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Case Report

Inflammatory fibroid polyps of small and large intestine: Report of two cases

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ABSTRACT

Inflammatory fibroid polyps (IFPs) are uncommon benign solitary mesenchymal neoplasms which may develop anywhere in the gastrointestinal tract. These submucosal lesions can occur in individuals of any age but are more frequently observed in the 6th or 7th decade of life. Although rarely associated with adenoma or adenocarcinoma, IFPs are typically managed through local excision. The clinical presentation varies depending on the anatomical location of the polyp. Gross examination typically reveals localized submucosal sessile polypoidal masses. Microscopically, IFPs are characterized by inflammatory cell infiltrate, spindle shaped cells and prominent capillaries. These benign lesions have unknown pathogenesis. Below mentioned are two case report of inflammatory fibroid polyps which lead to intussusception.

1. Introduction

Inflammatory fibroid polyps (IFPs) are rare, solitary, benign mesenchymal polypoid neoplasm centered in the submucosa of the gastrointestinal tract. Uncommonly known as Vanek's polyps, this terminology is not used now. They are also described as submucosal fibroma, eosinophilic granuloma, fibroma, inflammatory pseudotumor and hemangiopericytoma.[1] IFPs may occur throughout the gastrointestinal tract but are more frequently found in the gastric antrum.[2] Their incidence rate ranges from 0.1% to 2%.[3] IFPs can appear at any age but are most common in individuals in their 6th to 7th decades of life, with an equal distribution between sexes.[4] Clinical features vary depending on the lesion's location, and they are often diagnosed incidentally in asymptomatic patients.[5] It is uncommon for inflammatory fibroid polyps to cause ileal intussusception.[6] Recognized as benign mesenchymal neoplasms specific to the gastrointestinal tract, the exact cause of IFPs is not fully understood. However, continuous mutations in the PDGFRA (platelet derived growth factor receptor alpha) gene have been identified. The below given table shows details of two cases of inflammatory fibroid polyps in the small and large intestines.

2. Case Report

2.1. Case 1

A 47-year old female came with acute, sudden lower abdominal pain, non radiating in nature since 2 days in surgical department. Radiological examination revealed evidence of bowel within bowel appearance (target sign) is noted in hypogastric region suggestive of Intussusception. The length of involved segment measures approximately 6 cm. On exploratory laparotomy, a mass was appreciated at the site of intussusception therefore Intussusception reduction, jejunojejunal resection and anastomosis was done and a mass appreciated at the site of intussusception was sent for histopathological evaluation.

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Table 1:

S. No.	Age	Gender	Clinical presentation	Radiological findings	Histopathology
1.	47 years	Female	Lower abdominal pain	Intussusception	Inflammatory fibroid polyp
2.	65 years	Male	Right lumber pain	Intraluminal polyp lesions-neoplastic likely	Inflammatory fibroid polyp

2.1.1. Gross examination

Specimen labelled as mass from jejunal loop measuring 3.7 × 3.5 × 1.7 cm. Polypoidal submucosal mass is measuring 2.5 × 2.0 × 1.7 cm. On cut-section, it is whitish and smooth.

Figure 1:

2.1.2. Microscopic examination

On H & E sections from jejunal mass shows unremarkable jejunal mucosa with submucosal lesion composed of proliferation of bland, ovoid to spindled cells in loose edematous and collagenous stroma. There is presence of marked vascularity with eosinophilic inflammatory cell infiltrate. No evidence of giant cells, mitosis or necrosis is seen. Immunohistochemistry was advised. The immunohistochemistry report showed CD 34 positive and CD 117 negative.

2.1.3. Diagnosis - inflammatory fibroid polyp of jejunum

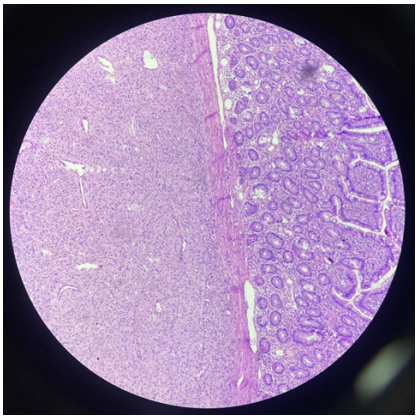

Figure 2: Inflammatory fibroid polyp arising from the submucosa of the jejunum (H& E, 10x)

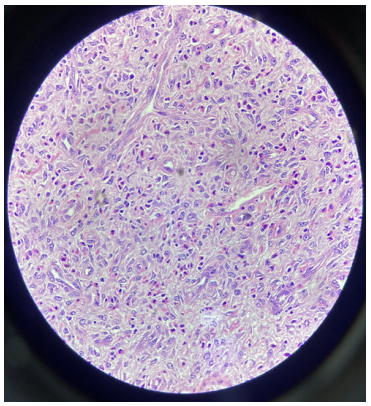

Figure 3: Bland spindle shaped cells and background of inflammatory cells with prominent numbers of eosinophils (H& E, 40x)

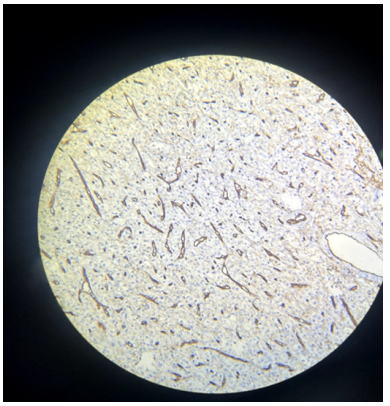

Figure 4: IHC study for CD34 at 10x magnification showing positivity

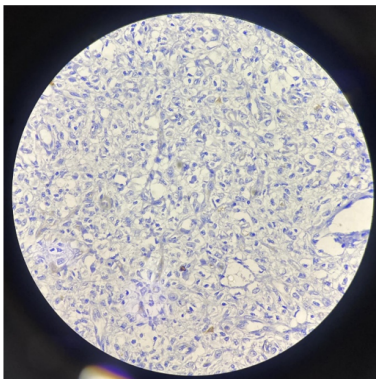

Figure 5: IHC study for CD117 is negative

2.2. Case 2

A 65 years old male came with right lumber pain, spasmodic on & off since 10 days. Radiological examination revealed colocolonic Intussusception at hepatic flexor of colon with circumferential enhancing wall thickening and intraluminal polyp lesion - neoplastic likely.

2.2.1. Gross examination

Specimen labelled as colonic polyp consists of single brownish tissue portion total measuring 3 × 2.4 × 1.6 cm.

2.2.2. Microscopic examination

Sections show unremarkable colonic mucosa with submucosal lesion composed of proliferation of bland, ovoid to spindled cells in loose edematous and collagenous stroma. There is presence of marked vascularity with eosinophilic inflammatory cell infiltrate. No evidence of giant cells, mitosis or necrosis is seen.

2.2.3. Diagnosis - inflammatory fibroid polyp

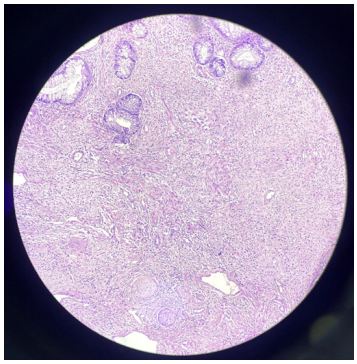

Figure 6: Bland spindle shaped cells and background of inflammatory cells with prominent numbers of eosinophils (H& E, 10x)

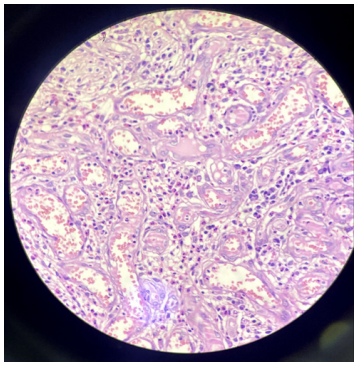

Figure 7: Vascular proliferation and background of inflammatory cells with prominent numbers of eosinophils (H& E, 40x)

3. Discussion

Inflammatory fibroid polyps (IFPs) are uncommon, benign, usually solitary, sessile or pedunculated submucosal polyps found in the gastrointestinal tract.[6] IFPs predominantly arise in the gastric antrum (70%) near the pyloric sphincter, followed by the small & large intestines (25%), esophagus (01%), gallbladder (01%), duodenum (01%), anal canal (01%), and appendix.[2] These polyps show variation in size ranging from 1.5 to 13 cm and are polypoidal with a broad base.[1] Patients with gastric IFPs may present with symptoms such as vomiting and bleeding, while those with lesions in the small bowel might experience intussusception and obstruction.[7] In the colon, symptoms can include weight loss, diarrhea, bleeding, and anemia.[8] Adult intussusception is relatively rare, accounting for only 1% of bowel obstruction cases, and IFPs are an uncommon cause of ileal intussusception. Duodenal IFPs, which account for 1% of cases, typically present with non-specific symptoms. The exact pathogenesis of IFPs remains unclear, though trauma, allergic reactions, genetic predispositions, and various physical, chemical or metabolic triggers have been suggested.[9] IFPs are usually limited to submucosa but it can infiltrate the mucosa or muscularis propria and may extend to the serosa.[1] Grossly, they appear tan, yellow or gray with potential ulceration of the overlying mucosa. Microscopically, these are mesenchymal lesions featuring an inflammatory infiltrate and a variable vascular component. The fibroblasts are stellate or spindle-shaped with indistinct basophilic cytoplasm. Certain regions of the lesions can also exhibit sparse cellularity with a notable myxoid component. In more cellular areas, cells may display mitotic figures, with few lesions showing up to 2 mitoses per high-power field (HPF). The inflammatory infiltrate includes eosinophils, plasma cells, lymphocytes, mast cells and macrophages, with eosinophils varying in number. Lesions with fewer eosinophils may present areas of significant stromal hyalinization. The vascular component can be significant, with many lesions showing distinctive zone of loose connective tissue around larger blood vessels.[1] Inflammatory fibroid polyps exhibit various histopathological patterns, including classical fibrovascular, nodular, sclerotic, edematous, and atypical. Differential diagnoses include eosinophilic gastroenteritis, gastrointestinal stromal tumors (GISTs), schwannoma, leiomyoma and other benign nerve sheath lesions.[1] Morphologically, IFPs can mimic many tumorous and non-tumorous processes, including inflammatory myofibroblastic tumors, gastrointestinal stromal tumors, eosinophilic gastroenteritis and various other mesenchymal lesions. Differentiating between inflammatory fibroid polyps and gastrointestinal stromal tumors (GIST) can be particularly challenging.[10] On histomorphological features differential diagnosis include GIST, leiomyoma/sarcoma, schwannoma, intra-abdominal

fibromatosis. The IHC plays a key role in the final diagnosis of GIST. Immunohistochemically GISTs uniformly express CD117 (95%), CD 34 and SMA. The 5% of tumors are positive for PDGFR mutation.[11] Unlike GISTs, it exhibits a coarsely granular cytoplasmic texture with prominent membranous accentuation. Immunohistochemically, stromal cells in IFPs are positive for PDGFRA, vimentin, CD34, fascin, cyclin D1, CD35 and calponin, and negative for CD117.[12] Care must be taken when interpreting CD117 stains in these lesions because mast cells, which are often abundant, will also stain positively. Activating PDGFRA mutations are commonly seen, with exon 12 mutations prevalent in small intestine lesions and exon 18 mutations in gastric lesions.[1] Although IFPs are uncommonly associated with adenoma or adenocarcinoma, local excision is usually curative, and no further endoscopic surveillance is required after histological confirmation.

4. Conclusion

Inflammatory fibroid polyps (IFPs) are an extremely rare histologic entity that must be distinguished from other mesenchymal tumors of the gastrointestinal tract, especially those with spindle shaped tumor cells. Clinically, Inflammatory fibroid polyps may be confused with neoplastic polyps, particularly in adults, making preoperative diagnosis challenging. Therefore, a combination of clinical, endoscopic, and histopathologic evaluation is necessary for accurate diagnosis. Immunohistochemical (IHC) staining is valuable for confirming the diagnosis and excluding other differential diagnoses. Inflammatory fibroid polyps are benign lesions that are typically treated with local excision.

5. Source of Funding

None.

6. Conflict of Interest

None.

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