CASE REPORT

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Vanek's tumor: a rare cause of gastric outlet obstruction. Case report and literature review

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Abstract

Vanek's tumor or inflammatory fibroid polyp (IFP) is a rare submucosal, mesenchymal tumor of the digestive tract, of unknown etiology. We present an unusual case of a 62-year-old female patient, investigated for intermittent nausea, vomiting, abdominal pain and anemia. Narrow-band imaging (NBI) endoscopy showed a polypoid tumor with normal coverage mucosa prolapsing through the pylorus. The tumor was surgically removed. Histopathological (HP) examination revealed the presence of spindle cells with uniform nuclei exhibiting no mitotic activity. The inflammatory cells were predominantly represented by eosinophils. The vascular component was prominent suggesting a vascular lesion. We put emphasis on the presentation of this particular case because of its scarcity among the other types of gastric polyps, suspicion for features of malignancy, and the particularities of the clinical presentation, consisting of intermittent gastric outlet obstruction ("ball valve syndrome") accompanied by weight loss and anemia. The differential diagnosis of other benign or malignant gastric lesions was based on endoscopic, computed tomography and HP aspects.

Keywords: ball valve syndrome, inflammatory fibroid polyp, benign, gastric.

Introduction

Vanek's tumor or the inflammatory fibroid polyp (IFP) was first described in 1949 by Vanek [1]. The name of inflammatory fibroid polyp was proposed by Helwig & Ranier, in 1953 [2]. This type of polyp occurs mainly in adults, typically between the fifth and seventh decades of life [2].

IFP can be most frequently localized in the stomach, at the level of the digestive tract (66–75%) [3]. It is a rare benign gastric tumor, representing approximately 4.5% of stomach polyps [3]. This type of tumor consists of mesenchymal cells that originate from the submucosa and promote proliferation. Usually, the lesion is less than 1 cm in size. Larger lesions (>4 cm in diameter) causing secondary complications (bleeding, obstruction) have also been reported [4].

The differential diagnosis with other submucosal gastrointestinal tract tumors, especially gastrointestinal stromal tumor (GIST), should be considered. The role of biopsy samples obtained during endoscopy is limited in the majority of the cases, which it makes difficult to establish the final diagnosis. A final conclusion can be drawn only after the removal of the tumor.

The endoscopic diagnosis of the lesion was performed with narrow-band imaging (NBI) technology, by using an Olympus Exera III endoscope. NBI endoscopy is an optical image-enhancing technology that allows a detailed investigation of the vascular and mucosal patterns, thus predicting histology in real-time endoscopy [5, 6].

Our report aims to present a rare benign gastric lesion with a particular clinical presentation (occlusive syndrome, accompanied by weight loss and anemia), and to emphasize specific diagnostic criteria that help us to distinguish IFP from other benign or malignant polyps.

Case presentation

B.A., a 62-year-old female patient was admitted to the Department of Gastroenterology, Mureş County Hospital, Romania, during 20–27 December 2016, complaining of nausea, vomiting, and abdominal pain. On further questioning, the patient admitted that she had been suffering from intermittent bloating, nausea, vomiting and weight loss for two months. She denied any history of visible gastrointestinal bleeding. The patient did not have significant pathologies in her family and personal medical history. She denied smoking and drinking alcohol and did not work in a toxic environment.

The objective clinical examination revealed the following: body temperature 36.4ºC, blood pressure 105/95 mmHg, resting heart rate 100 beats/min, respiratory rate 21 breaths/min, pale skin and mucosa. Pain was felt in the epigastrium on profound palpation, and there were no palpable abdominal masses detected. The laboratory tests confirmed the microcytic anemia: hemoglobin was 7.33 g/dL (reference range 12–15 g/dL), and hematocrit was 29% (reference range 39–49%). The peripheral blood smear showed anisocytosis, anisochromia.

Considering the upper gastrointestinal tract symptoms,

gastroscopy was indicated. Endoscopy was performed by using NBI technique with magnifying endoscopy (ME). A polypoid tumor with a large base and normal coverage mucosa was detected at the antrum level (Figures 1 and 2). Duodenal examination showed fresh blood in a small quantity and ulcerations of the overlying mucosa. The biopsies were taken from the lesion and the surrounding mucosa, but the histopathological (HP) results based on Giemsa staining and specific immunohistochemical (IHC) examination showed only alterations caused by reactive gastropathy, without significant inflammation in the mucosa and the absence of *Helicobacter pylori (H. pylori)*. Periodic Acid–Schiff (PAS)–Alcian Blue staining showed no evidence of intestinal metaplasia. The biopsy was also negative for dysplasia.

The computed tomography (CT) scan with an intravenous and oral contrast substance revealed a non-iodophilic, well-defined, hypodense formation located at the level of the gastric antrum and duodenal bulb, protruding inside the lumen, without any sign of invasion, suggesting a benign lesion (Figure 3).

The patient underwent surgery, and antrum resection was performed with gastrointestinal anastomosis. The surgical specimen received for HP examination was represented by a 50×60 mm gastric antrum, with a protruding 40×40×25 mm polyp, with abroad base and smooth surface, covered by intact mucosa, accompanied by erosions in the duodenal slope (Figure 4). On the cut surface, the polyp was solid, with well-delineated margins, white-gray, with prominent edematous features.

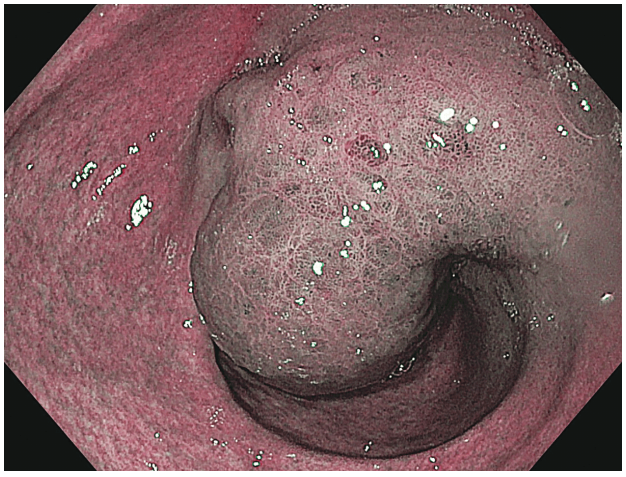

Figure 1– ***Narrow-band imaging endoscopic appearance of the formation covering the pylorus.***

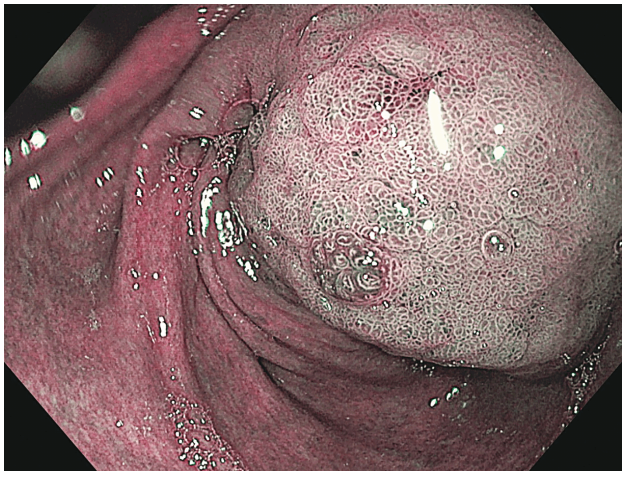

Figure 2 – ***A coil-shaped appearance of subepithelial capillary network in normal antral mucosa (narrow-band imaging endoscopy, near focus mode).***

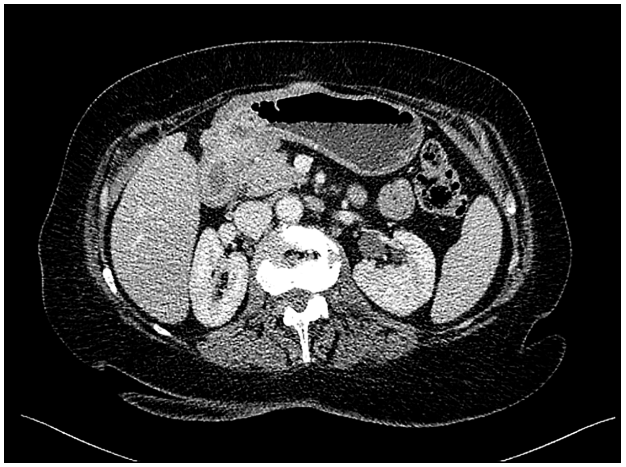

Figure 3 – ***Axial computed tomography scan detected non-iodophilic, well-defined, hypodensial formation, located at the level of the gastric antrum and duodenal bulb.***

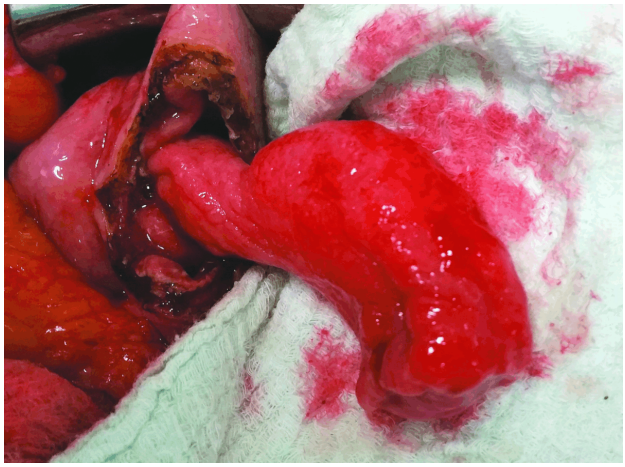

Figure 4 – ***Resected specimen: a 40×40×25 mm polyp, with broad base and smooth surface, covered by intact mucosa, accompanied by erosions in the duodenal slope.***

The microscopic examination of the surgical specimen revealed a polypoid lesion covered by a mucosa of normal appearance. The tumor was localized in the submucosa and was clearly delimited from the overlying mucosa through the muscle of the mucosa (Figure 5). The vascular component was prominent with structures of variable size, predominantly small and medium size, some of which were dilated and of variable thickness of the walls. The prominent vascular component suggested a possible vascular lesion. The fusiform cells identified in the stroma presented small, uniform, ovoid nuclei, without nucleoli and mitoses. The fusiform cells in the stroma were concentrically positioned, like onion leaves (Figure 6). Typically, the polyp consisted of myxoid stroma, containing spindle cells, inflammatory cells, and blood vessels (Figure 7). The inflammatory cells were predominantly

represented by eosinophils, but lymphocytes, plasma cells, and mastocytes were also present (Figure 8).

Modifications caused by reactive gastropathy were present in the mucosa above the lesion and in the rest of the antrum. Edema in the lamina propria and marked foveolar hyperplasia with minimal inflammation were also detected. Giemsa staining and IHC examination, similar to the biopsy, did not identify the presence of *H. pylori*. Neither intestinal metaplasia nor dysplasia was found. The surgical margins were tumor free.

Although the routine HP examination was characteristic of an inflammatory polyp, further IHC staining was performed to confirm the diagnosis. Spindle cell lipoma was strongly positive for CD34, with diffuse cytoplasmic expression, while factor VIII highlighted the vascular component and the positivity of the endothelial cells. Smooth muscle actin (SMA) was focally positive in the fusiform cells, but a prominent expression was identified in the vessel walls. Cytokeratin AE1/AE3, desmin, S100 protein, CD117, and DOG1 expression was not detected in the polyp (Figures 9 and 10).

The patient was discharged one week after admission, without subjective complaints and the recommendation of endoscopic monitoring.

Discussions

Inflammatory fibroid polyps are generally diagnosed in adults [3]. This is a rare disease, with only 18 pediatric cases described in the literature [7]. Gastric fibrovascular polyps are most frequently located in the antrum (70%). They can appear in other segments of the digestive tract like the small intestine or colon [1, 8].

As far as the etiology of IFP is concerned, an abnormal inflammatory response is suspected to be the trigger of this condition, but its mechanical, chemical (such as bile reflux) or biological trigger remains unknown. Some published reports describe *H. pylori* as a possible etiology [8–10]. A case of an IFP, which presented morphological changes after the eradication of *H. pylori*, has been reported [11]. In our case, *H. pylori* was not present in the antral and corporeal biopsy specimens. The etiological role and mechanism of *H. pylori* in IFP remain speculative [10].

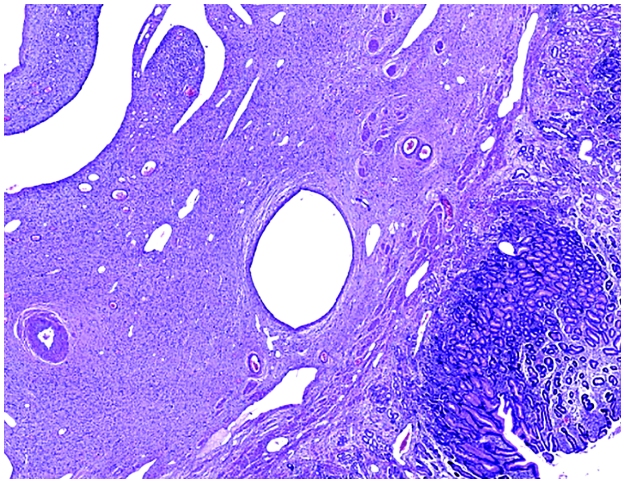

Figure 5 – ***Well-defined lesion in the submucosa, clearly delimited from the overlying mucosa through the muscle of the mucosa [Hematoxylin–Eosin (HE) staining, ×20].***

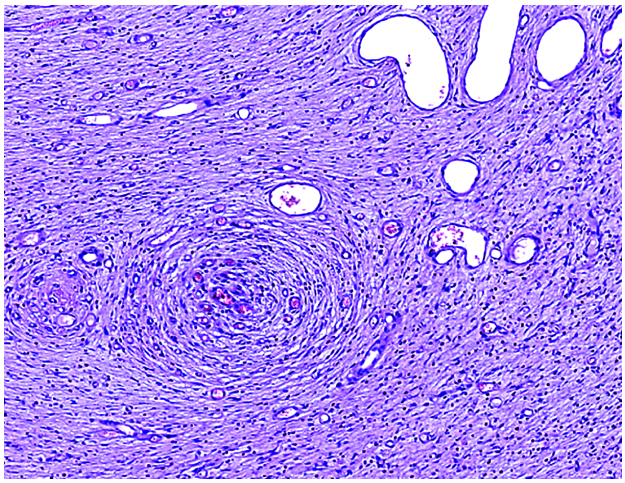

Figure 6 – ***The prominent vascular component suggesting a possible vascular lesion. The fusiform cells identified in the stroma presents small, uniform, ovoid nuclei, without nucleoli and mitoses. Focally, the fusiform cells in the stroma are concentrically positioned, like onion leaves (HE staining, ×40).***

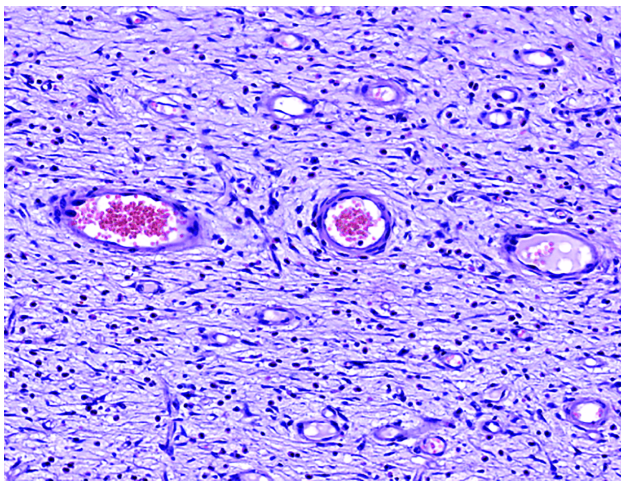

Figure 7 – ***Myxoid stroma, with spindle cells, inflammatory cells, and blood vessels (HE staining, ×100).***

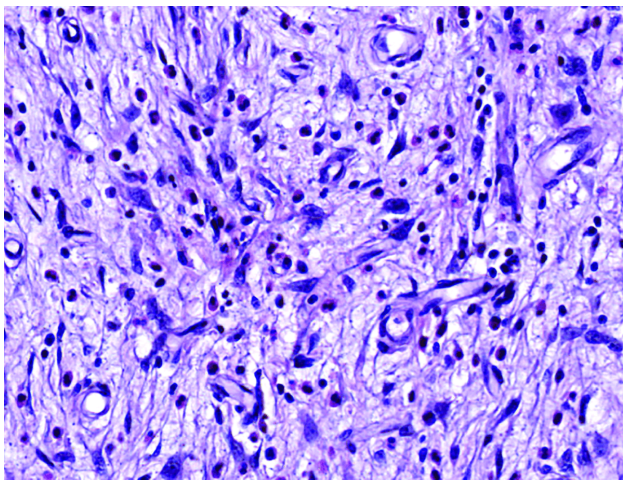

Figure 8 – ***The inflammatory cells were predominantly represented by eosinophils; lymphocytes, plasma cells and mastocytes are also present (HE staining, ×200).***

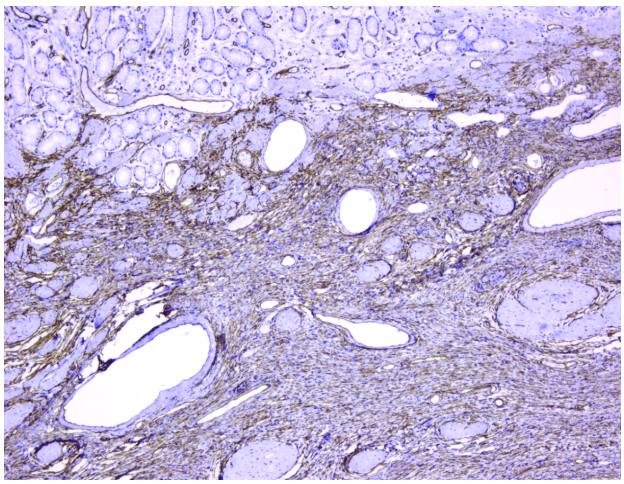

Figure 9 – ***CD34 immunohistochemical staining positive in the spindle cells, with diffuse cytoplasmatic expression (×20).***

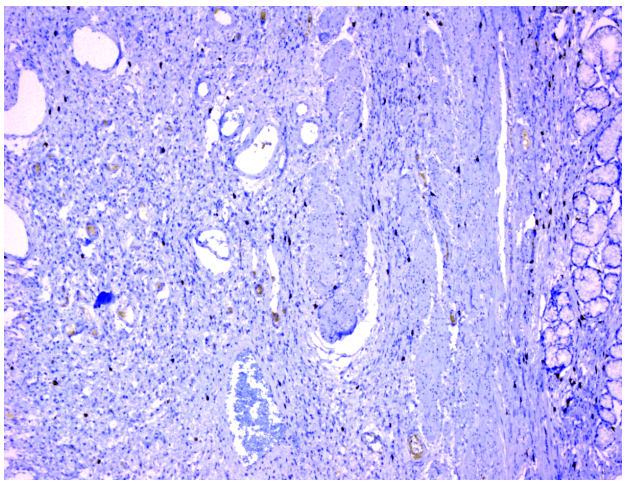

Figure 10 – ***Tumor cells with negative reaction to the anti-CD117 antibody. Imunohistochemical staining (×20).***

Most fibro-inflammatory polyps are discovered accidentally, during a superior digestive endoscopy [12]. Patients are predominantly asymptomatic or present uncharacteristic symptomatology: dyspepsia, abdominal discomfort [9, 13]. In our case, the patient's symptomatology was significant, similar to those of malignant tumors: nausea, vomiting, abdominal pain. The gastric outlet obstruction was intermittent due to the prolapse of the pedunculated polyp through the pylorus into the duodenum. This rare presentation was named by Hobbs & Cohen "ball valve syndrome" [14]. Gastroduodenal intussusception caused by a prolapsed gastric polyp is rarely documented [15]. Intussusception and obstruction are the most frequent initial symptoms when the polyp is located in the small intestine [16]. The symptomatology is associated with the size of the polyp [17]. These formations rarely exceed 4 cm. However, a retroperitoneal IFP measuring approximately 20 cm was reported [18].

There is a single report of a correct pre-operative diagnosis of gastric IFP using a combination of conventional endoscopy and endoscopic ultrasonography (EUS) [19]. Because of the large size and the characteristic of the tumor, the diagnosis cannot be established only endoscopically (without histological assessment), since it is impossible to determine whether the tumor has a mucosal or submucosal location. We examined our patient with NBI and ME and did not detect any modified mucosal or vascular patterns characteristic to epithelial polyps, so the diagnosis was submucosal tumor with normal covering mucosa.

Mori *et al.* (1988) found that 8% of 50 patients present both gastric IFP and adenoma [20], while Mucientes *et al.* (2012) described gastric IFP associated with gastric adenocarcinoma [21]. In our case, there was no dysplasia in the mucosa overlying the polyp or in the rest of the gastric antrum. Multiple small bowel IFPs were documented in two families, affecting only the women, and this condition was named "Devon polyposis syndrome" [22].

Regarding the treatment of IFP, some authors used submucosal dissection for the treatment of small submucosal tumors, in case of accessible lesions [9, 19]. Local excision with free surgical margins represents the curative remedy. The follow-up of patients with IFP revealed the absence of recurrences [4]. Therefore, after the complete resection of the tumor, additional therapy is not necessary. If the IFP is sessile, or with a large base, like in our case, endoscopic resection may result in perforation or incomplete resection of the polyp, and increased risk of local recurrence. Zinkiewicz *et al.* reported a case of recurrent IFP at the level of cardio-esophageal junction following endoscopic submucosal excision [23]. In our patient with wide base polyp, large tumor size, and pylorus involvement the recommended treatment was surgical resection. Partial antrectomy with gastroenteric anastomosis was carried out.

The final diagnosis was established only after the HP assessment of the resected specimen. Typically, the tumor arises from the submucosa and is characterized by a stroma with variable myxoid appearance, but with numerous inflammatory cells (mainly eosinophils) and fibroblast proliferation with a perivascular distribution [12]. Sometimes, small lesions are restricted to the mucosa, while large lesion may extend in the muscularis propria.

The differential diagnosis of other lesions can be challenging in practice. Eosinophilic gastroenteritis, inflammatory myofibroblastic tumor, plexiform fibromyxoma, solitary fibrous tumor, GIST or mesenteric fibromatosis invading the gastric wall should be considered. Because of the prominent vascular component, hemangioma and arteriovenous malformations should be excluded. Other benign mesenchymal tumors such as leiomyomas, neurofibromas, and schwannoma may be taken into consideration [3, 17].

Because there were multiple eosinophils in the lesion, some authors considered the lesion to be a variant of the localized eosinophilic gastroenteritis and name it eosinophilic granuloma. In case of eosinophilic gastroenteritis, patients present allergies to a significant degree in their medical history, and transmural inflammatory infiltrate is identified histopathologically [9, 17]. In case of our patient, blood eosinophilia was not revealed, and she had no known history of allergies. We found that the number of eosinophils was normal in the surrounding antrum, including the mucosa.

Inflammatory myofibroblastic tumor (IMT) rarely appears in the stomach. IMT is frequently associated with systemic signs, and the inflammatory component is represented by plasma cells, lacking regular vascular pattern. The IHC expression is different, IMT is positive for desmin, keratin, and anaplastic lymphoma kinase (ALK) and is negative for CD34 [24]. Plexiform fibromyxoma is a multinodular tumor centered in the muscularis propria, with a myxoid appearance, but without concentric distribution of CD34 fusiform cells around the vessels [25]. Solitary fibrous tumors are arising from the serosa in the gastrointestinal tract and are characterized by hemangiopericytoma (HPC)-like vessels with ropy collagen in the stroma [26].

The clinical differentiation between IFP and GIST can be difficult since the endoscopic appearance may be similar. Edematous or myxoid stroma with prominent vascular form and numerous eosinophils, associated with IHC examination lead to a correct diagnosis. Both tumors are positive for CD34, but CD117 and DOG1 are negative incase of IFP and positive in case of GIST [27, 28].

Mesenteric fibromatosis can invade the gastric wall, but the lesions are extensive, centered in the serosa, with bland spindle or stellate shape cells evenly deposited in a collagenous or sometimes myxoid stroma with thin-walled vessels. They express β-catenin and are negative for CD34 [29]. Because of the abundance of vascular structures, it is difficult to differentiate IFP from the vascular lesions, such as hemangioma or arteriovenous malformation, but the presence of the fusiform cells positive for CD34 confirms the diagnosis [30]. Other benign mesenchymal tumors like leiomyomas, neurofibromas, and schwannomas can be easily excluded, because, apart from the spindle cell component, they present a different architecture and a specific IHC profile. They are also negative for CD34 [3].

Conclusions

We presented a rare case of gastric IFP with salient, atypical symptomatology, similar to that of malignant tumors. This symptomatology was caused by intermittent pyloric obstruction. To the best of our knowledge, this case of IFP causing "ball valve syndrome" is a very rare case reported in the literature. The final diagnosis was based on the HP assessment, but the IHC reaction can assist the right diagnosis in difficult cases. Considering this rare benign pathology as a possible cause of pyloric obstruction prevents the over diagnosis of a malignant disease.

Conflict of interests

The authors declare that there is no conflict of interests.

Consent

The patient's written informed consent and the approval of the Hospital Ethics Committee were obtained for the publication of this Case Report and images.

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