

Gastric plexiform fibromyxoma, an uncommon mesenchymal tumor

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Received: 08/04/2020 · **Accepted:** 02/06/2020

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ABSTRACT

Plexiform fibromyxoma (PF) is an uncommon primary tumor of the gastrointestinal tract, with a mesenchymal origin and a benign behavior. Herein, we report a case and provide a literature review. A 41-year-old male patient underwent surgery in our unit for a PF at the gastric antrum, after being admitted due to vomiting and weight loss. As illustrated by our case, the mean age at presentation is around 40 years, the antrum is the most common location and abdominal pain the most widely reported manifestation. None of the reviewed cases involved regional or distant spread.

Keywords: Plexiform angiomyxoid myofibroblastic tumor. Plexiform fibromyxoma. Plexiform angiomyxoma. Plexiform angiomyxoid tumor.

INTRODUCTION

Plexiform fibromyxoma (PF) is a mesenchymal tumor of the gastrointestinal tract with highly peculiar histopathological characteristics, hence its denomination (1,2). It is an uncommon tumor, with only 19 confirmed cases reported in the literature since 2016. The incidence of gastrointestinal stromal tumor (GIST), with which a differential diagnosis should be considered, has been estimated to be 150 times that of PF (3-5). Typically, it has a benign behavior with a very high long-term survival rate. We report the case of a middle-aged male patient with gastric PF who underwent surgery in our unit and provide a review of the literature.

Conflict of interests: the authors declare no conflict of interest.

This case report was presented at the XXXII National Surgical Conference on November 12-15, 2018 in Madrid, Spain, and was included in the "Best of Conference" category.

METHODS

A brief case report and literature review search was performed in PubMed using the terms "plexiform angiomyxoid myofibroblastic tumor, plexiform fibromyxoma, plexiform angiomyxoma or plexiform angiomyxoid tumor" from May 2007 to February 2018. Case reports in English and Spanish were selected and 15 publications were retrieved. Demographic data, manifestations, location, size, type of procedure, immunohistochemistry, follow-up and mortality were analyzed in these cases.

RESULTS

Case report

We report the case of a 41-year-old male patient who was admitted due to recurrent vomiting and a 40 kg loss of body weight over two years. Computed tomography (CT) scans showed a 5 cm-long thickening of the gastric wall at the lesser curvature, with poorly defined borders and the development of perigastric collateral vascularization (Fig. 1). The study was completed with gastroscopy, which revealed a benign-looking antropyloric stricture, with negative histology for malignancy. Hence, the management initially consisted of endoscopic dilation. Surgery was decided after repeated failed attempts at endoscopic management and a

Magadán Álvarez C, Olmos Martínez JM, Toledo Martínez E, Trugeda Carrera MS, Fernández Díaz MJ, Martín Rivas B, Mazorra Horts R, Mayorga Fernández MM, Arias Pacheco RD. Gastric plexiform fibromyxoma, an uncommon mesenchymal tumor. *Rev Esp Enferm Dig* 2021;113(3):183-185

DOI: 10.17235/reed.2020.7048/2020

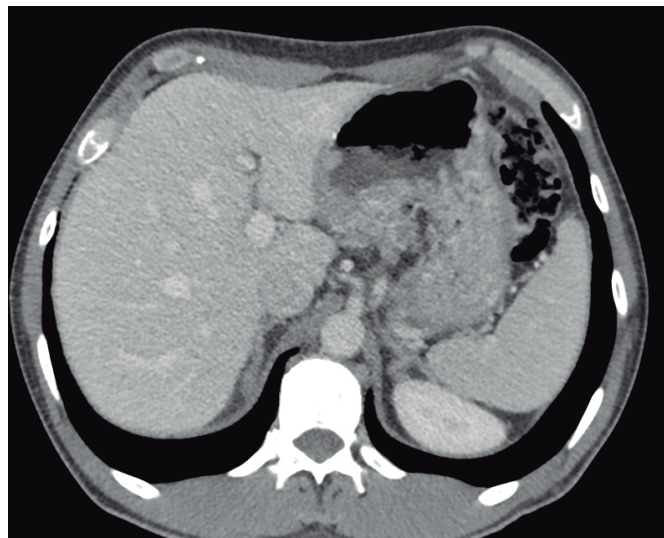


Fig. 1. The image shows a 5 cm thickening of the gastric wall in the lesser curvature, with poorly defined borders and the development of perigastric collateral vascularization.

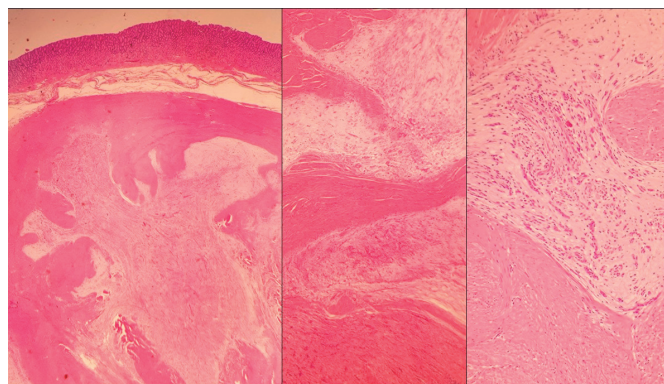


Fig. 2. A neoformation of a nodular growth with a plexiform pattern was observed under the mucosa, which is inserted between the fibers of the muscularis propria. Tumor cells are spindle-shaped and located in a myxoid matrix.

laparoscopic distal gastrectomy was performed. Plexiform fibromyxoma was diagnosed histologically (Fig. 2) and immunohistochemistry (IHC) was positive for muscle-specific actin (MSA), vimentin, smooth muscle actin (SMA), desmin, caldesmon, calponin and CD10. C-Kit, S100 protein, CD34, NSE, DOG1, NF, progesterone and β -catenin staining were negative.

Review findings

PF cases were collected and analyzed from 13 studies. The mean age was 42 years, ranging from seven to 83 years (1-10) and there were no differences in sex distribution (1,3-13). A total of eight studies were rejected due to a lack of access to the full text, failure to report detailed data, or being written in a language other than Spanish or English.

Manifestations varied from asymptomatic cases to nonspecific gastrointestinal (GI) complaints, which rendered a diagnosis incidental on many occasions (1-3,8,9). Predominant symptoms included abdominal pain followed by upper GI bleeding (1,3-5,7,8,10,11,13). Obstructive complaints (7,13,14), weight loss and a palpable mass (1,9,13) with fistulization were less frequently reported (1).

The most commonly involved site was the gastric antrum (80 %) (3,5-8,10,12) particularly the pyloric region (1,2,9). Extension to perigastric soft tissues or duodenal bulb was described (10) in 20 % (2,8) to 50 % of patients (15) and cases were described in the esophagus, duodenum, jejunum, gallbladder, mediastinum and colon (1,3,5,10).

The mean tumor size was 5.5 cm, with a range from 1.5 to 15 cm (3,6-11,13). Morphologically, these tumors were described as multinodular, non-encapsulated lesions projecting onto the serosa layer (1,8,10,11) and up to 75 % had mucosal ulceration (2,7).

No cases of malignant transformation (1), local recurrence or distant spread (3-10,12) have been reported and several authors found no evidence of tumor-related mortality (8). Some authors report vascular invasion and suggest the potential for intravascular tumor spread within the gastric wall and subserosal layer, which does not worsen prognosis (1,10,14).

DISCUSSION

PF is a primary, mesenchymal-originating tumor of the GI tract that was first reported in 2007 by Takahashi (1-8). It is an uncommon tumor, with an estimated incidence of one PF case per 150 GISTs (3-5). The World Health Organization adopted the plexiform fibromyxoma designation in 2010, replacing other terms such as plexiform angiomyxoma and plexiform angiomyxoid myofibroblastic tumor (2).

The preoperative diagnosis of PF may be challenging, as the condition may mimic a GIST, with similar radiographic and endoscopic characteristics (12). Given the submucosal nature of these lesions and the important role of histology for their diagnosis, endoscopic ultrasound associated with puncture-aspiration is considered as the diagnostic test of choice (1). Major radiographic characteristics include heterogeneous attenuation with enhancement in the arterial phase and progressive enhancement during the venous and delayed phases, which have been associated with its myxoid component (8). Growths with a cystic component (8,10) and strongly enhanced small nodules in their periphery have also been reported (4). Magnetic resonance imaging is superior to CT for the visualization of tumor extent and its components (9).

Macroscopic, microscopic, IHC and molecular findings are crucial for the exclusion of the different types of GIST, which are more common and potentially have a more aggressive behavior (2,5,8-10,13). In contrast with PF, c-kit expression is considered to be key for their diagnosis and the lesions are typically positive for CD117 or DOG1 (1,4-6,8,10,14).

A plexiform growth pattern involving fusiform cells in a myxoid or fibromyxoid stroma with positive Alcian blue

stain is typical of PF (8). From an immunohistochemical viewpoint, the presence of SMA is characteristic (1,3-5,7,8,10-12,14). The expression of MSA, CD10, vimentin, desmin, caldesmon and calponin have also been registered, which would point at a myofibroblastic origin (1,4,5,7,8,10).

Primary resection is recommended for the accurate treatment and histological diagnosis (1,2,13). Currently, partial gastrectomy remains the treatment of choice (13). Atypical resection, a combined approach and advanced endoscopic resection using endoscopic submucosal dissection (ESD) have all been reported (13,14). In the past few years, their use has been extended not only to gastric submucosal tumors but also to early gastric carcinoma. These techniques obtain cure rates higher than 90 % and morbidity and mortality rates lower than those of surgery (15).

PF behavior is typically benign, with no mortality or distant spread reported in the literature. However, further and more extensive studies are needed to improve perioperative diagnosis and standardize treatment and follow-up.

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