

# A Rare Case of Gastric Plexiform Fibromyxoma Mimicking Gastrointestinal Stromal Tumor.

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## Abstract

We present a 54-year-old male with a gastric mass with pre-operative biopsy being consistent with a gastrointestinal stromal tumor. After surgical resection, final pathology revealed a plexiform fibromyxoma. One must consider the histopathological and gross features of plexiform fibromyxoma to distinguish it from other mesenchymal tumors of the GI tract.

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Consent was obtained from patient for publication of this article.

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## Introduction:

Plexiform fibromyxoma (PF) is a rare benign mesenchymal tumor of the gastrointestinal tract typically occurring in the gastric antrum and fundus. The pathogenesis and associated molecular alterations are currently unknown, and these tumors are difficult to distinguish from other mesenchymal tumors arising in the gastrointestinal tract. The most common clinical features of PF are abdominal pain, anemia, weight loss, abdominal distension and melena. Many cases of PF are misdiagnosed as gastrointestinal stromal tumors (GISTs) or other neoplastic masses. This is because endoscopies and other radiological studies do not identify the distinguishing features of PF, which are usually seen after resection and histopathological examination.

## Case Presentation:

A 54-year-old male with a past medical history of prostate cancer, gastroesophageal reflux disease and hypertension had undergone a screening CT scan for a rising PSA level status post radical prostatectomy six years prior and was found to have an incidental soft tissue density arising from the fundus of the stomach (Figures 1a and 1b). He subsequently underwent an endoscopic ultrasound which revealed a 28.5 x 26 mm hypoechoic mass in the gastric submucosal area arising from the muscularis propria (Figure 2). A fine needle aspiration was performed with pathology showing a bland spindle cell neoplasm consistent with a gastrointestinal stromal tumor. He was then referred to surgery and underwent an uncomplicated laparoscopic, hand-assisted wedge resection of the mass. The patient had an unremarkable post-operative course and was discharged on post-operative day two. Final pathology revealed a plexiform fibromyxoma measuring 4 x 3.2 x 2 cm with negative margins. Histology revealed bland appearing spindle cell proliferation with some myxoid degeneration with patchy positive staining for CD117 (Figures 3-4). Furthermore, there was nonspecific staining of the spindle cells with pancytokeratin and cytokeratin CAM 5.2 (Figures 5-6). His case was discussed at a multidisciplinary tumor board with the consensus of no further treatments necessary. The patient has been seen in follow-up and continues to do well. b

## Discussion:

Plexiform fibromyxoma is an extremely rare mesenchymal tumor, and only 130 cases have been reported in literature as of May 2021. The median age of presentation is between 40 and 50 years, but there have also been reports of pediatric cases. PF is difficult to distinguish from other submucosal tumors such as GISTs using typical radiological studies. The patient discussed in this case report had a prior history of GERD, but the discovery of the mass was incidental. This tumor is often an incidental finding; however, may also present with clinical features such as weight loss, gastrointestinal bleeding, dyspepsia, and abdominal pain. PF has a gross appearance described as a pale tan to rose colored mass that is hemorrhagic, and either mural or partially extramural. The surface of these neoplasms is often described as mucoid and gelatinous in nature. The size of the tumor typically ranges anywhere from 0.8 cm to 16 cm. The median tumor size of PF is 14 cm.

Histologically, PF is characterized by a distinctive plexiform architecture, with an extensive capillary network throughout the tumor, contributing to the hemorrhage sometimes seen upon presentation. Histologically, the characteristic features of a PF is plexiform and multinodular involvement in the muscularis propria of the gastric antrum. The cells are characterized as bland and scattered ovoid to spindle shaped cells present in a myxoid or fibromyxoid and collagenous matrix. Upon immunohistochemistry analysis, the most common positive markers for PF include SMA, MSA, and vimentin<sup>2</sup>. PF typically stain negative for CD117 (c-kit), an important distinguishing feature between it and GISTs. However, there have also been cases where these tumors have stained positive for CD117.

The treatment of choice for PF is surgical resection. In this case, the patient was treated with a laparoscopic, hand-assisted wedge resection. As PF is a benign tumor, there have not been reports of metastases or malignant transformations. The most common features that indicate aggressive behavior such as vascular and lymphatic invasion are usually absent in PF, and this tumor has not been known to metastasize or recur.

### Conclusion:

In conclusion, we find that the clinical, histological, and gross tumor appearance are important in distinguishing PF from GISTs and other mesenchymal tumors of the GI tract. Data further describing the epidemiology, risk factors, and cell origin should be conducted to develop a much more detailed understanding of this tumor

### Author Contributions:

1. Ramya Mohankumar: contributed to the design of the manuscript, drafting and revisions, approval of version to be published and agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.
2. Kyle Schank, MD: contributed to the design of the manuscript, drafting and revisions, approval of version to be published and agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.
3. Sajjaad Samat, MD: contributed to the design of the manuscript, drafting and revisions, approval of version to be published and agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.
4. Srinivas Kavuturu, MD: contributed to the design of the manuscript, drafting and revisions, approval of version to be published and agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

### References:

#### Figures:

Figure 1a and b: CT scan demonstrating a mass-like density in the gastric fundus.

Figure 2: Endoscopic ultrasound demonstrating a 28.5 x 26 mm gastric mass arising from the muscularis propria.

Figure 3: CD117 stained cells.

Figure 4: Spindle Cells.

Figure 5: CAM 5.2 stained cells.

Figure 6: Pancytokeratin stained cells

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