



Jornal Brasileiro de Patologia e Medicina  
Laboratorial

ISSN: 1676-2444

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Sociedade Brasileira de Patologia  
Clínica/Medicina Laboratorial  
Brasil

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Jornal Brasileiro de Patologia e Medicina Laboratorial, vol. 51, núm. 2, março-abril, 2015,  
pp. 117-120

Sociedade Brasileira de Patologia Clínica/Medicina Laboratorial  
Rio de Janeiro, Brasil

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# Inflammatory fibroid polyp (Vanek's polyp): a case report and literature review

## *Pólipo fibroide inflamatório (pólipo de Vanek): relato de caso e revisão da literatura*

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### ABSTRACT

Inflammatory fibroid polyp (IFP) is a benign uncommon lesion (1%-4% of gastric benign lesions), originated from the submucosa of the gastrointestinal tract. Its origin is controversial and immunohistochemical studies of lesions have largely refuted the possible vascular, neural or smooth muscle origin. Recent studies suggest a neoplastic etiology due to a mutation, in some cases, in the alpha-type platelet-derived growth factor receptor gene (PDGFRa). Our objective is to report the case of a patient aged 70 years, with gastric IFP, comparing her immunohistochemical profile with those of other studies, and a brief review of the literature.

**Key words:** gastric polyp; inflammatory fibroid polyp; Vanek; immunohistochemistry.

### INTRODUCTION

The inflammatory fibroid polyp (IFP) is a benign lesion that arises from the submucosa of the gastrointestinal (GI) tract<sup>(1-3)</sup>, most commonly in the antrum (70%) and ileum (20%) and, only occasionally, in the duodenum and jejunum<sup>(4-7)</sup>. Its frequency ranges from 1% to 4% of diagnoses among benign stomach lesions, and usually occurs between the fifth and the seventh decade of life<sup>(1, 2)</sup>. Malignancy is a rare event<sup>(2)</sup>, however, up to 8% of IFPs have been described as concomitant lesions with adenoma or carcinoma<sup>(8)</sup>.

The lesion may be polypoid or sessile, usually single and covered with normal or ulcerated mucosa<sup>(2, 9, 10)</sup>. Histologically, IFP is composed of proliferation of spindle cell elements and a prominent network of small capillaries, in an inflammatory background<sup>(1, 2, 5, 8, 11, 12)</sup> composed of plasma cells, lymphocytes and eosinophils, more predominantly<sup>(4, 10, 12, 13, 14)</sup>.

Several studies on structural and immunohistochemical analysis of lesion has been conducted in order to clarify the origin of the IFPs, which, however, remains controversial<sup>(4, 15-17)</sup>. Several immunohistochemical studies have refuted a potential to vascular or neural origin for the lesion, since IFPs resulted negative for factor VIII-Ra and S-100, respectively<sup>(18, 19)</sup>, or a smooth muscle

origin due to a negative reaction for alpha smooth muscle actin (SMA), desmin and myoglobin<sup>(18)</sup>. Some studies have promoted the idea that fibroblasts are the main cells, primitive mesenchymal cells<sup>(18-21)</sup> or even dendritic cells<sup>(22)</sup>. Other, more recent, have shown a mutation in the alpha type receptor for the platelet derived growth factor (PDGFRa), suggesting a possible neoplastic factor in the etiology of this lesion<sup>(8, 14, 16)</sup>.

We report the case of a female patient aged 70 years with gastric IFP comparing her morphological and immunohistochemical features with the actual studies, and we reviewed the literature on this lesion.

### CASE REPORT

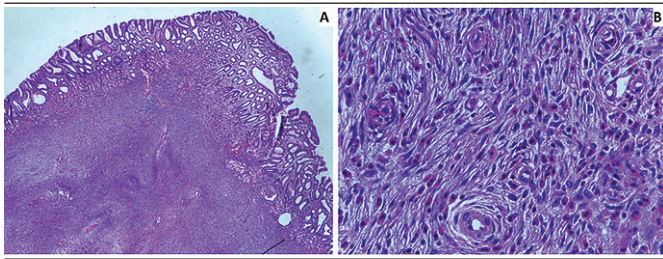
#### Case

A 70-year-old female patient with diabetes *mellitus*, undergoing well differentiated endometrioid endometrial adenocarcinoma follow-up, presented nausea, vomiting, and bloody stools lasting one week. Upper digestive endoscopy demonstrated expanded diaphragmatic hiatus and hiatal hernia with 3 cm slip, and pedunculated polyp in the antrum region, covered by preserved mucosa, similar to the adjacent, measuring

3 cm in diameter. There were no other abnormalities, and the endoscopic polypectomy of the lesion was analyzed.

### Histopathological analysis

Histopathological examination revealed a well-defined polypoid lesion, composed of proliferation of spindle cell elements, a prominent network of small capillaries and inflammatory cells located in the submucosa. The cells have ovoid nuclei, uniform chromatin, and abundant cytoplasm. The stroma is myxoid edematous, concentrically arranged around blood vessels with “onion skin” pattern. The inflammatory cells are predominantly composed of eosinophils (**Figure 1**). The overlying mucosa was preserved. Special staining for *H. pylori* caused negative result.



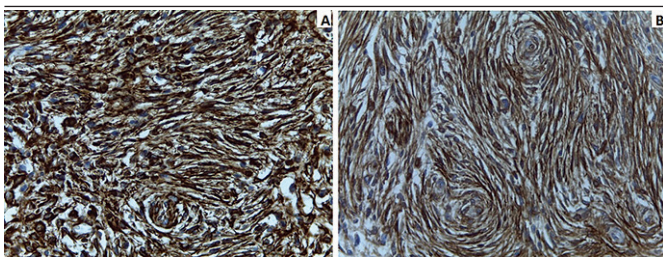
**FIGURE 1** – Histopathological images of the lesion

A) antral mucosa and submucosal IFP (HE, 25×); B) edematous myxoid stroma arranged in concentric layers around blood vessels, and inflammatory background predominantly consisted of eosinophils (HE, 400×).

IFP: inflammatory fibroid polyps; HE: hematoxylin and eosin.

### Immunohistochemistry

For immunohistochemical technique, the spindle cells of lesion were positive for CD34 (monoclonal, dilution 1:100, Novocastra) and vimentin (monoclonal, dilution 1:200, DAKO) (**Figure 2**) and negative for muscle actin smooth – SMA (monoclonal, dilution 1:50, DAKO), VIII-Ra factor (dilution 1:10, DAKO), CD117 – c-kit (polyclonal, dilution 1:100, DAKO – and S-100 protein (polyclonal, dilution 1:1200, DAKO). However, was positive for VIII-Ra factor and smooth muscle actin in the blood vessels walls, showing the rich vascularization of the lesion. IFP diagnosis was assigned.



**Figure 2** – Immunohistochemical profile of the lesion

A) vimentin-positive; B) CD34-positive.

### DISCUSSION

IFP was first described as “polypoid fibroma” in 1920 by Konjetzny<sup>(7)</sup>. In 1949, Vanek named it as “gastric submucosal granuloma with eosinophilic infiltration” and described six cases<sup>(2, 9, 23)</sup>. Later, it became recognized by a variety of different names: eosinophilic granuloma, polypoid fibroma, gastric fibroma with eosinophilic infiltration, polypoid eosinophilic granuloma, inflammatory pseudotumor, and Vanek’s polyp<sup>(2, 15, 16)</sup>. In 1953, Helwig *et al.* introduced the term “inflammatory fibroid polyp” to define the disease<sup>(1, 13, 15)</sup>.

IFP lesion that arises from the GI tract submucosa<sup>(2, 3, 15)</sup>, and are classified as submucosal tumor of connective tissue<sup>(4, 5, 12)</sup>; it is most commonly found in the antrum (70%) and ileum (20%)<sup>(2, 6, 11, 12)</sup>. Other sites in descending order are colon, jejunum, duodenum, and esophagus<sup>(9, 10)</sup>. Rarely it reaches more than 6 cm<sup>(4, 12, 15)</sup>, however there is a case of IFP reported with 12.5 cm diameter<sup>(3, 15)</sup>.

Most often, IFPs are incidentally discovered in asymptomatic patients<sup>(3, 10, 12)</sup>. However, depending on the size and location of the lesion, patients may present dyspepsia symptoms, such as abdominal epigastralgia-type pain, nausea and, sometimes, vomiting, in addition to gastrointestinal bleeding, iron-deficiency anemia and change in bowel habits<sup>(1, 4, 8, 12, 16, 24)</sup>.

The typical macroscopic aspect is a polypoid, sessile or pedunculated lesion, usually single, well defined, without capsule, firm, with grayish surface and covered with normal or ulcerated mucosa<sup>(2, 9, 10)</sup>. Histologically, it is characterized by a submucosal lesion<sup>(12, 22)</sup>, which rarely extends to the muscle layer<sup>(9)</sup>, composed of small blood vessels and mesenchymal spindle cell proliferation<sup>(1, 2, 11, 12, 14)</sup> with ovoid nuclei, fine granular chromatin, small nucleoli and eosinophilic cytoplasm<sup>(9)</sup>. It also presents inflammatory infiltrates containing lymphocytes, plasma cells and eosinophils, predominantly<sup>(2, 5, 13, 11, 12, 14)</sup>, and a edematous myxoid stroma arranged, generally concentrically around blood vessels, with “onion skin” pattern<sup>(2, 10)</sup>. The overlying mucosa may be normal or ulcerated, inflammation or regeneration secondary to erosion<sup>(8)</sup>. Mitotic activity and reactive changes are occasionally seen in the superficial epithelium<sup>(9)</sup>.

The differential diagnosis is challenging, even at the microscopic level, and immunohistochemical analysis is performed to differentiate from other tumors, such as gastrointestinal stromal tumor (GIST), inflammatory myofibroblastic tumor (IMT) and schwannoma<sup>(14, 16)</sup>. IMT is very similar to IFP, with myofibroblast proliferation and mixed inflammatory component<sup>(9)</sup>. The

immunohistochemical analysis of this lesion is positive for smooth muscle actin, and negative for CD34<sup>(13, 14, 16)</sup>. GIST is a intramural tumor, positive to CD117<sup>(8, 9, 13, 14)</sup> and CD34<sup>(14)</sup>. Schwannoma arise in the lamina propria, has no eosinophils or other inflammatory cells scattered in the stroma and is positive for S-100 protein<sup>(14, 25)</sup>.

IFP may have variable reactivity for smooth muscle actin, CD34, CD117 (c-kit), and S-100 protein<sup>(5, 7, 10, 12, 22)</sup>. In this case, the immunohistochemical analysis excluded GIST (negative for CD117), inflammatory myofibroblastic tumor (negative for smooth muscle actin) and schwannoma (negative for S-100). Its etiology is still controversial<sup>(5, 10-12, 15-17, 22)</sup>, but several hypotheses have been proposed, the most accepted, until recently, the inflammatory theory<sup>(2)</sup>, according to which, IFP originates due to excessive tissue reaction to damage applied on the gastrointestinal mucosa, as chemical, mechanical or biological factors<sup>(2, 5, 6, 10-12, 15)</sup>. It is believed that these factors act on mucosal fibroblasts and myofibroblasts, stimulating its growth<sup>(2)</sup>. An allergic etiology has also been proposed due to the presence of eosinophilic infiltration<sup>(9, 6, 11, 17)</sup>, however, in most cases there is no medical history of allergies and eosinophilia, which is present in only 4% of patients<sup>(16, 17)</sup>. The emergence of these polyps in gastric location has been associated to *Helicobacter pylori* infection, entailing the hypothesis of an immune response during a chronic infection by this bacterium<sup>(1, 2, 8, 17, 26)</sup>. Some studies suggest that gastric IFPs are more frequent in patients with atrophic gastritis and pernicious anemia<sup>(8)</sup>. Other autoimmune diseases have also been linked to IFP, such as sarcoidosis, rheumatoid arthritis and ankylosing spondylitis, which confirms the possibility of an immune reaction as a contributing factor<sup>(1, 11, 17)</sup>.

In this report, the spindle cells of the lesion were positive for CD34 and vimentin, and negative for SMA actin, factor VIII-Ra,

CD117 (c-kit) and S-100 protein. However, the positivity for factor VIII-Ra and smooth muscle actin in the vessels walls showed the rich vascularity of the lesion, which may indicate a secondary involvement of vascular proliferation<sup>(18)</sup>. Our results corroborate the literature when indicating that the lesion presents a possible neural, vascular or smooth muscle origin<sup>(18, 19)</sup>, encouraging the idea that the main cells are fibroblasts or primitive mesenchymal cells, primarily because it is strongly positive for vimentin and CD34<sup>(18-21)</sup>. In a study of 14 IFP cases, three cases were negative for CD-34, and all of positive cases for this marker had concentric stromal proliferation<sup>(18)</sup>. The authors concluded that IFP with concentric proliferation may have a different histogenesis from IFP without this type of stromal pattern, and that it may originate from a subpopulation of dendritic interstitial cells<sup>(18, 22)</sup>. Our case presented stroma concentrically organized around the vessels and positive for marker CD34.

Recent studies have shown mutations in exons 12<sup>(26, 27)</sup> or 18<sup>(26, 28)</sup> and, less frequently, in exon 14<sup>(26)</sup> of PDGFRa gene (chromosome 4q12), suggesting a possible neoplastic factor in the etiology of this lesion<sup>(26-28)</sup>. A total of 145 IFP cases, 55.2% had mutations in exons 12 or 18 of PDGFRa gene, and only two cases presented mutation in exon 14<sup>(26)</sup>. Studies on mutation of this lesion have shown that a type of unidentified mesenchymal stem cells of the GI tract submucosa is mutated, developing a tumor<sup>(26, 27)</sup>.

Thus, IFPs present a possible major component of spindle cells, recognized as fibroblast and primitive mesenchymal cells, and studies on Mutation Research should be conducted to elucidate the mechanisms that lead to the origin of this lesion.

## RESUMO

*Pólipo fibroide inflamatório (PFI) é uma lesão benigna, pouco frequente (1%-4% das lesões benignas gástricas), originada na submucosa do trato gastrointestinal. Sua origem é controversa e vários estudos imuno-bistoquímicos da lesão refutaram uma possível origem vascular, neural ou muscular lisa. Recentes estudos sugerem etiologia neoplásica devida à mutação, em alguns casos, no gene receptor tipo alfa para fator de crescimento derivado de plaquetas (PDGFRa). Nosso objetivo é relatar o caso de uma paciente de 70 anos de idade, com PFI gástrico, comparando seu perfil imuno-bistoquímico com o de outros estudos, além de breve revisão da literatura.*

*Unitermos: pólipo gástrico; pólipo fibroide inflamatório; Vanek; imuno-bistoquímica.*

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