Eosinophilic enteritis: A case report

Ilavarasi Lakshmanan, Ratnakar Kini, Pugazhendhi Thangavelu, Mohammed Ali

ABSTRACT

Introduction: Eosinophils have a key role in the pathogenesis of a number of gastrointestinal diseases. Eosinophilic gastroenteritis is a rare benign disease characterized by tissue eosinophilic infiltration that may involve several digestive tract layers. The clinical manifestations are related to the layers and extent of the bowel involved. The disease may involve any part of the gastrointestinal tract, but the stomach and the small intestine are the most common sites involved.

Case Report: In this report, we present a case of intractable abdominal pain caused by jejunal mucosal eosinophilic infiltration, without any secondary causes of eosinophilia, diagnosed by deep endoscopic biopsies. The patient was successfully treated with steroids without need for surgery for diagnosis or therapy.

Conclusion: This entity emphasizes the importance of an invasive approach along with a thorough medical history and a careful histological assessment. Our patient had a drastic response to steroid therapy and currently on follow-up without any complications.

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Keywords: Eosinophilic gastroenteritis, Eosinophilic infiltration, Jejunal mucosa, Steroids

INTRODUCTION

Eosinophilic infiltration of the digestive tract was first described by Kaijser in 1937 [1]. It is diagnosed in the biopsies taken during endoscopic examination in patients presenting with abdominal pain and chronic diarrhea [2]. The disorders associated with gastrointestinal eosinophilia are largely categorized into primary and secondary. Primary gastrointestinal eosinophilia, also termed eosinophil associated gastrointestinal disorders (EGID) selectively affect the gastrointestinal tract with eosinophil-rich inflammation in the absence of known causes for eosinophilia. It is also known as allergic gastroenteropathy or eosinophilic allergic gastroenteropathy [3]. EGIDs, or primary eosinophil-associated gastrointestinal disorders, include eosinophilic esophagitis (EoE),
eosinophilic gastritis, eosinophilic gastroenteritis, eosinophilic enteritis, and eosinophilic colitis or proctitis. The pathogenesis is not fully understood but hypersensitivity along with chemokines play a central role in eosinophilic migration and inflammation in both tissues and blood. EGID inflammation is believed to be driven by CD4+ Th2 cells [4]. These cells are the key component in the production of IL-4, IL-13, IL-5, IL-10 and the control of allergic inflammation responses. IL-4 and 13 are responsible for Th2 cell production and IgE by B cells and IL-5 controls eosinophil production, activation and survival. Eotaxin is a chemokine, constitutively expressed in the gastrointestinal tract. In exotoxin-deficient mice, eosinophil recruitment into the mucosal lining was not seen with allergen stimulation, proposing its importance in pathogenesis of eosinophilic gastroenteritis [5, 6]. Secondary eosinophil-associated gastrointestinal disorders include those disorders where eosinophil accumulation can be attributed to a distinct cause. These disorders are further categorized as those due to eosinophilic disorders (usually hypereosinophilic syndrome) or those due to non-eosinophilic disorders (such as inflammatory bowel diseases, parasitic infestations, infectious diseases, connective tissue disorders, vasculitides, neoplasia, and drug reactions).

CASE REPORT

A 25-year-old male presented to the medical gastroenterology department with intractable, intermittent, left lower quadrant pain of three months duration. There was no fever, weight loss, change in bowel habits, nausea, vomiting or rash. He had no food or pollen allergies. He did not have any relevant medical history of atopy. He also denied any history of illicit drug use. He was an occasional alcohol consumer and a non-smoker. He has not taken any medications recently. The physical examination was unremarkable. Of the baseline investigation hemogram showed leukocytosis of 12,500 per mm$^3$ (a differential of neutrophils 36%, lymphocytes 25%, eosinophils 34% and monocytes 5%) with normal hemoglobin and platelets. His absolute eosinophil count (AEC) was 3200 cells/mm$^3$ and serum immunoglobulin E levels were 100 IU/ml. Serum amylase, lipase and liver functions were normal. Abdominal ultrasonography, chest X-ray and echocardiogram were also normal. Repeated stool examination was negative for ova and parasites. A contrast enhanced computed tomography scan of abdomen revealed long segment jejunal wall thickening with few enlarged mesenteric lymph nodes (Figure 1). He was submitted for push enteroscopy under intravenous sedation which showed edematous, erosive mucosa in the proximal jejunum. Multiple biopsies were taken from the abnormal jejunal mucosa which revealed >50 eosinophils/high power field favoring eosinophilic enteritis (Figures 2 and 3). Gastroduodenoscopy and ileocolonoscopy were performed and found to be normal. Random biopsies were taken from esophagus, stomach, duodenum, colon and ileum which revealed no eosinophil, hence a diagnosis of primary eosinophilic enteritis (jejunitis) was concluded in this patient. The patient was started on steroid therapy, prednisolone 20 mg/day and he had complete symptomatic recovery within a few days. He was kept on the same dosage of steroids for two weeks and was subsequently tapered over the next few weeks. Currently the patient is off steroids for the last three months without any complaints.

Figure 1: Contrast enhanced computed tomography scan of abdomen showing long segment jejunal wall thickening with few mesenteric lymph nodes (Pointed by arrows).

Figure 2: Jejunal mucosal biopsy showing numerous eosinophils per high power field in the lamina propria with no evidence of granuloma or malignancy.
DISCUSSION

Eosinophilic gastroenteritis is a rare, benign condition characterized by significant eosinophilic infiltration of the wall of the gastrointestinal tract that usually involves the stomach and small intestine and, rarely, the colon [7]. The stomach and duodenum are the most common sites of involvement [8]. It may or may not be accompanied by elevated numbers of eosinophils in the peripheral blood.

Little is known about the prevalence of this disorder. It can affect both sexes although it seems to be more common in men. The peak age at presentation is in the third decade.

Four criteria are required for the diagnosis of EGE: the presence of gastrointestinal symptoms, eosinophilic infiltration of the gastrointestinal tract, exclusion of parasitic disease or other identifiable cause of eosinophilia and the absence of other systemic involvement [9, 10].

Patients with eosinophilic gastroenteritis (EGE) have also been divided into two groups. First group of patients have atopy, asthma, elevated serum IgE levels, a peripheral eosinophilia and positive allergy tests. Patients in the second group have no history of atopy, usually have normal serum IgE levels, and negative response to allergy tests.

Clinical features vary depending upon the location and layer of gastrointestinal tissue involved. EGID can be classified into various types based on etiology, atopic component, site and layer of involvement (Table 1). Klein in 1970 classified EGE into three types based on the layer of involvement of the gastrointestinal system [11]. The mucosal subtype is the most common one which presents with decreased appetite, early satiety, abdominal pain, nausea, vomiting, diarrhea, occult gastrointestinal bleeding, anemia, weight loss, or protein-losing enteropathy. Submucosal form comprehends infiltration of eosinophils in the muscularis layer leading on to thickening of the bowel wall that may result in gastric outlet obstruction or intestinal obstruction. The serosal layer involvement is least common and manifest as painful peritonitis or exudative ascites with higher peripheral eosinophil counts. Combinations of these symptoms occur with involvement of more than one tissue layer. The prevalence of each subtype is unknown because of reporting and referral biases. Along with eosinophilic cholangitis and acute pancreatitis, eosinophilic cholecystitis can also occur.

Evaluation is mainly to rule out any secondary causes of eosinophilic gastroenteritis. A careful search for food allergy, drug intake and gut parasites should be undertaken in all patients. A personal history of allergic disease, such as eczema, asthma, allergic rhinitis, or animal dander sensitivity, suggests the possibility of EGIDs. A complete medication history, including allergen immunotherapy and the use of over-the-counter medications, vitamin supplements, and herbal remedies should be obtained. Medications induce gastrointestinal eosinophilia include interferon, gemfibrozil, enalapril, carbamazepine, clofazimine and co-trimoxazole. Occupational exposures, foreign travel, camping, ingestion of well or stream water should also be questioned. Among parasites, tissue eosinophilia also may be found with hookworms (Ankylostoma caninum), pinworms (Enterobius vermicularis), Giardia lamblia, Anisakis, Trichinella spiralis, Ascaris, Trichuris, Schistosomiasis, Toxocara canis, and Strongyloides stercoralis. Various connective tissue disorders (e.g., scleroderma, dermatomyositis, lupus) and vasculitis (e.g., Churg–Strauss syndrome, polyarteritis nodosa) are also associated with variable peripheral and gastrointestinal eosinophilia. Hypereosinophilic syndrome should also be ruled out which is a rare disorder with unexplained

Table 1: Classification of eosinophilic gastroenteritis (EGE)

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<td>• Primary (absence of known causes of eosinophilia)</td>
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<td>• Secondary (associated with secondary causes like drugs, atopy, parasitic infection)</td>
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<th>Based on Atopy</th>
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<td>• With Atopy (asthma, elevated serum IgE levels, a peripheral eosinophilia and positive responses to allergy tests)</td>
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<tr>
<td>• Without Atopy (no history of atopy, usually have normal serum IgE levels, and negative response to allergy tests)</td>
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<tr>
<th>Based on layer of digestive tract involvement</th>
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<td>• Mucosal (abdominal pain, nausea, vomiting, malabsorption)</td>
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<td>• Muscular (gastric outlet/small intestinal obstruction)</td>
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<td>• Serosal (ascites, peritonitis)</td>
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<th>Based on the site of involvement</th>
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<td>• Eosinophilic esophagitis</td>
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<td>• Eosinophilic gastritis</td>
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<td>• Eosinophilic gastroenteritis</td>
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<td>• Eosinophilic enteritis</td>
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<td>• Eosinophilic colitis</td>
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<td>• Eosinophilic proctitis</td>
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marked peripheral hypereosinophilia (>1500 cells/L for more than six consecutive months) and presence of organ damage (heart or CNS).

Endoscopically the gastrointestinal mucosa may vary in appearance from normal gastric/small intestinal mucosa to ulcerated, erythematous and nodular mucosa with multiple polyps.

There are no consensus guidelines on the requisite number of eosinophils needed for the diagnosis of EGE. Eosinophil counts of 30 eos/hpf (eosinophils per high powered field) in the stomach and 50 eos/hpf in the duodenum have been proposed for the diagnosis of eosinophilic gastroenteritis and duodenitis, respectively. In addition, the location of eosinophils (intraepithelial eosinophils, intraglandular eosinophils, and eosinophils in the muscularis), and epithelial hyperplasia also point towards the diagnosis of EGE.

Steroids remain the cornerstone of medical treatment in all types of eosinophilic gastroenteritis. Improvement is observed in up to 90% of cases [12]. However, the duration is not specified, and relapse is not uncommon. Prednisone 20–40 mg daily or its equivalent usually results in dramatic improvement of all manifestations of the disease. After 7–10 days of therapy the dose can frequently be tapered slowly over several weeks (approximately six to eight weeks). Budesonide proposed by Tan et al. has been found to be effective and safer than prednisolone as it inhibits both eosinophilic activation and survival [13]. Mast cell stabilizer (sodium cromoglycate), antihistamines (ketotifen) are the other drugs that have been proposed as therapeutic measures with inconclusive results. Recently, montelukast, has been successfully been used in a patient with relapsing eosinophilic gastroenteritis. Surgery becomes useful in cases of obstructive symptoms lacking improvement with medical treatment. Mortality is generally rare and there is no increased risk of gastrointestinal malignancies. Response assessment is mainly clinically and repeat endoscopy and biopsies.

CONCLUSION

Eosinophilic disorders of the gastrointestinal tract emphasize the importance of an invasive approach along with a thorough medical history and a careful histological assessment. Our patient had a drastic response to steroid therapy and currently on follow up without any complications.

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Author Contributions
Ilavarasi Lakshmanan – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Ratnakar Kini – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Pugazhendhi Thangavelu – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Mohammed Ali – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

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