A Challenging Case of Eosinophilic Esophagitis

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Background

- Eosinophilic gastroenteritis (EGE) is an uncommon infiltrative eosinophilic disease.
- Presentation of EGE is non-specific making the diagnosis challenging.

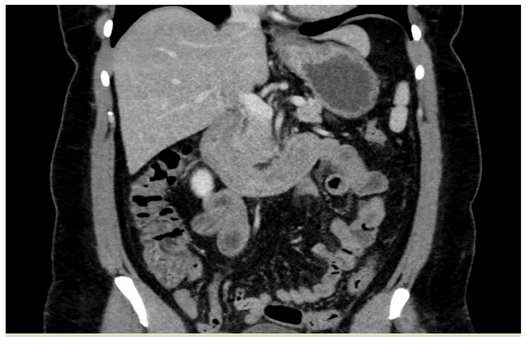
Learning Objectives

- To understand presentation, diagnosis, disease course and differentials of EGE.

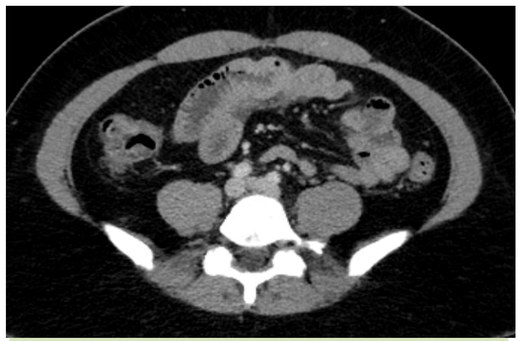
Case Presentation

- A 43-year-old woman with history of hypertension and cholecystectomy presented with chronic worsening epigastric pain associated with postprandial nausea and vomiting.
- Onset was 15 years ago, lasting for few weeks and resolving spontaneously.
- Medications consisted of lisinopril and felodipine.
- On physical exam, the patient had epigastric tenderness.
- Labs were remarkable for:
  - WBC 8.4 ($x10^9$/L)
  - High eosinophil count of 3.21 ($x10^9$/L)
  - High IgE of 320 IU/mL
- Renal panel, hepatic panel, peripheral blood smear, HIV serology, B12 and tryptase levels, stool ova and parasites were unrevealing.
- CT Abdomen with contrast: Mucosal thickening of the stomach and small bowel.
- EGD: Erythema and nodularity of the gastric and duodenal mucosa.
- Gastric biopsies: 40 eosinophils per high per field (Eo/HPF); Duodenal biopsies showed up to 100 Eo/HPF
- Colonoscopy and colon biopsies were unremarkable.

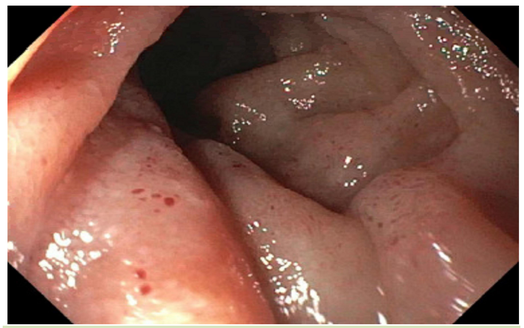
Clinical Images



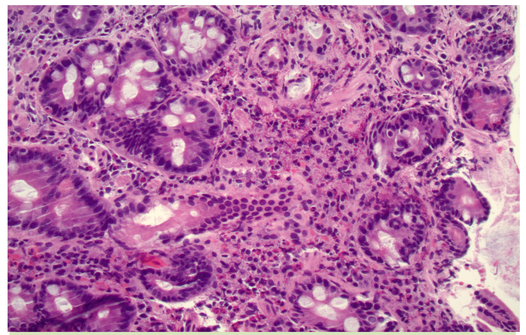
CT Abdomen: Mucosal thickening of the stomach and small bowel



CT Abdomen: "Saw-tooth" appearance of the intestinal mucosa



EGD: Erythema and nodularity of the gastric and duodenal mucosa



EGD: Eosinophilic infiltration of bowel wall

Excluding Differentials

- Chronic recurrent abdominal pain, emesis and eosinophilia in the absence of esophageal, malabsorptive and biliary pathologies amongst others suggested an eosinophilic gastroduodenal etiology.
- Symptoms started long before lisinopril initiation. The timeline of symptoms, unremarkable C4 level and the absence of diffuse bowel wall edema, ascites and/or inflammation of surrounding organs on abdominal CT scan ruled out ACE inhibitor-induced angioedema.
- Secondary causes of eosinophilia such as parasitic infections, celiac disease, inflammatory bowel disease, neoplasia and hypereosinophilic syndrome were excluded

Epidemiology and Diagnosis

- Prevalence rate is 5.1/100,000.
- More common in Caucasians.
- Occurs typically around 30 to 50 years of age.
- Pathogenesis speculated to be related to a hypersensitivity response with eosinophilic infiltration of the bowel wall.
- A personal or familial history of atopy is frequently described.
- Diagnosed with > 20 Eo/HPF on gastric biopsies and >30 Eo/HPF on duodenal biopsies.
- Secondary causes of eosinophilia should be excluded

Histological Types

- Klein classified EGE into predominant mucosal, predominant muscle layer and predominant subserosal disease.
- In our case, the sole involvement of the mucosal layer by eosinophilic infiltration, absence of bowel obstruction and eosinophilic ascites on abdominal CT scan favored the mucosal EGE subtype.

Treatment

- Spontaneous remission can occur in more than a third of patients.
- Most of the patients require treatment and steroids are firstline.
- Disease course is different between the subtypes of EGE.
- Course is continuous in predominant mucosal disease, recurring in predominant muscle layer disease and manifests as a single flare in predominant subserosal disease.
- High rates of relapse are seen in proximal small bowel disease, extensive bowel involvement in mucosal EGE, higher eosinophilia and absence of spontaneous remission.
- Our patient received prednisone 40 mg daily and tapered over 8 weeks leading to resolution of symptoms and eosinophilia.