Adult Intussusception from an Inflammatory Fibroid Polyp: A Case Report And Review Of The Literature
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Introduction
Approximately 95% of all intussusceptions occur in children (1). Adult intussusceptions are rare entities, accounting for 0.003% to 0.02% of all hospital admissions (2,3). Unlike their childhood counterparts, 80-90% of adult intussusceptions are due to demonstrable pathology (1,2,3,4,5). We report a case of adult intussusception due to an inflammatory fibroid polyp (IFP) - a benign mesenchymal lesion of the gastrointestinal tract.

Case Report
A 50 year old man with no known medical illnesses presented to the University Hospital of the West Indies after experiencing colicky abdominal pain, vomiting and abdominal distention that had been worsening over the past three days. There was no history of prior abdominal operations.

Significant examination findings were confined to the abdomen that was distended, with mild peri-umbilical tenderness and hyperactive bowel sounds. Plain abdominal radiographs revealed multiple loops of dilated small bowel in the left upper quadrant of the abdomen.

A diagnosis of mechanical small bowel obstruction was entertained. Exploratory laparotomy revealed an intussusception at the jejunum with a 2cm tumor palpable transmurally as a lead point. The intussusception was resected without prior reduction and intestinal continuity was restored by primary anastamosis. The post-operative course was unremarkable.

Gross pathologic examination revealed a 3.0 x 2.5 x 2.5 cm sessile hemorrhagic polyp within the small bowel. Sectioning of the lesion revealed a homogenous, hemorrhagic, tan, submucosal tumor that did not invade the muscularis propria. The mucosa overlying the tumor was hemorrhagic and ulcerated (Fig. 1).

Figure 1
Figure 1: Whole mount view showing polypoid small bowel lesion covered by ulcerated mucosa
Figure 2
Figure 2: Low power view showing proliferative lesion within submucosa of small bowel

The mass was composed of a proliferation of spindle-shaped and stellate fibroblasts embedded within loose, oedematous stroma, which contained a dense chronic inflammatory cell infiltrate composed predominantly of eosinophils (Fig. 3). Numerous small blood vessels were scattered throughout the stroma (Fig. 3) and larger vessels with distinctive zones of loose connective tissue around them were also present.

Figure 3
Figure 3: High power view of submucosal mass showing proliferation of fibroblasts within oedematous stroma. Blood vessels and eosinophils are evident.

The overall features were consistent with IFP. Immunohistochemistry revealed positive staining for vimentin and absence of staining for CD34, CD117 (c-kit), S-100 and Cathepsin-D. Six lymph nodes were present in the attached mesentery, all with reactive changes only.

DISCUSSION
Although adult intussusceptions are uncommon, they are reported to account for approximately 1% of all admissions for bowel obstruction in adults (s). Preliminary regional data from Trinidad suggests that there is a slightly higher prevalence in the Caribbean, where 11% of intussusceptions occurred in adults, accounting for 4% of all admissions for bowel obstruction (s).

The jejunum is the least likely location for intussusceptions to occur because mass lesions are uncommon at this part of the gastrointestinal tract (s). A jejuno-jejunal intussusception resulting from an IFP was first described by Winker et al in 1986 (s). Since that time, there have been a handful of case reports that document the frequent association between enteric intussusceptions and IFPs (s).

Inflammatory fibroid polyps can be found anywhere within the gastrointestinal tract, but they are most prominent within the gastric antrum and distal ileum (s). The lesion was first described in 1949 by Vanek who referred to the lesion as a “submucosal granuloma with eosinophilic infiltration” (s). The now commonly accepted nomenclature, IFP, was coined by Ranier and Helwig in 1953 (s).

IFPs have been reported to occur across a wide age range, but are most commonly identified in the sixth decade of life (s). This is likely because the lesions produce local symptoms that are size dependent.

The majority of cases are sporadic, although there are isolated reports of IFP occurring in familial clusters (s) and as polyposis syndromes (s). There is also a noted association between IFPs and coexistent gastric neoplasms (4 carcinomas and 2 adenomas) (s).

The aetiology of IFPs is unclear, and it is controversial whether they are true neoplasms; many authors believe that they are not (s). One theory suggests that they occur as a chronic inflammatory reaction in response to local noxious stimuli, with incomplete differentiation of myofibroblasts and primitive submucosal stromal cells (s). Others have suggested that the heavy eosinophilic infiltrate is in keeping with an uncontrolled allergic response (s) or that the occasional smooth muscle bundles may represent degenerating leiomyomata (s). Neither of these theories has been widely accepted as the aetiology of IFPs (s).

Macroscopically, IFPs tend to appear as solitary sessile
polyps that are well circumscribed. Larger lesions may become pedunculated (s31), and the overlying mucosa may be ulcerated (s118-20). The polyps are usually confined to the submucosa, but may extend into the mucosa, or into the muscularis propria and serosa (s3). The classic microscopic features are as described in the index case. That is, a proliferation of fibroblasts within loose, oedematous connective tissue stroma, accompanied by infiltrates of eosinophils, plasma cells, lymphocytes, macrophages and mast cells (s14-15). Several variably sized blood vessels are usually present within the stroma, and larger vessels are often surrounded by a distinct zone of loose connective tissue (s34-35).

The gastrointestinal stromal tumour (GIST) is an important consideration in the histological differential diagnosis of IFPs, and in morphologically ambiguous cases, immunohistochemistry is used to make a distinction between the two. Both tumours are positive for CD34 and vimentin, but GISTs are positive for CD117 (c-kit), while IFPs are not (s34-35). Other histological differential diagnoses include neurogenic tumours and desmoid tumours. Neurogenic tumours are positive for S-100, while IFPs are negative for this marker, and desmoid tumours can be distinguished by their origin outside of, rather than within, the bowel wall (s34-35), as well as positive staining for cathepsin-D. In addition to positivity for CD34 and vimentin, some IFPs may also be positive for smooth muscle actin, calponin, CD35 and cyclin-D1 (s35). Some IFPs, like ours, fail to show positive staining for CD34, and this may be related to the state of evolution of the tumour (s35). It has also been suggested that the entity IFP may encompass two different tumours (s35), accounting for the absence of CD34 staining in some.

IFPs have no metastatic potential. They remain dormant until they are large enough to produce local symptoms that are dependent on their location. Small bowel lesions are not usually diagnosed pre-operatively because they present with vague symptoms of bowel obstruction due to intussusception. Laboratory investigations and plain radiographs are not helpful in making the diagnosis as they will demonstrate non-specific findings that are more in keeping with bowel obstruction.

The current trends of liberal diagnostic imaging have increased the pre-operative detection of adult intussusceptions. The “target” or “sausage” sign has been described on ultrasound and computed tomography (CT) scans, representing the central intussusceptum surrounded by concentric layers formed by the thickened oedematous intussusceptum (s39-40). Collective analysis of three retrospective studies that examined CT findings in over 70 patients with surgically confirmed intussusceptions has shown that abdomino-pelvic CT has 78% sensitivity and 100% specificity to diagnose intussusceptions pre-operatively (s38-41).

There are no unique identifying features on imaging that can distinguish the nature of the lead points. The absence of pathognomonic clinical or radiological features makes operative resection and microscopic assessment mandatory.

Operative resection of the involved intestine with macroscopically clear margins is sufficient treatment for IFPs. It is important to ensure uninvolved resection margins to ensure complete excision of the affected bowel segments. Inadequate resection margins may leave involved bowel and predispose to disease recurrence. To date, there have been two reported cases in the literature where IFPs recurred after presumably incomplete resection (s16-42).

CONCLUSIONS

Inflammatory fibroid polyps may commonly present as enteric intussusceptions in adults. This diagnosis should, therefore, be borne in mind when intussusceptions are encountered in adult patients. They can be treated adequately by resection of the involved bowel segments with macroscopically clear margins.

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References

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