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Inflammatory Fibroid Polyp of the Small Intestine: An Extremely Rare Cause of Intussusception

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Abstract: ***Introduction: Inflammatory fibroid polyps (IFP) are rare, benign lesions of the gastrointestinal tract. Recent molecular studies of IFPs identified platelet - derived growth factor receptor α (PDGFRA) - activating mutations, suggesting possible neoplastic qualities to IFPs. IFPs originate from the submucosa and often extend into the overlying mucosa. Although certain IFPs infiltrate the muscularis propria focally, disruption of the muscularis propria and penetration into the subserosa is extremely rare. Methods: The current report presents an unusual case of an ileal IFP. A 48 - year - old female visited the Department of Surgery, SGT Hospital due to abdominal pain. Radiological study demonstrated an ileocecal - type intussusception due to a luminal polypoid mass of the ileum. Results: Excised specimen sent for a histopathology had a histologic diagnosis of inflammatory fibroid polyp. After six months of follow - up, the patient is in good health and no evident recurrence of the tumor has been detected.***

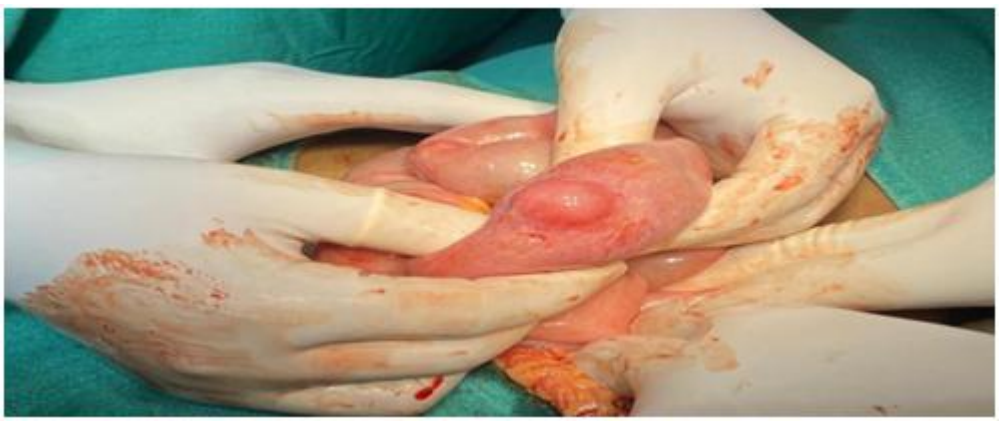
Keywords: Inflammatory fibroid polyp (IFP), Intussusception

1. Introduction

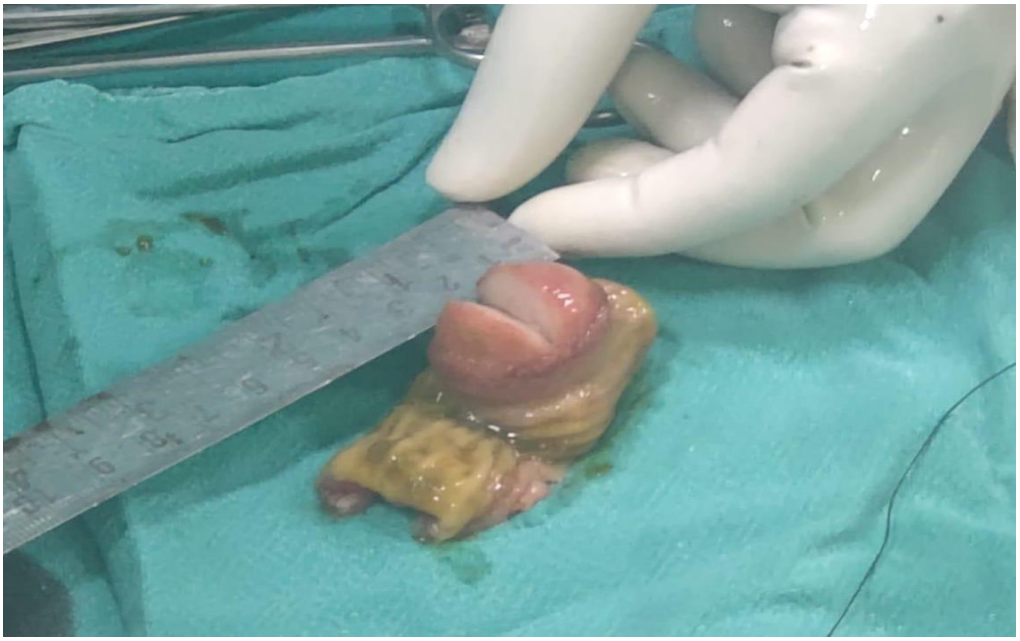
Inflammatory fibroid polyp (IFP), a rare benign polypoid lesion that arises in the submucosa of the gastrointestinal tract is composed of fibrous connective tissue, blood vessels, and variable inflammatory infiltrates, composed primarily of eosinophils. The lesion is usually solitary attd arises in the gastric antrum, although the ileum was the primary site in 25 cases recently described, First described by Vanek in 1949 as submucosal granulomas with eosinophilic infiltration, inflammatory fibroid polyps have been referred to by a variety of names, including eosinophilic granuloma, granuloma with eosinophils, inflammatory pseudotumor, gastric fibroma with eosinophilic infiltration, eosinophilic gastroenteritis, and polyp with eosinophilic granuloma. Due to the lack of a known etiologic factor and to the fact that most authors favor an inflammatory pathogenesis, the term inflammatory fibroid polyp, first proposed by Helwig and Rainer, seems to be the most appropriate label. We present a case of IFP of the terminal ileum; the lesion, the largest reported to date, had an extensive extramural component, a feature that has not been described previously.

2. Case Report

A 48 - year - old female visited the Department of Surgery, SGT Hospital due to abdominal pain. Radiological study demonstrated an ileocecal - type intussusception due to a luminal polypoid mass of the ileum. Patient was posted for exploratory laparotomy. Intra - op Findings were:



Intra - operative photograph - Showing subserosal extension of IFP



Excised specimen of mass – Showing submucosal extension of IFP

3. Discussion

IFPs are rare mesenchymal lesions of the digestive system that were originally described by Vanek (9); they are considered reparative processes. Recent molecular studies of IFPs identified PDGFRA - activating mutations, suggesting a neoplastic nature to this lesion. Histologically, IFPs are characterized by proliferation of spindle, stellate or epithelioid cells accompanied by an inflammatory reaction, particularly eosinophilic granulocytes. The tumor cells are embedded in an edematous, fibromyxoid stroma, with prominent vasculature. A characteristic finding is the presence of concentric cuffing of the glands or vessels by spindle shaped tumor cells. Concentric cuffing of tumor cells is more pronounced in gastric IFP than in intestinal IFP. IFPs tend to arise within the submucosa and often involve the mucosa. In the present case, the IFP exhibited the following unusual features: Firstly, the tumor cells obliterated the muscularis propria and extended into the subserosa. IFPs are considered benign tumors without any risk for recurrence or metastasis following complete excision. The majority of IFPs are centered within the submucosa and rarely spread to the muscularis propria (8). A study by Makhlouf and Sobin demonstrated that among 45 reported cases of IFP, none extended into the serosa or subserosa tissue, although 7% of small intestinal IFPs infiltrated the muscularis propria. A search of the literature found only one similar case study describing a rectal IFP that exhibited attachment to the sacral bone, thus mimicking rectal cancer (10). Secondly, despite low tumor cellularity, the tumor cells demonstrated relatively high proliferative activity, as shown by Ki - 67 labeling and mitotic figures. The majority of IFP cases have few or no mitotic figures, and Ki - 67 labeling is <1% (7, 8). In the study by Makhlouf and Sobin, only two out of the 45 IFPs showed 2 - 4 mitoses/50 HPFs, and no IFPs had more than 5 mitoses/50 HPFs (8). The present findings of subserosal extension of the lesion with disruption of the muscularis propria by infiltrating tumor cells and proliferative activity of the tumor cells supported a neoplastic nature of the current IFP. The mutation and activation of PDGFRA contribute to the development of IFPs. Huss *et al* reported that exon 12 PDGFRA mutations are more often associated with small intestinal IFPs, whereas exon 18 PDGFRA mutations occur frequently in gastric IFPs. A PDGFRA mutation or PDGFR expression was not identified in the present case. The reason for the lack of PDGFRA mutation for the current IFP is unclear. A possible explanation is a false - negative sequence analysis possibly resulting from the cellularity of these lesions, or the degradation of tumor DNA in the paraffin blocks, among other factors. This assumption is less likely in the present case, which showed no PDGFR expression, as none of the PDGFR - negative IFPs revealed PDGFRA mutations, although they showed typical histological features of IFPs (7). It has also been suggested that certain small intestinal IFPs are caused by molecular mechanisms other than PDGFRA activation (6). Indeed, previous studies showed that only 47 of 81 (58.0%) IFPs of the small intestine carried mutations in the PDGFR gene. It is difficult at present to elucidate the underlying mechanisms of this condition; further studies with a larger number of similar cases will be necessary to characterize the phenotype and nature of this tumor. After six months of follow - up, the patient is in good health and no evident recurrence of the tumor has been detected.

4. Conclusion

In conclusion, we report a case of inflammatory fibroid polyp of the small intestine presented with intussusception. Adult intussusception due to IFP is even more rare condition. A CT scan is most important diagnostic tool to detect IFP, however a histopathological evaluation is needed to confirm diagnosis.

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