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Gastric Plexiform Fibromyxoma with Ulceration and Bleeding: An Unusual Presentation of a Rare Tumour

Hiren Vasava[1*], Keyur Bhatt[2], Dhaval Mangukiya[2] and Jigar Jariwala[2]

[1]1st Year FNB- Minimal Access Surgery, SIDS Hospital, Surat, India
[2]Consultants, Surgical Gastroenterology, SIDS Hospital, Surat, India

[*]**Corresponding author:** Hiren Vasava, 1st Year FNB- Minimal Access Surgery, SIDS Hospital, Surat, India, Tel: 099099 07475; E-mail: hiren.vasava9619@gmail.com

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Abstract

Background: Plexiform fibromyxoma (PFM), also known as plexiform Angio myxoid myofibroblast tumour, is a rare benign mesenchymal neoplasm of the stomach. It typically arises from the gastric antrum and can mimic GIST both clinically and radiologically.

Case Presentation: A 38Year/female with complaints of black coloured stool since last 20 days UGI Scopy - Stomach showed ulcero - proliferative lesion? Extrinsic infiltration at antral region. OGD Biopsy report- Chronic active gastritis with severe ulceration. No granuloma /dysplasia, Malignancy. CECT- demonstrated a lobulated submucosal homogenously enhancing soft tissue lesion in the Antro pyloric region, causing moderate compromise of the lumen. Findings are in favour of neoplastic Lesion. The patient underwent Distal Gastrectomy with Retro Colic Gastrojejunostomy.

HPE - Neoplasm with spindle to stellate cells - Suggestive of mesenchymal Origin, involving wall of stomach IHC - Positive for smooth muscle actin (SMA) and CD10, and negative for ALK, CD34, SOX 10, S100, CK AE1/AE3, Beta catenin, DOG 1. Features are consistent with Plexiform Fibromyxoma. The postoperative course was uneventful.

Conclusion: Plexiform fibromyxoma is a distinct benign gastric tumour that closely mimics GIST. Awareness of its characteristic histopathological and immunohistochemical features is essential for accurate diagnosis and to prevent unnecessary aggressive treatment.

Keywords: ***Plexiform fibromyxoma; Benign mesenchymal tumour; GIST mimic***

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1. Introduction

Plexiform fibromyxoma (PFM), also known as plexiform Angio myxoid myofibroblastic tumour (PAMT), is a rare benign mesenchymal neoplasm of the stomach. It was first described in 2007 and predominantly arises from the gastric antrum [1]. PFM is characterized by a plexiform growth pattern of spindle and stellate cells within a myxoid stroma [2]. Clinically and radiologically, it often mimics gastrointestinal stromal tumour (GIST), but unlike GIST, it lacks malignant potential [3]. Accurate diagnosis relies on histopathological features and immunohistochemical profiling [2-4].

2. Case Presentation

A 38-year female patient presented to the Outpatient department with black stool for 10 days associated with generalised weakness & fatigue. Past history of blood transfusion 1 year back (Haemoglobin - 4 gm /dl).

Ultrasound (Abdomen) - Normal.

Esophagogastroduodenoscopy Report - Stomach shows Ulcer proliferative lesion. Extrinsic Infiltration at antral region (FIG. 1).

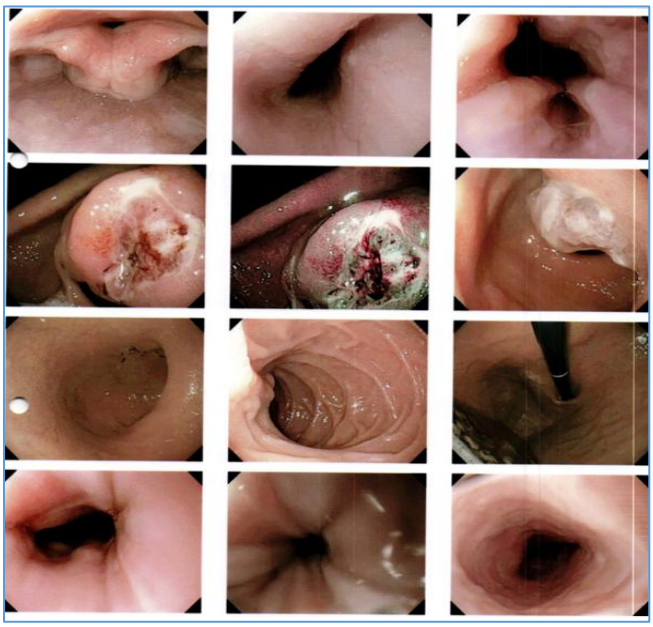

FIG. 1. **OGD findings.**

Stomach shows ulcer proliferative lesion with extrinsic infiltration at antral region.

Biopsy Report - Chronic active gastritis with severe ulceration. No granuloma /dysplasia, malignancy.

Contrast enhanced CT scan - Abdomen

A lobulated submucosal homogenously enhancing soft tissue lesion in the Antro pyloric region, causing moderate compromise of lumen- without any proximal gastric overdistension. Findings are suggestive of neoplastic lesion - (? Gastro Intestinal Stromal Tumour) FIG. 2.

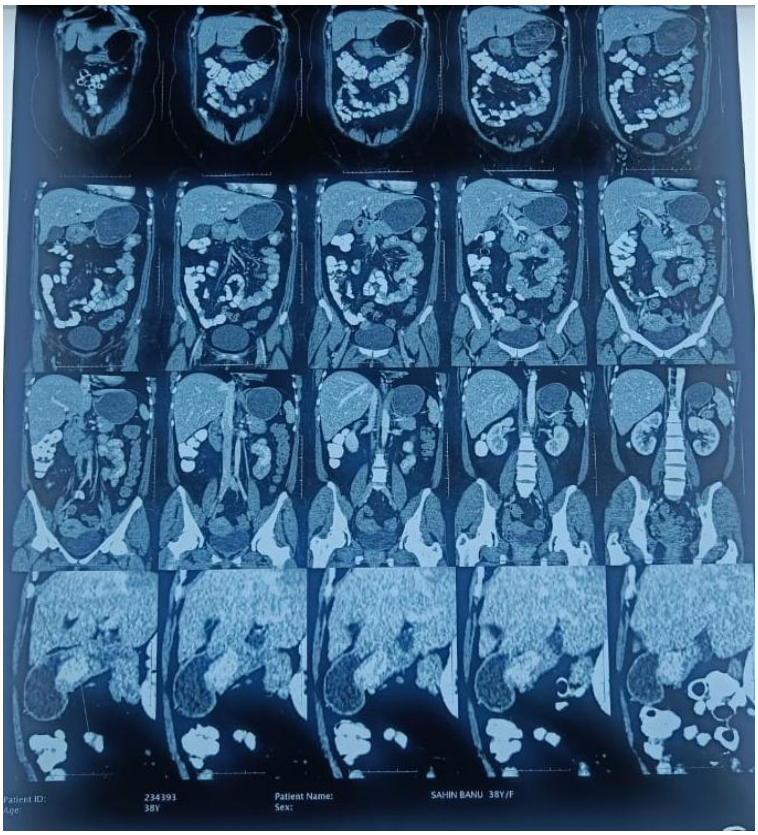

FIG. 2. **CECT (Abdomen & Pelvis).**

Planned for routine Laparoscopic Radical Distal Gastrectomy. Post operative course was uneventful.

HPE - Neoplasm with spindle to stellate cells – Suggestive of mesenchymal Origin, involving wall of stomach IHC - Positive for smooth muscle actin SMA and CD10, and negative for ALK, CD34, SOX 10, S100, CK AE1/AE3, Beta catenin, DOG 1. Features are consistent with Plexiform Fibromyxoma. The postoperative course was uneventful.

3. Discussion

Plexiform fibromyxoma (PFM), also known as plexiform angiomyxoid myofibroblastic tumour (PAMT), is a rare benign mesenchymal neoplasm of the stomach, first described by Takahashi et al. in 2007 [1]. Since its initial recognition, fewer than 200 cases have been reported in the literature [5]. It typically arises from the gastric antrum and pyloric region, although occasional cases have been documented in the duodenum and small intestine.

PFM usually affects adults with a slight female preponderance, but it can occur over a wide age range [6]. The clinical presentation is nonspecific and depends on tumour size and location. Most patients present with upper abdominal pain,

gastrointestinal bleeding, anaemia, or are incidentally diagnosed during endoscopy or imaging for unrelated reasons. Endoscopically, the lesion appears as a submucosal mass, sometimes with mucosal ulceration.

Radiologically, PFM often appears as a well-circumscribed, intramural, or submucosal mass with heterogeneous enhancement due to its myxoid and vascular stroma. However, imaging alone cannot reliably differentiate it from gastrointestinal stromal tumour (GIST), the most common gastric mesenchymal tumour. Therefore, histopathological and immunohistochemical examination is essential for diagnosis.

Histologically, PFM is characterized by a multinodular or plexiform growth pattern composed of bland spindle cells in an abundant myxoid stroma with prominent arborizing vasculature. Mitotic activity is low, and necrosis is absent. Immunohistochemically, the tumour cells are typically positive for smooth muscle actin (SMA) and vimentin, variably positive for desmin, and negative for CD117 (c-KIT), DOG1, CD34, and S-100 protein. This profile distinguishes PFM from GIST and other myxoid mesenchymal neoplasms.

The pathogenesis of PFM remains unclear, but molecular studies have shown no KIT or PDGFRA mutations (5), further supporting its distinction from GIST. Rare cases have shown GLI1 gene rearrangements, suggesting a possible molecular pathway of tumorigenesis.

Surgical resection remains the mainstay of treatment and is usually curative. Depending on the tumour size and location, options range from local wedge resection to distal gastrectomy. Recurrence or metastasis has not been documented in long-term follow-up, emphasizing its benign clinical course. Hence, accurate recognition is crucial to avoid overtreatment with tyrosine kinase inhibitors, which are ineffective for this entity.

4. Consent

Written informed consent was obtained from the patient.

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