



Eosinophilic Enteritis Presenting As Intestinal Obstruction – Report of Two Cases

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ABSTRACT

Eosinophilic gastroenteritis (EG) is an unusual disorder. It is characterized by eosinophil infiltration of the gut wall histologically and is manifested by gastrointestinal (GI) symptoms clinically. This disease entity preferentially affects the stomach and proximal small intestine. Transmural variety is a rare form of this disease causing stricture and intestinal obstruction.

We present two cases of Eosinophilic enteritis causing distal small intestinal stricture and proximal colonic mass both presenting as acute intestinal obstruction.

INTRODUCTION

Eosinophilic enteritis is an uncommon disease characterized by eosinophilic infiltration involving any layer of the bowel wall ^[1]. It can affect any area of gastrointestinal tract from oesophagus to rectum although stomach and proximal small intestine especially duodenum are sites most frequently involved. It is a spectrum of gastrointestinal (GI) disorders characterized by inflammation rich in eosinophils without evidence

of other known causes of eosinophilia (i.e. parasitic, infectious, drug reaction, or malignancy) ^{[1][2]}.

In 1937, Kaijser first described the disease in two patients with syphilis who were allergic to neoarsphenamine ^[1]. It is an extremely rare disease, with less than 200 patients described in the literature. The largest published series includes only 40 patients. We report two cases of eosinophilic enteritis which presented to us with

acute intestinal obstruction. We present two cases of Eosinophilic Gastro Enteritis with transmural distal small intestinal stricture and proximal colonic mass both presenting as acute intestinal obstruction. Both the cases presented to us were acute abdominal emergencies and hence the choice of management was surgery.

CASE REPORTS

CASE 1

A 55Y/Male Pt came with the c/o Right lower abdominal pain -1 week, Vomiting - 4 days, Fever - 3days. He is not a known DM/HT/TB. No history of asthma or any other allergic disorder was present. On Examination patient was dehydrated with a pulse of 120/min. Abdomen Examination :Tenderness ++ in RIF & Hypogastrium, Guarding+++ in RIF, Distension++, No organomegaly, No ascites. Other system examinations were normal .P/R: Normal. Provisional diagnosis of Appendicular perforation with localised peritonitis was made. Blood investigations revealed moderate leukocytosis (TC 10,000) and eosinophilia of 10%. Laparotomy findings were Firm mass involving the whole caecum and causing stenosis of the lumen with multiple nodes in the mesocolon Appendix was normal and rest of the bowel was normal. Considering the patient's age with intra operative findings of hard mass and multiple nodes, malignancy was suspected and the Procedure done was Rt Hemicolectomy with ileo-transverse anastomosis.

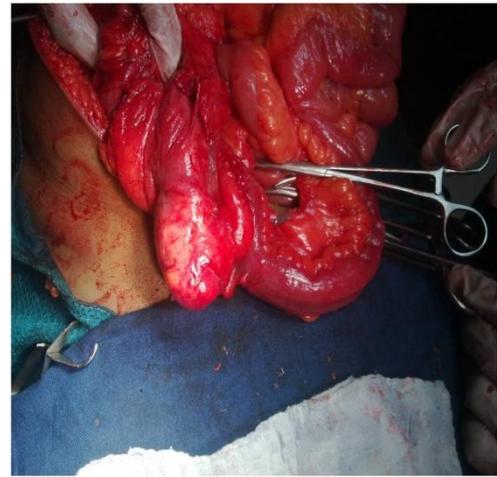


Figure 1: Showing Caecal mass lesion

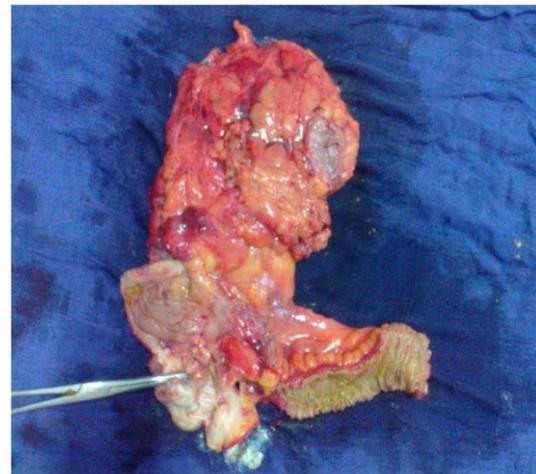


Figure 2: Cut section showing Luminal narrowing

Postoperative period: Uneventful. Post operative HPE revealed eosinophilic enteritis. Patient followed up till date and there are no complaints.

CASE 2

50 year old patient came with complaint of abdomen pain 5 days intermittent colicky type, abdomen distension past 2 days, vomiting 2 days, constipation and obstipation 1 day. He is a Known asthmatic, not a known DM/HT/TB. On Examination patient was dehydrated, tachycardia was present. Abdomen examination revealed distended abdomen, tense abdomen and with guarding in right iliac fossa and right lumbar

regions. Ultrasonogram of abdomen revealed distended bowel loops occupying the whole abdomen, other viscera namely liver spleen and kidney were normal.



Figure 3: Dilated bowel loops with multiple air fluid level

CT of the abdomen revealed distended bowel loops with cut off at the terminal ileum. Due to increasing abdomen distension and abdominal pain patient was taken up for laparotomy. Intra operatively there was a stricture in the terminal ileum about 25cm from ileocaecal junction causing luminal obstruction. The strictured part was removed and end to end ileo-ileal anastomosis was done.



Figure 4: Cut section showing stricture segment of the ileum

Post op period was uneventful and patient is on regular follow up with no complaints at present. Post operative HPE revealed eosinophilic enteritis. In both cases there was histologically, dense infiltration of eosinophils throughout the entire thickness of bowel wall (Trans mural)

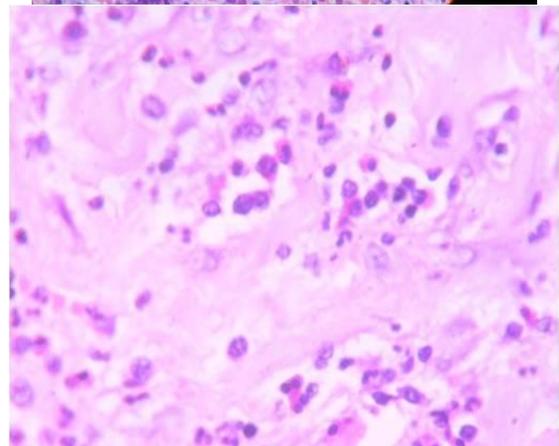
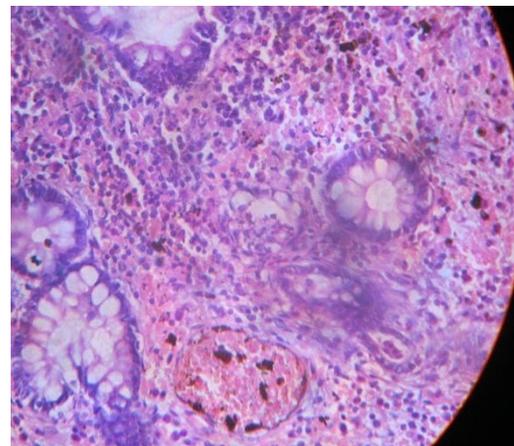


Figure 5: HPE - showing dense eosinophilic infiltrates

DISCUSSION

Primary eosinophilic gastroenteritis (EG) is defined as a disorder that primarily affects the gastrointestinal tract with eosinophil rich inflammation in the absence of a known cause for eosinophilia, including drug reaction, parasitic, infections and malignancy ^{[1][4]}. Eosinophilic intestinal inflammation can occur secondarily in the GI tract in inflammatory bowel disease,

autoimmune diseases, reaction to medications, infections, hypereosinophilic syndrome and after solid organ transplantation.

A study at Mayo Clinic showed that 50% of patients with Eosinophilic gastrointestinal disorders have a history of allergy such as asthma, allergic rhinitis, urticaria, drug allergy and eczema [7].

EPIDEMIOLOGY

Eosinophilic enteritis is a rare disease with less than 200 cases since it was first described by Kaijser in 1937. The etiology and pathogenesis was shaped in large part by case reports and series over the years.

EG is predominantly a male disorder that affects children as well as adults. The incidence of EG peaks between the age of 30 to 50 years [5].

ETIOPATHOLOGY

The exact etiology of EG remains unknown, although it is now recognized as a result of both IgE and non-IgE mediated sensitivity. Interleukin (IL)-5 is a chemoattractant responsible for tissue eosinophilia [9][10].

Desreumaux, et al found that among patients with EG, the levels of IL-3, IL-5, and granulocyte macrophage colony stimulating factor (GM-CSF) were significantly increased as compared to control patients. Once recruited to the tissue, eosinophils may further recruit similar cells through their own production of IL-3 and IL-5 [9].

CLINICAL FEATURES

The majority of patients with eosinophilic gastroenteritis are adults' age 20 to 50 years who have nonspecific gastrointestinal symptoms correlating with a classification scheme based on [8]

1. The depth of infiltration by the eosinophils (mucosa, serosal or transmural)
2. The pattern of involvement (diffuse, localized), and
3. The location of the disease (oesophagus, stomach or bowel).

The stomach is the most frequent site of involvement, followed by the small intestine, the duodenum, and (rarely) the colon.

Diffuse mucosal infiltration (MUCOSAL FORM) by eosinophils is the most common pattern, resulting in symptoms of abdominal pain, nausea and vomiting, and diarrhea, malabsorption syndrome, anemia, bleeding, protein-losing enteropathy, esophagitis.

Eosinophilic gastroenteritis (TRANS MURAL FORM) involving the muscle layer often involves the gastric antrum, and affected patients can present with pyloric or small-bowel obstruction [3][11].

The serosal involvement (SEROSAL FORM) occurs only in a minority of the patients and is characterized by exudative ascites with higher peripheral eosinophilic counts as compared to other forms.

Most cases in transmural form present as acute abdominal emergencies as they can be dismissed as non specific abdominal pain in initial stages. In some cases it can be picked up on incidental

radiographical examination as thickened loop of bowel in which cases we need to confirm it with laparoendoscopic full thickness biopsy ^{[3][10]}.

Pre operative diagnosis of cases presenting as intestinal obstruction is difficult as more common ulcero-constrictive lesions like tuberculosis and malignancy are suspected.

Most reported cases of muscular disease were diagnosed during surgical resection of an intestinal obstruction or suspected malignancy as was in our case.

In first case stricture in terminal ileum is rare as reported cases of stricture formation is in proximal small intestine.

Isolated colonic involvement is a rare presentation of EG. Our review of the literature shows that colitis is the most common clinical presentation of colonic EG (54%), followed by a colon mass or tumor (31%), and intestinal obstruction (23%).
^{[10][5][6]}

INVESTIGATIONS AND EVALUATION

Talley *et al* have identified three main diagnostic criteria ^[9]

1. non-specific gastrointestinal symptoms;
2. eosinophilic infiltration of one or more areas of the GIT
3. exclusion of other causes for the intestinal eosinophilia.

The absolute requirement of establishing a diagnosing of eosinophilic enteritis is demonstration of tissue eosinophilia.

LABORATORY INVESTIGATIONS

Laboratory study often reveals peripheral eosinophilia in EG patients (67% - 100%). This is not a universal finding and thus does not constitute a prerequisite for diagnosis ^[2].

Between 60 - 70% of adult EG patients have elevated total IgE levels. Iron deficiency anemia and positive occult blood in the stool are also commonly found, probably due to mucosal involvement with GI blood loss.

Stool analysis is an important part of the laboratory investigation, as exclusion of parasitic infestation is needed for diagnosis. In our patient eosinophilia was present in the first case but not in second. In Both cases there was absence of any ova or parasites on repeated stool examinations.

ENDOSCOPY AND IMAGING

Endoscopic findings vary widely. Abnormal endoscopic findings include prominent mucosal folds, hyperemia, ulcerations, and nodularity. In EG that involves the terminal ileum or colon, aphthoid ulcers may also be seen most commonly at the cecum and ascending colon. To establish a diagnosis of EG, endoscopic biopsy is often done. Because EG involvement is usually patchy in character, multiple biopsies from both the normal and abnormal mucosa have to be done.

CT scan may show nodular and irregular thickening of the gut which may mimic other conditions like Crohns disease, tuberculosis or lymphoma.

Despite all the clinical features, definitive diagnosis can only be made based on presence of increased eosinophils in biopsy specimens from

the GI tract wall, the lack of involvement of other organs, and the exclusion of other causes of eosinophilia ^{[4][7]}.

The histological characteristic is edema and an inflammatory cell infiltrate composed of eosinophils, which may appear in clumps.

MANAGEMENT

Acute obstructive presentation is dealt with laparotomy and segmental resection anastomosis as in our case.

With advent of rampant laparoscopic practice there are reports describing laparoscopic biopsy in suspicious cases. Diagnostic laparoscopy may show a thickened intestine with or without mesenteric adenopathies.

Intestinal biopsy can be taken as described by Edelman, in patients with abdominal pain in whom exists the clinical suspicion of this disease based on a history of atopy, increased eosinophil count in peripheral blood, and suggestive ultrasound findings ^{[5][7]}.

ROLE OF STEROIDS

Steroids have a role in the management of patients with non-allergic eosinophilic enteritis ^{[7][10]}. Serosal disease and mucosal disease responds dramatically to steroids. They have produced symptomatic improvement in controlling diarrhoea and protein losing enteropathy and preventing recurrence of ascites.

For patients who underwent surgery for acute event, adjunctive use of steroids as part of the treatment regimen decreased the recurrence rate. Both our patients were treated with steroids for 6

weeks in post op period and continue to remain well for follow up period of 1 year and 6 months respectively.

Mortality related to eosinophilic gastroenteritis has not been reported. However, progressive weight loss leading to profound cachexia unresponsive to treatment can occur. Eosinophilic gastroenteritis does not predispose a patient for GI malignancy.

The differential diagnosis for patients with similar presenting complaints would include:^[4]

1. Parasitic infections (which could also elevate the peripheral blood eosinophil count)
2. Inflammatory bowel disease (alteration of bowel habits and produce both small and large bowel wall thickening on CT scan)
3. Idiopathic HES is a condition of sustained hypereosinophilia that affects multiple organs including the lung, skin, blood vessels, and nervous system.
4. Lastly, a variety of medications and malignancies can produce eosinophilia and abdominal pain as side effects.

All these conditions were ruled out in our patients through detailed history taking and laboratory investigations.

CONCLUSION

Eosinophilic enteritis continues to be diagnostic dilemma as clinical presentations with investigations are only contributory.

Eosinophilic enteritis should be considered in the differential diagnosis of patients with unexplained acute or recurrent abdominal pain and peripheral

hypereosinophilia Patients with mild and sporadic symptoms can usually be managed with assurance and observation. Surgical intervention might be necessary in patients with obstructive complications or refractory disease.

Both of our patients experienced relief from symptoms after resection. The cases have been presented because of the rarity of occurrence and presentation and to acquaint the surgeon community with the condition.

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