxylin and Eosin, $\times 40$; c: Hematoxylin and Eosin $\times 100$; d: Alcian Blue, $\times 400$).

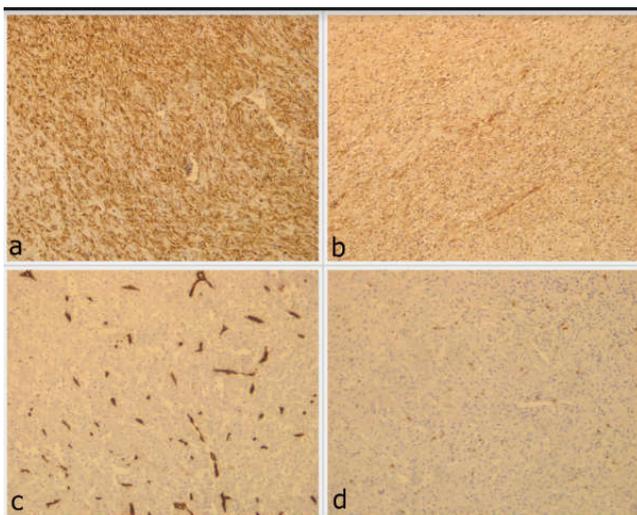


Figure 3 In immunohistochemical study, tumor cells showed diffuse cytoplasmic positivity for SMA (a), and focal positivity for CD10 (b), but were negative for CD117 (d). CD34 highlighted the rich arborizing capillary network but was negative in tumor cells (c).

DISCUSSION

Plexiform fibromyxoma (PF), also known as a plexiform angiomyxoid myofibroblastic tumor (PAMT), is a relatively new pathological entity described for the first time in 2007 by Takahashi [1]. This tumor is a very rare mesenchymal non-gastrointestinal stromal tumor (non-GIST) with fewer than 100 cases [2].

The mean patient age is 43 years, with a range from 7 to 75 years; thus far, the male-to-female ratio has been approximately 1:1.

The size ranges from 1.5 to 15 cm and the average is 5.3 cm. The tumor location seems to be exclusive to the gastric antrum, especially the pyloric region [3].

The clinical presentation is not specific. It includes anemia, gastric ulcer, nausea, abdominal pain, abdominal distension, abdominal discomfort, abdominal mass, weight loss and rarely, pyloric obstruction.

Endoscopic visualization reveals typically a glistening elastic tumor and covered with ulcerative, erosive, or smooth mucosa. Endoscopic ultrasonography shows a hypoechoic tumor with mild heterogeneity [4].

Macroscopic examination reveals a lobulated intramural or submucosal mass with a tan-white or grayish-whitish color that is usually glistening and mucoid. It may protrude from the serosa or mucosa.

As disclosed in the name PAMT, it is characterized by the presence of bland ovoid to spindle cells arranged in a plexiform or multinodular pattern and separated by abundant myxoid and a variably collagenized extracellular

matrix, with prominent arborizing thin-walled capillary network [4]. Atypia and mitosis are both rare.

Immunohistochemically, spindle cells are focal to diffuse positive for Vimentin and SMA, and sometimes for CD10, Desmin, and/or H Caldesmon or Calponin [2]. However, tumor cells of PF are negative for CD117 (C-kit), Dog1, CD34, ALK, and S100 protein. Those markers can be useful to distinguish PF from GIST and others differential diagnoses such as plexiform neurofibroma, angiomyxoma and Inflammatory myofibroblastic tumor.

PF appears to be a benign tumor. Neither recurrences nor metastatic disease have been reported after surgical resection [2,3]. This tumor is clearly distinguishable from GIST in terms of postoperative management.

A more conservative treatment than distal gastrectomy or partial gastrectomy might be considered, at least in selected cases with a smaller tumor size.

CONCLUSION

PF is a rare benign mesenchymal tumor of stomach characterized by a non-specific clinical and endoscopic presentation. Recognition of this rare entity is important to avoid misdiagnosis with other mesenchymal tumor of stomach. The main differential diagnosis is the GIST. Immunohistochemistry is the basis of the differential diagnosis.

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