An Unusual Case of Eosinophilic Ascites with Pleural Effusion - A Rare Manifestation of Eosinophilic Gastroenteritis (EGE)

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Abstract

Eosinophilic gastroenteritis (EGE) is a rare disease characterized by tissue eosinophilia and can affect any part of gastrointestinal (GI) tract from the esophagus to the rectum, although stomach and small intestine are sites most frequently involved. We hereby describe an unusual case of eosinophilic gastroenteritis affecting the stomach, small intestine, colon and rectum involving the mucosa and serosa. A twenty-oneyearold student presented with fever, diarrhea, ascites and right pleural effusion. Total leucocyte count was high with marked eosinophilia. Ascitic and pleural fluid were exudates with low adenosine deaminase (ADA) level and predominant eosinophils. Biopsy specimens of the stomach, duodenum, ileum, colon and rectum showed dense eosinophilic infiltration of lamina propria. Based on the constellation of clinical features and investigations, a diagnosis of EGE was made, and therapy with prednisone was started. Symptoms and peripheral eosinophilia rapidly resolved. It is thus imperative to diagnose this disease early and institute the necessary treatment.

Keywords: Eosinophils; Distension; Diarrhea; Ascites; Biopsy; Gastrointestinal.

Key points

- Diagnosis of eosinophilic gastroenteritis (EGE) remains a diagnostic challenge.
- It is characterized by eosinophilic infiltration of the bowel wall, various gastrointestinal symptoms and exclusion of secondary causes of eosinophilic infiltration.
- Eosinophilic ascites with pleural effusion are rare presenting manifestations of EGE and are caused by serosal involvement.
- Peripheral eosinophilia may or may not be present.
- Diagnosis requires a high index of suspicion.
- Corticosteroids are the mainstay of therapy with a 90% response rate.

Introduction

Eosinophilic gastroenterocolitis (EGE) is uncommon gastrointestinal (GI) disease, characterized by eosinophilic infiltration in the gut[1]. It is diagnosed by a triad of GI symptoms, presence of eosinophils in at least one segment of GI tract, and exclusion of secondary causes of tissue eosinophilia[2]. We present a hitherto an unreported case of EGE who had both mucosal and serosal involvement who presented with diarrhea, ascites and right pleural effusion.

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Case Report

A twenty-one years old, previously healthy student was admitted for fever and passing watery, yellowish stools without blood and mucous and abdominal

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distension for 8 days. She reported colicky abdominal pain before passing stool. She did not have addictions and allergies. On examination, she had no pallor, icterus and pedal edema. Her vitals were normal. Breath sounds were diminished in right infrascapular region with dullness on percussion. Abdomen was soft with fluid thrill. There was no organomegaly. Other systems were normal.

Her hemoglobin was 14.1gm/dl, WBC count 25,500/µL (52% neutrophils, 12% lymphocytes, 29% eosinophils, 7% monocytes, absolute eosinophil count – 7395/µL) and platelet count 3.5L/cu mm. Liver and renal function tests were normal. Total serum proteins were 6.0g/dl, serum albumin 2.97 g/dl, C- reactive protein 2.28 mg/dl. Urine and stool routine examination were normal. Abdominal ultrasound showed grade 1 fatty liver with moderate ascites. Chest radiograph showed moderate right pleural effusion. Echocardiography was normal. Serum IgE level was 2500IU/ml. Anti-nuclear antibodies (ANA) were not detected.

Ascitic fluid was straw colored. Analysis showed total 1,040 cells - 60% neutrophils, 25% eosinophils and 15% lymphocytes, glucose 93 mg/dl, LDH 131.6U/dl, proteins 4.3g/dl, albumin 2.45g/d, adenosine deaminase (ADA) 2.6U/L. Smear revealed neutrophils and eosinophils. Malignant cells and parasites were not found. Cultures for bacteria and fungi were negative. Pleural fluid was also exudate with ADA 4.3U/dl and eosinophilic predominance.

Contrast enhanced computed tomography (CECT thorax) showed moderate right pleural effusion (Figure 1a). CECT of abdomen revealed mild hepatomegaly, moderate ascites, thickening of the pylorus, collapsed colon with submucosal edema and proximal jejunal thickening. No abnormal mass was detected (Figure 1b-d).

The differentials considered were inflammatory bowel disease, intestinal tuberculosis and eosinophilc gastroenteritis (EGE). She underwent esophagogastroduodenoscopy with colonoscopy for tissue diagnosis (Figures 2a-d). There was patchy hyperemia of gastric mucosa especially pylorus, duodenum first part, (D1) terminal ileum, sigmoid colon and rectum. No ulcers or growth were found. Rapid urease test was negative for Helicobacter pylori.

Histopathological examination of biopsies of gastric antral mucosa and duodenal biopsy specimen showed diffuse eosinophilic infiltrate mixed with few neutrophils and lymphocytes in the lamina propria. Biopsy of terminal ileum and rectum also revealed focal neutrophilic aggregates and diffusely infiltrating eosinophils in lamina propria. There was no evidence of crypt abscess, granuloma formation, dysplasia or malignancy (Figures 3A-D). Acid fast bacilli and parasites were not found. These findings were suggestive of EGE.

Patient was treated with intravenous hydrocortisone 100 mg thrice/day. Her diarrhea responded to steroids within 48 hours. On tenth day, her eosinophil count was 120/ µL. She continued oral prednisolone 40 mg /day for 2 weeks and was subsequently advised to taper by 5 mg every week.

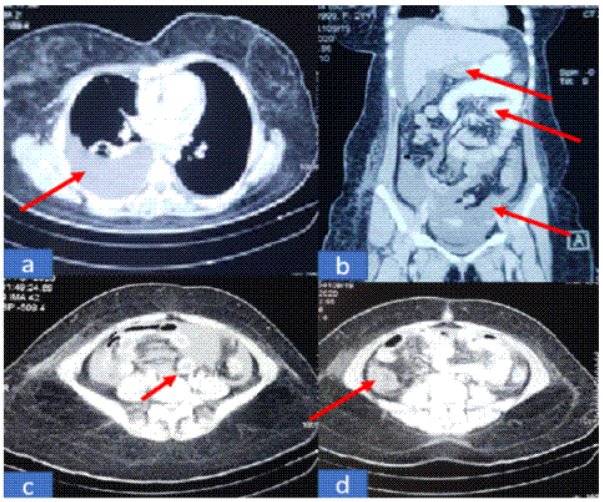

Figure 1 a - d: Showing Contrast enhanced computed tomography (CECT thorax) of Chest and Abdomen

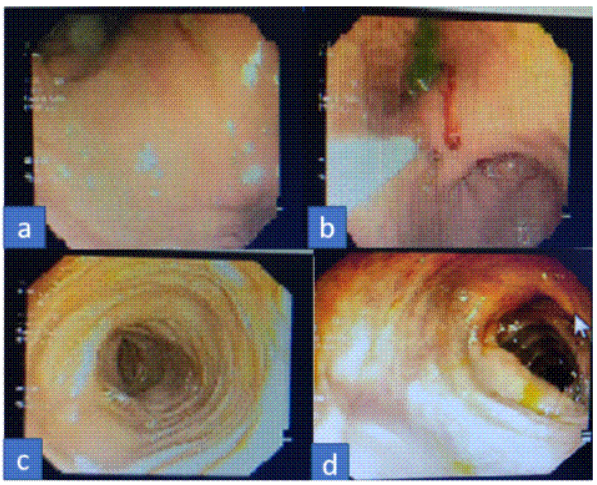

Figure 2 a - d: Showing esophagogastroduodenoscopy and colonoscopy images

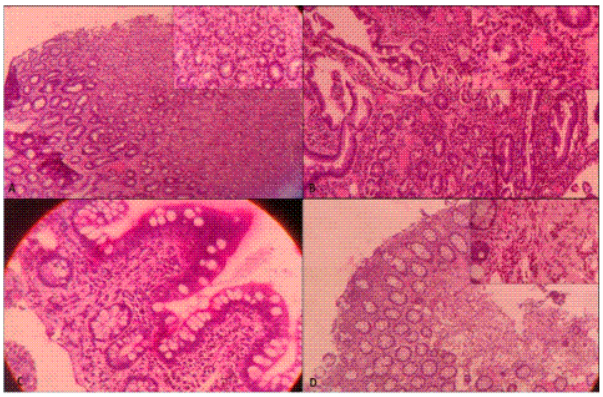

Figure 3 a - d: Showing Histopathological examination of biopsies of the gastric antral mucosa and duodenum.

Discussion

EGE is a rare inflammatory condition of unclear etiology. It affects the entire GI tract from the esophagus to colon. It is characterized by presence of tissue eosinophils in at least one segment of GI tract, GI symptoms and absence of other causes of tissue eosinophilia[2]. It was initially described by Kaijser in 1937[3]. It usually affects adults in third to fifth decade and uncommonly pediatric populationand shows male preponderance [3].The estimated prevalence reported is 1 in 100,000[4].

It is important to differentiate it from secondary diseases associated with eosinophilia like hypereosinophilic syndrome, inflammatory bowel disease, parasitic infestationslike Toxocara and Strongloides, vasculitis like Churg-Strauss syndrome, polyarteritis nodosa, connective tissue diseases like systemic lupus erythematosus, scleroderma, neoplasms, allergic reactions to drugs like nonsteroidal anti-inflammatory drugs, enalapril, carbamazepine, clopidogrel, interferon and IgE mediated food allergies[4]. EGE is associated withatopic conditions in 80% of cases, and food allergyin 62% of cases[3]. Also, 16% of patients have a family member with similar disorder[5].

Eosinophils are normally present in GI tract in few numbers; to provide innate anti-parasite immunity. Eosinophilic recruitment and degranulation is mediated by interleukins - IL3, IL5, granulocyte–macrophage colony-stimulating factor (GM-CSFs) and eotaxin (chemokine) [1,4,6]. Their degranulation causes severe inflammatoryresponse via reactive oxygen formation, eosinophil derived neurotoxins and halide acids[7].

It is classified into 3 types (Klein classification) - mucosal, muscularis and serosal forms[8]. Clinical manifestations depend on the affected layers. Mucosal variant is the commonest form and manifests as diarrhea, vomiting, abdominal pain or protein-losing enteropathy. Muscularis involvement results in gut wall thickening and manifests as intestinal obstruction[1,4]. Serosal form is uncommon and leads to eosinophilic ascites (EA). It may be associated with pleural effusion[9] as in our case. It may take a relapsingremittingcourse in 25% of patients[4].

Peripheral blood eosinophilia should raise the suspicion of EGE. Our patient had severe peripheral eosinophilia. Hypoalbuminemia may be present, especially in patients with mucosal involvement[1,6]. Total serum IgE ⩾100 IU/mL is reportedly present in about two-thirds of EGE cases[1]. Upper GI endoscopy may show normal mucosa or erythematous, nodular, ulcerated mucosa, pseudopolyps, and polyps[6,10].

The diagnosis can be confirmed on histopathological examination of the affected region which demonstrates increased numbers of eosinophils (> 30 per high-power field) in the lamina propria, sometimes infiltrating the muscularis and serosa. Infiltration is often patchy, can be missed[6]. CT abdomen is useful in assessing the extent of disease and response to treatment but has little value in diagnosis[1].

Corticosteroids remain the mainstay of therapy with rapid response. Duration of steroid treatment is unknown and relapse often necessitates long term treatment. Other steroidsparing agents like sodium cromoglycate, ketotifen andmontelukast have been used, considering the allergichypothesis, with variable results. Dietary modifications may be needed. Long-term prognosis is good, mortality is rare. EGE does not predispose to GI malignancy.

The predominance of eosinophils in ascitic and pleural fluid, tissue eosinophilia as evidenced in endoscopic biopsies, absence of extra-intestinal involvement, no evidence of secondary cause of eosinophilia and dramatic response to corticosteroids made us diagnose EGE. Our case had simultaneous involvement of the serosal (characterized by ascites, pleural effusion) and mucosal layer (hypoalbuminemia, anorexia, diarrhoea of 2 weeks). Pleural effusion has been described in 11% of patients with eosinophilic ascites by Durieu I et al. Further 75% of the patients with eosinophilic ascites were

females of ⩾ 40 years and >55% had history of atopy[11]. Our patient was young, without any history of atopy, had right pleural effusion and had bilayer involvement, making the case unique for presentation.

Consent: Informed consent was obtained from the patient

Conflict of interest: Nil

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