EOSINOPHILIC GASTROENTERITIS - A RARE CAUSE OF INTESTINAL OBSTRUCTION

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Abstract: Intestinal obstruction is one of the most common gastrointestinal emergencies in children attending pediatric gastroenterology clinic. Among the many causes that cause intestinal obstruction in children, one rare but a distinct clinical entity is eosinophilic enteritis which responds dramatically to steroids. We report 2 rare cases of Eosinophilic enteritis presenting as intestinal obstruction in children. The first case was a seven year old female who came with complaints of acute onset of vomiting, abdominal pain with abdominal distention, underwent laparotomy and resection on suspicion of intestinal obstruction, didn't improve and was referred to GE dept. On examination, she had generalised distention of abdomen with tenderness and normal bowel sounds. Routine investigations were done which were normal. Serum IgE levels were 444 IU. Child was subjected to emergency re-laparotomy which revealed anastomotic stricture in distal ileum with dilated proximal bowel. Resection of the diseased ileum and end to end ileoileal anastomosis was done and specimen sent for histopathology which revealed inflammatory infiltrate with eosinophil more than 20HPF in mucosa and submucosa extending to serosa. Findings which were consistent with eosinophilic gastroenteritis. The child was started on oral steroids which were tapered over six months. During regular follow up she was found to be asymptomatic. The second case was that of a twelve year old male child who came with complaints of vomiting, abdominal pain and distention for one week. General examination revealed mild pallor with generalized distended tender abdomen with dilated loops. Routine investigations revealed eosinophilia. Serum IgE level was 923 IU. He was subjected to emergency laparotomy which revealed dilated jejunum and ileum with two ileal strictures at 15 and 20 cms from ileocaecal junction. Strictures were resected and ileoileal anastomosis was done and the resected segment was sent for histopathology which revealed eosinophils extending from mucosa to serosa. Findings which were consistent with eosinophilic gastroenteritis.
Post operative oral steroids were started and gradually tapered. He is on regular follow up and currently asymptomatic.

**Keyword**: Eosinophilia, Intestinal Obstruction, Serum IgE, Oral steroids

**Introduction**

Eosinophilic enteritis is a rare disease that is characterized by tissue eosinophilia. The clinical signs and symptoms depend on the layer of the gut predominantly involved. The most prevalent form is the one with predominant involvement of the mucosal layer, but sometimes it can involve the muscular layer presenting with symptoms of gastrointestinal obstruction mimicking pyloric stenosis or gastric outlet syndrome. The exact pathogenesis of the disease remains unknown but, at least in some of these patients, high serum IgE levels have been documented. Most patients will have history of food allergy or family history of allergy.

**CASE I**

7 year old female child born to 3 degree consanguineous marriage came with complaint suggestive of intestinal obstruction. There was no history of food sensitization, allergic diathesis, asthma, parasitic infestations, chronic disease like tuberculosis or any abdominal surgery. She was admitted in May 2010 with above complaints in a private hospital where a clinical diagnosis of sub-acute intestinal obstruction was made. She underwent emergency laparotomy with resection and anastomosis of parts of bowel, details of which are not known. Since the procedure did not relieve the symptoms, she was referred to our institute.

**ON EXAMINATION:**

Child was pale and undernourished with grade II PEM. On examination she had no jaundice, pedal oedema, or lymphadenopathy. Examination of abdomen revealed generalized distension with tenderness and normal bowel sounds. Surgical scar was present. Per rectal examination was unremarkable. Investigations revealed hemoglobin of 7.4g/l, total WBC count of 8600/mm3 with 43% neutrophils, 55% lymphocytes and 2% eosinophils. Peripheral smear revealed microcytic hypochromic anaemia. Serum IgE level was 444 IU. Electrophoretic pattern did not reveal significant hypergammaglobulinemia hence ANA/dsDNA not done. Three consecutive motion examinations did not reveal any parasitic infestation. Her renal function, serum electrolytes were normal. Ultrasonography of abdomen revealed hepatomegaly, echogenic kidneys with ascites and right sided pleural effusion with thickened small bowel. Child was taken for re-laparotomy. Per operative findings revealed an anastomotic stricture in distal ileum with dilated proximal bowel. Resection of the diseased ileum and end ileoileal anastomosis was done and specimen was sent for histopathology. Histopathology showed inflammatory infiltrate with eosinophils >20/HPF in the mucosa and submucosa, extending up to serosa with focal congestion and fibrosis consistent with eosinophilic gastroenteritis. Oral steroids were started post operatively at 1mg/kg for 12 weeks and gradually tapered and maintained on low dose steroids for 6 months. She was followed up for one year and is asymptomatic till date.

**CASE II**

A 12 year old male child first born to non-consanguineous marriage was admitted in our hospital with complaints of vomiting, colicky abdominal pain and abdominal distension of one week duration. There was no past history of similar illness, food sensitization, allergic disease, asthma,
parasitic infestations, chronic disease like tuberculosis or any abdominal surgery. There was no family history of allergy or asthma. Clinical examination revealed pallor with no icterus, pedal oedema or lymphadenopathy. Per abdomen examination revealed generalized distension with tenderness with dilated bowel loops and increased bowel movements. Per rectal examination revealed fecal pellets. Blood investigations revealed hemoglobin of 8.1 g/l, total WBC count of 11500/mm$^3$ with 45% neutrophils, 36% lymphocytes and 19% eosinophils. Peripheral smear revealed microcytic hypochromic anaemia. Electrophoretic pattern did not reveal significant hypergammaglobulinemia hence ANA/dsDNA not done. Three consecutive motion examinations did not reveal parasitic infestation. Serum IgE level was 923 IU/m. The renal function, serum electrolytes were within normal limits. Plain abdominal radiograph revealed dilated bowel loops. The boy was subjected to emergency laparotomy and intra operative findings included dilated jejunum and ileum and two ileal strictures at 15 and 20cms from ileocaecal junction. The strictures were resected and ileo-ileal anastomosis was done. The resected segments were sent for histopathology which revealed focal area of ulceration, submucosal oedema with ganglion cells in muscular and submucosal layers. Eosinophils were seen extending from mucosa to serosa. Findings were consistent with eosinophilic gastroenteritis. Post operative oral steroids 1mg/kg was started and gradually tapered. He is on regular follow up and currently asymptomatic.

**DISCUSSION**

Eosinophilic gastroenteritis (EG) is a rare and heterogeneous condition characterized by patchy or diffuse eosinophilic infiltration of gastrointestinal (GI) tissue, first described by Kaijser in 1937(1,2) A history of atopy or food allergies is often present.(1). Eosinophilic gastroenteritis is characterized by the following: a) Presence of abnormal GI symptoms, most often abdominal pain. b) Eosinophilic infiltration in one or more areas of the GI tract, defined as 20 or more eosinophils per high-power field, the absence of an identifiable cause of eosinophilia and the exclusion of eosinophilic involvement in organs other than the GI tract. Eosinophilic gastritis, enteritis, or gastroenteritis are diseases characterized by the selective infiltration of eosinophils in the stomach, small intestine, or both respectively. The mucosal form is characterized by vomiting, dyspepsia, abdominal pain, diarrhea, blood loss in the stools, iron deficiency anemia, malabsorption, protein-losing enteropathy, and failure to thrive. The muscularis form, characterized by infiltration of eosinophils predominantly in the muscularis layer, may present with gastrointestinal obstructive symptoms mimicking pyloric stenosis or gastric outlet syndrome. The serosal form, which is less common, presents with significant bloating, exudative ascites, and higher peripheral eosinophil counts. The disorders are classified into primary and secondary subtypes. The primary subtypes, which have also been called idiopathic or allergic GE, include the atopic, nonatopic, and familial subtypes. The secondary subtypes may be divided into 2 groups: systemic eosinophilic disorders (ie, hypereosinophilic disorders) and noneosinophilic disorders (eg, celiac disease inflammatory bowel disease, vasculitis). (1,2,3). The exact pathology in predisposing to the clinical manifestation of eosinophilic gastroenteritis is unknown. Although these diseases
are idiopathic, recent investigations support the role of eosinophils, T helper 2 (Th2) cytokines (interleukin [IL]-3, IL-4, IL-5, and IL-13), and eotaxin as the critical factors in the pathogenesis of eosinophilic gastroenteritis. Eosinophils function as antigen presenting cells as they express major histocompatibility complex (MHC) class II molecules and mediator of proinflammatory effects as well as up-regulation of adhesion systems, modulation of cell trafficking, and cellular activation states by releasing cytokines, chemokines, lipid mediators (platelet activating factor [PAF] and leukotriene C4). Tissue damage and cellular dysfunction occur by released toxic granule proteins (major basic protein [MBP], eosinophil cationic protein [ECP], eosinophil peroxidase [EPO], and eosinophil-derived neurotoxin [EDN]) and lipid mediators, which are cytotoxic.(4,5) A subset of patients may have atopy, non IgE mediated food allergy etc in 50% of children.

1. Peripheral blood eosinophilia is found in 20-80% of cases(1,2,4) with average count of 2000 eosinophils (eos) /µL in patients with mucosal layer involvement, 1000 eos/µL in patients with muscular layer involvement, and 8000 eos/µL in patients with serosal involvement. Iron-deficiency anemia may occur. Hypoalbuminemia occur especially in patients with mucosal layer involvement. One third of patients can have mild-to-moderate steatorrhea. Skin prick tests to inhalant allergens and food, help identify sensitization to specific allergens. Histopathology shows tissue infiltrations with eosinophils crypt hyperplasia, epithelial cell necrosis, and villous atrophy. Diffuse enteritis with complete loss of villi, submucosal edema, infiltration of the GI wall, and fibrosis may be apparent. Mast cell infiltrates and hyperplastic mesenteric lymph nodes infiltrated with eosinophils may be present.

2. In absence of other causes of tissue eosinophilia is highly diagnostic of this condition. Radiographically, eosinophilic gastroenteritis does not have a pathognomonic appearance and the findings are variable, nonspecific, and/or absent in at least 40% of patients. Gastric folds can be enlarged, with or without nodular filling defects. Valvular conniventes may be thickened and flattened. Strictures, ulceration, or polypoid lesions may occur. The small intestine also may be dilated, with an increase in the thickness of the folds. Rarely, diffuse esophageal narrowing or achalasia like motor abnormalities may occur. Ultrasound and CT scans may show thickened bowel walls and sometimes, local- ized lymphadenopathy. In patients with serosal layer involvement ascitic fluid is usually detectable. Exploratory laparotomy may be indicated, especially in patients with serosal eosinophilic gastroenteritis and muscular involvement. Endoscopy may be normal or show erythema, nodularity and ulceration.

3. Oral glucocorticosteroids, is indicated for those with obstructive symptom with or without diet elimination therapy especially in children with history of food intolerance or allergy. Most patients respond to conservative measures and oral glucocorticosteroids. Some of the drugs like montelukast, ketotifen, cromoglycin, and mycophenolate mofetil have been tried in some of the cases. (6,7) Surgery is rarely indicated and may be done to relieve persistent pyloric or small bowel obstruction. Recurrence is possible, even after surgical excision.
References


