

## Review Article

# Inflammatory fibroid polyp (Vanek's tumour) of the bowel

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### ABSTRACT

Inflammatory fibroid polyp (IFP) or Vanek's tumour is a rare benign tumour of the gastrointestinal tract. Lower gastrointestinal IFP's may present as an acute abdomen in the form of an intussusception in an adult. The surgeon needs to be aware of this entity to avert misdiagnosis. An abdominal CT or an MRI is helpful in diagnosing an acute abdomen due to IFP in an adult. Surgery is the mainstay of treatment. The etiopathology and management of this rare tumour is discussed.

**Keywords:** Inflammatory fibroid polyp, Vanek's tumour

## INTRODUCTION

Inflammatory fibroid poly (IFP) is one of the rare tumours arising from any part of the gut more commonly in the stomach followed by the small intestine. It was first described by Vanek in 1949.

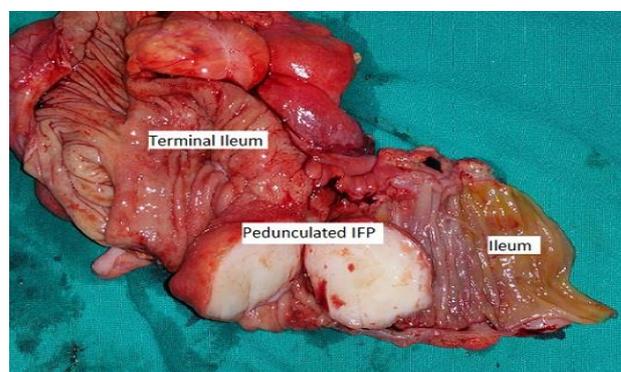
He described the lesion in the stomach as a benign, non-encapsulated, submucosal granuloma composed mainly of loose connective tissue, vessels and abundant eosinophilic inflammatory component.<sup>1</sup> It is also called as eosinophilic granuloma, submucosal fibroma or inflammatory pseudotumor. It is equally seen in both the sexes involving all age groups.

### Site of origin

IFP's are idiopathic, localized, benign neoplastic lesions originating in the submucosa of the gastrointestinal tract. Gastric antrum is the commonest site followed by small bowel, colorectal, gall bladder, oesophagus and duodenum.<sup>2,3</sup> Ileal IFP's deserve special mention as they usually present as intussusception in adults.<sup>3</sup>

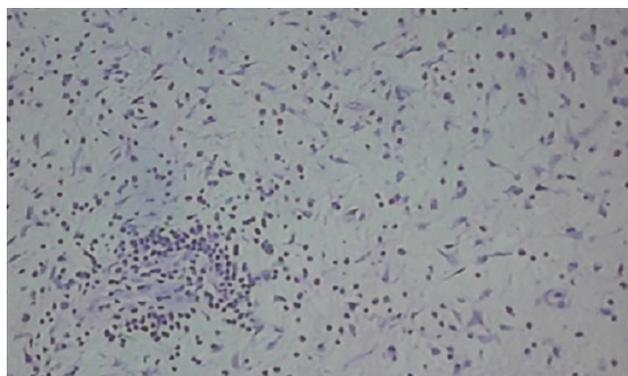
### Pathology

Macroscopically the tumours are either pedunculated or sessile in nature. Majority of them are in the form of pedunculated polyps (Figure 1). Their size may vary from 0.2 cm to 20 cms. They project into the lumen of the bowel. The surface of the tumour is ulcerated and pale.



**Figure 1: Pedunculated IFP arising from the terminal ileum was the apex of the intussusception.**

Microscopically IFP's exhibit peculiar features. Vascular and fibroblastic proliferation accompanied by an inflammatory response which is predominantly eosinophilic in nature is characteristic. Mononuclear spindle shaped cells arranged in whorls around the blood vessels or mucosal glands are typically seen (Figure 2). There is an abundant inflammatory response comprising of eosinophils, lymphocytes, macrophages and mastocytes.<sup>4</sup> However the eosinophil component predominates. The background matrix contains collagen. This was described by Vanek.<sup>1</sup> Based on this component he classified IFP's into two types. The classic type typically seen in the stomach comprised of heavy inflammatory infiltrate or eosinophilic granuloma but sparse amount of collagen. The intestinal type was paucicellular but collagen rich. Fibroblastic spindle cells and inflammatory infiltrates were scant. Recurrence in completely resected tumours is extremely rare.<sup>2-4</sup>



**Figure 2: HP reveals spindle shaped cells in a background of eosinophilic inflammatory infiltrate. (H&E staining, Magnification 4X).**

#### **Genetics and immunohistochemistry**

IFP's are associated with genetic mutations in PDGFRA. The frequency of mutations ranges from 21% to 69%.<sup>5,6</sup>

In the gastric type of IFP, spindle cells are positive for CD 34. However in the intestinal type this is not seen.<sup>7</sup> Immunostaining for KIT, DOG -1, S 100 and EMA are negative. However IFP is strongly positive for vimentin but absent for HAM-56. This phenomenon is suggestive of a major component of spindle cells best recognizable as fibroblasts.<sup>2</sup> Thus IFP's exhibit variable activity for actin, CD 34, desmin, CD117 and S100.<sup>7</sup>

#### **Clinical features**

The site of the tumour determines the mode of presentation.<sup>8</sup> Tumours situated above the level of the ligament of Treitz are usually asymptomatic. These may present with vague symptoms of abdominal discomfort, upper abdominal pain, vomiting and bleeding. Tumours situated below the ligament of Treitz usually present as an acute abdomen. Acute onset colicky abdominal pain, lower GI bleeding and in certain cases a complicated

intussusception. IFP's can cause intussusception in adults just as other benign tumours such as lipomas.<sup>8</sup>

#### **Diagnosis and treatment**

An elaborate history can provide suggestive leads provided one is aware of such pathology. IFP's in the stomach can only be picked up by gastroscopy. This enables both diagnosis as well as therapy. However differentiation from GIST remains a big challenge. This can only be ascertained by detailed immune histochemical studies. Immunohistochemistry of the tumour tissue in GIST exhibits positivity for CD 34, vimentin and CD 117. Whereas gastric IFP exhibits positivity only for CD 117.<sup>9,10</sup> It is the lower gastrointestinal IFP's which pose a diagnostic dilemma. Push and pull enteroscopy or double balloon enteroscopy can aid in the diagnosis. Capsule endoscopy is yet another option but restricted to non-emergency situations only. Colonic lesions can be diagnosed by colonoscopy. Lower gastrointestinal IFP's usually present with lower GI symptoms and many a times with features suggestive of an acute abdomen. Physical examination in cases presenting as acute abdomen by virtue of intussusception may have a palpable lump. An abdominal x ray will reveal air fluid levels only in cases of complicated intussusception. Ultrasound may show a lump or mass. However diagnostic signs may not be picked up by USG as it is performer dependent.<sup>11</sup> An abdominal CT is invaluable for diagnosis. It will help in identifying intussusception. In a transverse view a typical target or doughnut sign is seen. While in a longitudinal view a pseudo kidney, sandwich or hayfork sign may be seen.<sup>12</sup> MRI of the abdomen can also help in diagnosis with atypical bowel in bowel or coiled spring appearance seen. The sensitivity, specificity and accuracy of MRI is higher than CT (Table 1).<sup>12,13</sup> Small bowel intussusception in adults can be caused by a variety of benign causes. Hence the final diagnosis can only be reached by a diagnostic laparoscopy or an open laparotomy followed by histopathological and immune histochemical studies of the specimen.<sup>14</sup>

**Table 1: Diagnostic efficacy of MRI vs CT.**

	MRI	CT
Sensitivity %	95	71
Specificity %	100	71
Accuracy %	96	71

Intraoperative reduction of the intussusception continues to be a contentious issue.<sup>14,15</sup> In cases of exclusive small bowel intussusceptions, intraoperative reduction may be attempted.<sup>13</sup> The advantage of reduction is that the length of the bowel to be resected is reduced.<sup>15,16</sup> However in cases of ileocolic or a colocolic intussusceptions, as the risk of a malignant lesion looms high it is preferable to do en bloc resection of the unreduced specimen based on oncological principles. The same applies to

intussusceptions complicated by ischaemia, necrosis or perforation.<sup>16,17</sup>

## CONCLUSION

Inflammatory fibroid polyp or Vanek's tumour is a rare type of small intestinal tumour. It presents as an acute abdomen in the form of an intussusception in an adult. This poses a diagnostic dilemma. Endoscopy is diagnostic for upper GI IFP's. Whereas abdominal CT or MRI can be diagnostic for lower GI IFP's. Diagnostic laparoscopy or open laparotomy is essential for safe surgical treatment and confirmation of the diagnosis.

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