A rare case of multifocal inflammatory pseudotumor of the ileum with regional lymph node involvement

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Citation

Abstract
Inflammatory pseudotumor involving ileum is a rare pathological entity and multiple involvements, including a lymph node, are a still rarer condition. We report such a rare case of multifocal inflammatory pseudotumor of the ileum presenting as small intestinal obstruction in a young adult male.

INTRODUCTION
Subacute obstruction of the small intestine has various etiologies. Inflammatory pseudotumor, as one of its causes, is a rare event that primarily affects the ileum. Transmural multifocal involvement of ileum with solitary regional lymph node involvement is still rarer. This account is of such an unusual case at our institute.

CASE HISTORY
A 35-year-old Hindu male priest presented with pain around the umbilicus, mild progressive abdominal distension, constipation and vomiting for 7-8 days. There was a history of loss of appetite for 15 days. The patient was non-alcoholic and had no past history of any major medical illnesses or any trauma to the abdomen. The patient took treatment from a private hospital for one week and was diagnosed of having a possibility of ileo-caecal tuberculosis or malignancy.

On examination, the patient was pale with vital parameters within normal limits. On per abdominal examination, there was mild distension associated with guarding and tenderness in the right iliac fossa and around the umbilicus. On per rectal examination, there was minimum tenderness but no ballooning and there were grade 2 piles at the 7 o’clock position without any active bleeding on proctoscopy.

The patient’s haemoglobin was 8.1g%, WBC count 9,200/cu.mm and ESR 70. RBC morphology revealed hypochromic, microcytic, anisocytic anemia. Stool routine showed 6-8 pus cells/hpf and mucus plugs with no RBCs. His HIV status was negative. The plain radiograph of the chest was normal. Ultrasonography of the abdomen revealed a bowel mass in the right iliac fossa.

Barium meal follow-through showed markedly delayed passage of barium through jejunum and ileum with grossly dilated proximal ileum. Terminal ileum and caecum were not seen. Delayed fluoroscopy showed retained dye in the ileum.

Preoperatively, the patient was given two units of blood to increase his haemoglobin and a decision for an exploratory laparotomy was made.

The patient was explored with right lower paramedian incision under spinal anesthesia with the following operative findings.

Tight stricture and induration with mass formation 1½ feet from the ileo-caecal junction.

There was another nodule and induration about 5cm distal to the above stricture.

A 3rd stricture with nodule was present about 10cm from the ileo-caecal junction which was not passable.

The proximal ileum was grossly dilated.

There was one large mesenteric non-contagious lymph node present at the root of the mesentery.

The rest of the viscera were normal without any evidence of other lymph nodes.

There was no free fluid in the abdomen.

Ileo-ileal resection with anastomosis in two layers was done.
The post-operative course was uneventful. Sutures were removed on the 7th post-operative day. At follow-up after more than four years, the patient is asymptomatic.

At pathology, on gross examination, the cut section showed the first polyp distal to the 1st stricture mass. There were 2 aphthous ulcers seen 5cm apart on the anti-mesenteric border, longitudinally over the bowel mucosa.

**Figure 1**
Fig. 1: Resected specimen

**Figure 2**
Fig. 2: Large nodule opened.

**Figure 3**
Fig. 3: Cut open small nodule.

**Figure 4**
Fig. 4: Two aphthous ulcers.

Microscopy showed a dense inflammatory infiltrate consisting of lymphocytes, plasma cells and eosinophils and transmural inflammation essentially extending from submucosa through muscle into serosa. There was no evidence of tuberculosis, malignancy, Crohn’s disease of the ileum or lymphoma. So the final diagnosis of a multifocal inflammatory pseudotumor of the ileum with involvement of a non-contagious lymph node was made.

**DISCUSSION**
Subacute intestinal obstruction has many etiologial factors. Commonest in India is small intestinal stricture involving ileum or the ileo-caecal region due to tuberculosis. Next are tuberculous adhesions, post-exploratory adhesions, immunocompromised states etc. In more than 50% of
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patients, mere good conservative treatment will be sufficient.

Presentation of an inflammatory pseudotumor is acute or insidious. Acute presentation manifests as signs of acute intestinal obstruction or intussusception. The insidious form presents as in this particular patient, with subacute intestinal obstruction.

A neoplastic nature of an inflammatory pseudotumor is debatable. It occurs rarely in the small intestine and normally there is a solitary or polypoidal lesion commonly in the mucosal and muscularis layer of the ileum and not involving serosa. Multifocal lesions with regional non-contiguous lymph node involvement have rich vascular stroma with plenty of eosinophils, lymphocytes and plasma cells without any granuloma formation.

Inflammatory pseudotumor, being a benign neoplasm, does not require radical treatment. Treatment recommendations include resection of the involved part and restoration of continuity. Post-operative chemotherapy is not needed. Few advocate localized radiotherapy in the lymph node area. Regular follow-up is a must and prognosis is excellent.

The most likely differential diagnosis could be Crohn’s disease of the small intestine and eosinophilic gastroenteritis showing pathological features of transmural involvement, skip lesions and intervening normal mucosa.

Our case shows a spectrum of lesions with two aphthoid ulcers on the anti-mesenteric border, stricture formation and polyp. This spectrum of lesions has not been described in any of the cases in the literature.

Recent molecular study shows a neoplastic nature of this condition; however, such a multifocal disease as seen in the presented case would support interrelationship with the localized form of eosinophilic gastroenteritis that takes the form of localized polypoidal disease. We prefer to retain the term “inflammatory pseudotumor” as the patient did not have any allergic history and there was no peripheral blood eosinophilia. The origin remains obscure.

Our case is a classic example which shows overlapping features of two entities. This lesion probably evolves from an ulcer and develops into a pseudotumor with the basic pathology remaining same.

References
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