Inflammatory Fibroid Polyp of the Ileum: A Case Report and Review of Literature

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Abstract

Introduction: Inflammatory fibroid polyps (IFPs) are rare, benign, tumour-like lesions of the gastrointestinal tract. IFPs usually present in the sixth or seventh decade. These polyps cause obstruction or intussusception when localised in the small intestine.

Case Report: A 70 years old female patient presented to the emergency with a history of diarrhoea, right sided lower abdominal pain, nausea and vomiting for two weeks. Abdominal ultrasound revealed a hypoechoic, round, soft tissue mass measuring 38 mm × 37 mm at the end of a dilated, thick-walled ileal loop at the right iliac fossa. Segmental resection of the intussuscepted ileum and end-to-end anastomosis were performed. On gross examination a tumour measuring 4 cm in diameter was identified. Cut surface of the tumour was grey white and homogeneous. Representative micro sections examined showed a submucosal lesion composed of an edematous stroma containing spindle-shaped, cytologically bland stromal cells, lymphoid nodules, thin walled blood vessels with onion skin arrangement of spindled cells around vessels. On immunohistochemical analysis, the spindle cells were negative for CD117, smooth muscle actin and positive for CD34. A diagnosis of Inflammatory fibroid polyp (Vanek's polyp) of the ileum was made.

Conclusion: When histopathologically diagnosing IFPs, other spindle cell lesions, such as GISTs, schwannomas, and inflammatory myofibroblastic tumors (IMTs), should be ruled out. In case of doubt, a limited panel of immunohistochemical stains (immunoreactivity for CD34, and absence of CD117 and S-100 protein) can be used to establish the correct diagnosis.

Keywords: Inflammatory Fibroid Polyp, GIST, CD-34

Introduction

Inflammatory fibroid polyps (IFPs) are rare, benign, tumour-like lesions of the gastrointestinal tract. Most commonly, they are localised in the gastric antrum, but can develop anywhere in the gastrointestinal tract. These polyps cause obstruction or intussusception when localised in the small intestine. (1)

IFPs usually present in the sixth or seventh decade, but cases have been reported over a wide age range from 5 months to 92 years old. (2)

Because there are usually asymptomatic, especially in their early stages and often go unrecognised until very severe symptoms ensue, which can create surgical emergencies. Clinical presentation is generally non-specific and chronic, including abdominal pain with or without nausea, vomiting, anemia, diarrhoea. (3)

Macroscopically, these lesions are pedunculated or sessile, measure 0.2-20 cm in diameter, arise from the submucosa, and project into the bowel lumen. The mucosal surface is usually ulcerated and pale. (1)

Microscopically, IFPs are submucosa-based lesions
of bland spindle cells with prominent vascularization. A characteristic finding and often a clue to the diagnosis is the presence of concentric cuffing of vessels by the lesional cells, referred to as an “onion skinning” appearance. Numerous eosinophils are also a common feature.(4)

CASE REPORT

A 70 years old female patient presented to the emergency with a history of diarrhoea, right sided lower abdominal pain, nausea and vomiting for two weeks. On abdominal examination, mild abdominal distension and tenderness was observed. The lab investigations performed upon admission were within normal limits. Abdominal ultrasound revealed a hypoechoic, round, soft tissue mass measuring 38 mm × 37 mm at the end of a dilated, thick-walled ileal loop at the right iliac fossa. The clinical symptoms and ultrasonographic findings were consistent with intussusception caused by a tumoral mass. Segmental resection of the intussuscepted ileum and end-to-end anastomosis were performed. The resected segment of the ileum was 6.5 cm in length.

On gross examination a tumour measuring 4 cm in diameter was identified. Cut surface of the tumour was grey white and homogeneous. Representative micro sections examined showed a submucosal lesion composed of an edematous stroma containing spindle-shaped, cytologically bland stromal cells, lymphoid nodules, thin walled blood vessels with onion skin arrangement of spindled cells around vessels. Diffuse inflammatory cells infiltrate comprising of large number of eosinophils, plasma cells and lymphocytes was also seen. The background showed collagen deposition, smooth muscle proliferation and occasional giant cell. The tumour at places was seen extending up to the mucosal layer and causing focal ulceration with granulation tissue, giving strophic appearance to the mucosa.

On immunohistochemical analysis, the spindle cells were negative for CD117, smooth muscle actin and positive for CD34. A diagnosis of Inflammatory fibroid polyp (Vanek's polyp) of the ileum was made.

DISCUSSION

IFPs were first described in the literature as "gastric submucosal granuloma with eosinophilic infiltration" in a study by Vanek in 1949. They later became recognised under a variety of different names: inflammatory pseudo-tumour, eosinophilic granuloma, and polyp with eosinophilic granuloma. Four years after their initial appearance in the literature, the term inflammatory fibroid polyp was introduced in a study by Helwig et al. and has since become the most widely used. (5)

IFPs can develop in many different locations in the gastrointestinal tract. The most common site is the gastric antrum (66%-75%), followed by the small bowel (18%-20%), colorectal region (4%-7%), gallbladder (1%), esophagus (1%), duodenum (1%), and appendix (<1%). However, the ileal segment is the most common site where these polyps cause intussusception. (2)

The aetiology of inflammatory fibroid polyps is unknown. Triggers including a foreign body, parasite and chronic Helicobacter pylori infection have been suggested but remain unsupported and theoretical. (6)

In 2008, Schildhaus et al. identified PDGFRA mutations in IFPs. (7) The presence of PDGFRA mutations provides strong evidence of clonal proliferation and suggests that IFPs have a neoplastic nature. Till date, IFPs are considered true neoplastic lesions rather than reactive lesions. (8) In 2012, Huss S et al. revealed that approximately 55% of IFPs have PDGFRA mutations and that most non-mutated IFPs are small lesions.

Clinical presentation depends on the site of involvement. Gastric and colon IFPs are typically identified incidentally, whereas small intestinal lesions are often encountered in the setting of intussusception. (4) Benign tumours, often lipomas, are the most common specific causes of adult intussusception. IFPs have been reported as a rare cause of adult intussusception. (9,10)

Histologically, IFPs are submucosa-based lesions containing bland spindle cells with prominent vascularization. (11) In addition to the typical morphologic features associated with IFP, two additional features were identified by Liu et al: a short fascicular growth pattern, in 36% of the cases, and an additional 13% of cases with sparse eosinophils but prominent hyalinization. (4)

Immunohistochemically, IFPs are positive for CD34, smooth muscle actin and CD68 and negative for
CD117, S100 protein and cytokeratin AE1/AE3 [11]. When histopathologically diagnosing IFPs, other spindle cell lesions, such as GISTs, schwannomas, and inflammatory myofibroblastic tumors (IMTs), should be ruled out. These lesions can be usually distinguished using immunohistochemical examination. For example, GISTs are positive for CD117, schwannomas are positive for S100 protein, and IMTs are positive for ALK and negative for CD34, whereas IFPs are not [11, 12-15]. In the present case, the most important differential diagnosis was that of an atypical GIST, because of CD117-negativity.

Genetically, IFPs overlap with GISTs and harbor mutations in the PDGFR-A gene. (4) Immunohistochemistry is used to distinguish between the two. Both tumours are positive for CD34 and vimentin, but GISTs are positive for CD117, while IFPs are not. (16,17)

Epithelial dysplasia and adenocarcinoma have rarely been reported in the associated mucosa overlying IFP, but IFPs can prove to be diagnostically challenging for pathologists. (4)

CONCLUSION

IFPs arise from the submucosa but often extend into the mucosa. A diagnosis of an IFP should be high on the differential diagnosis when there is presence of perivascular onion skinning, short fascicular growth, and abundant eosinophilic infiltration. In case of doubt, a limited panel of immunohistochemical stains (immunoreactivity for CD34, and absence of CD117 and S-100 protein) can be used to establish the correct diagnosis.

REFERENCES


LEGENDS

Figure 1: H&E 100x- Photomicrograph of Iliial polyp showing submucosa composed of edematous stroma containing thin walled blood vessels with onion skin arrangement of spindled cells around vessels.
Figure 2: H&E 400x- Photomicrograph showing edematous stroma with prominent vessels and bland spindle cells along with dense inflammatory infiltrate comprising mainly of eosinophils.

Figure 3: IHC 100x- Photomicrograph showing CD 34 positivity in the spindle cells.