

Ileal Inflammatory Fibroid Polyp: Report of Cases

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Abstract: Inflammatory fibroid polyp is a rare benign lesion of the gastro-intestinal tract. The symptoms are heterogeneous and depend on their location and size of the tumor. The pathogenesis of this lesion remains unclear. We report 2 cases of inflammatory fibroid polyp, the gross appearance showed polypoid masses. Histologically, the lesion consisted of spindle cell dispersed in a loose fibromyxoid stroma containing numerous blood vessels and inflammatory cells with abundant eosinophils. At immunohistochemistry, the stroma is positive for CD34 but negative for CD117. Definitive diagnosis is made through histopathology and it has excellent long-term prognosis.

Key words: Inflammatory fibroid polyp, Vanek's tumor, Gastrointestinal polyp.

I. Introduction

Inflammatory fibroid polyps (IFPs) are rare mesenchymal lesions of the gastrointestinal (GI) tract. First described by Vanek in 1949 as a submucosal gastric granuloma with eosinophilia [1]. Helwing and Ranier proposed the term IFP in 1953 [2]. The stomach is the most common site, but it has been described elsewhere. Its incidence is extremely low (0.1 %) and usually presents in the sixth and seventh decades of life. It is slightly more frequent in the male patients [2][3]. Its clinical symptoms are heterogeneous and essentially depend on the location and size of the tumor. Definitive diagnosis is made through histopathology [4][5] and this pathology has excellent long-term prognosis [6].

Observation 1:

A 18-year-old patient with no significant pathological history, admitted to the emergency department for an occlusive syndrome.

On clinical examination, there is a slightly distended abdomen, with no other abnormality.

Abdomen radiography without preparation showed dilated segments of the small intestine with marked hydro-aerobic levels. Abdominopelvic CT SCAN showed an occlusion on ileo-ileal intussusception most likely secondary to a Meckel diverticulum.

Resection of the invaginated part with end-terminal anastomosis and biopsy of mesenteric lymphadenopathy were performed.

Macroscopic examination showed a polyp of 4x2x2 cm, it's located at 8 cm from one limit and at 13 cm from the other (Figure 1).



Figure 1: Ileal segment showing a polyp with 4 cm in its longest axis.

Microscopic 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 2 and 3).

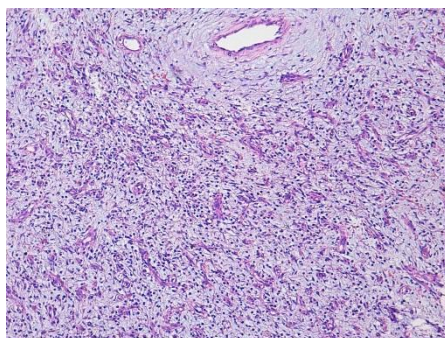


Figure 2: Loose, fibromyxoid spindled stroma.

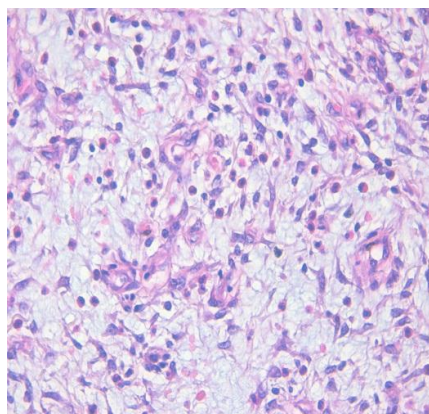


Figure 3: Numerous eosinophils in a typical inflammatory fibroid polyp

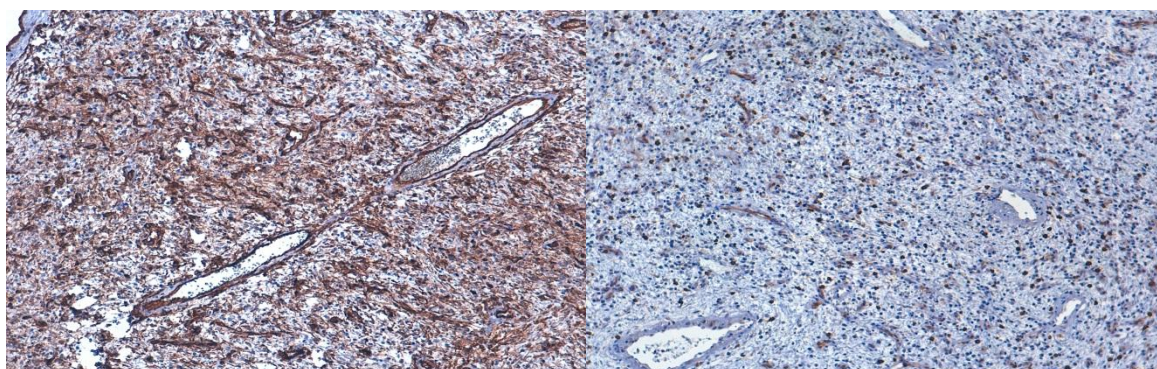


Figure 4: Spindle cells are CD34+, and CD 117- (mast cells are CD 117+).

Immunohistochemical studies showed that the large fusiform cells stained negative for antibodies CD 117, but positive for antibodies CD34 (Figure 4) .

Observation2:

45-year-old patient with a history of arterial hypertension under treatment who had abdominal pain, vomiting and nausea and anemic syndrome, he had also intermittent constipation associated with weight loss in the previous year.

Abdomen radiography without preparation showed dilated segments of the small intestine with marked hydro-aerobic levels. Esophagogastroduodenoscopy showed the presence of a polypoid lesion measuring 2 cm whose pedicle is long and wide with ulceration on the surface.

Abdominopelvic CT scan showed a grelic intussusception with no visible underlying cause. 3 hypodense liver nodules were also found.

Resection of the invaginated part with end-terminal anastomosis and liver biopsy were performed.

Macroscopic examination showed a pedonculated polyp of 3.2x2.5x2.5 cm. Located at 6.5 cm from one limit and at 6 cm from the other.

Histologically, the polyp was lined by a mucosecreting columnar epithelium which is largely ulcerated on the surface. The glands are of normal size, bordered by columnar cells, without lesions of dysplasia, the lamina propria and the submucosa contain a polymorphic inflammatory infiltrate made essentially of mononuclear cells and eosinophilicpolynuclear cells.

Multiple thick-walled vessels with an onion skin appearance were present.

The fibrous tissue harboring these elements is focally edematous.

The histological study of the liver biopsy concluded to a parasitic abscess.

II. Discussion

The IFP is a rare disease, it occurs mainly in adults between the sixth and seventh decades of life; it is rare in children, and can appear in all age groups. It affects the stomach (66%–75%), followed by the small intestine (18%–23%), mainly ileum, colon, and rectum (4%–7%), gallbladder (1%), esophagus (1%), duodenum (1%), and appendix (<1%). Since instances, synchronic and metachronic disease has already been described.

Clinical presentation depends on the site of involvement. Patients may present with abdominal pain, anemia, GI bleeding, or obstruction, the latter most often in the case of small intestine tumors, which may present emergently with intussusception.

On gross examination, IFPs are typically solitary, polypoid or sessile lesions measuring less than 5 cm. They are submucosal-based lesions, but the majority of cases also involve the mucosa, with smooth or ulcerated surfaces.

Histologically, inflammatory fibroid polyps are hypocellular lesions composed of bland ovoid, short spindled, stellate, or more epithelioid cells with fine chromatin, inconspicuous nucleoli, and small amounts of eosinophilic cytoplasm haphazardly arranged in a loose, predominantly edematous to myxoid stroma containing a conspicuous inflammatory infiltrate, chiefly of eosinophils, but also histiocytes and lymphocytes. Capillaries and small blood vessels are often prominent, a subset of which show perivascular, lamellar (onion-skin) fibrosis. Occasionally, tumors contain more collagenous stroma. The tumor often infiltrates and ulcerates the mucosa and may be difficult to distinguish from adjacent granulation tissue, particularly in small biopsies. Larger tumors may infiltrate through the muscularis propria into subserosa.

By immunohistochemistry, most inflammatory fibroid polyps are positive for CD34, whereas about 10%–20% of cases show focal reactivity for SMA. Desmin, KIT, DOG1, and S-100 are consistently negative[7].

Its etiology is unknown, but some authors suggest that it is an allergic reaction due to the presence of eosinophils. However, other factors have been implicated, such as neural hyperplasia, irritants, trauma, genetic alterations, and bacterial, physical or chemical stimulants[2][8]. Activating mutations in the platelet-derived growth factor receptor alpha (PDGFRA) gene have been associated with the development of IFPs and of gastrointestinal stromal tumor (GIST), showing a similar physiopathogenesis between these 2 neoplasms and lending support to the theory of a neoplastic origin.[8][9].

The differential diagnosis of inflammatory fibroid polyp primarily includes inflammatory myofibroblastic tumors (IMTs), Gastro-intestinal stromal tumors (GIST), and conventional inflammatory polyps.

IMTs are more cellular than inflammatory fibroid polyps, and are composed of plump, elongated spindle cells arranged in fascicles. In terms of the inflammatory infiltrate, plasma cells and lymphocytes usually predominate in IMT, whereas eosinophils are relatively uncommon. IMTs are usually positive for SMA and may also express desmin, whereas CD34 is consistently negative; 50% of tumors show reactivity for ALK, correlating with the presence of ALK gene rearrangements. GISTs are generally uniformly cellular and lack the loose, edematous stroma and prominent eosinophils and small blood vessels of inflammatory fibroid polyps. KIT and DOG1 expression are specific for GIST in this differential diagnosis. Small inflammatory fibroid polyps may be mistaken for inflammatory polyps, but inflammatory polyps often contain irregular and dilated crypts containing neutrophils, and the stroma lacks the uniform hypocellularity and stellate to epithelioid cytology of inflammatory fibroid polyps.

Among other digestive tumors with fusiform cells: the leiomyoma is more fasciculate architecture, with a clear positivity of muscle markers (actin, desmin, H-caldesmon). Schwannoma is much rarer in digestive localization; it consists of fusiform cells often expressing PS100, which may be associated with a rather typical lymphocyte infiltrate at the periphery.

IFPs are benign lesions. In symptomatic cases, the treatment of choice is endoscopic excision or surgical resection, depending on the size of the polyp. Following resection, IFPs typically do not recur.

III. Conclusion

The IFP is a rare disease. Its pathogenesis is still unknown; the symptoms depend on their location. The preoperative diagnosis is rarely established before the operation and is usually made after resection.

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