Eosinophilic Gastroenteritis as the Initial Manifestation of Hypereosinophilic Syndrome

NARISA SS¹, SHANTI P², JEEVINESH NA², SAKTHISWARY R²

¹Department of Medicine, Faculty of Medicine and Health Sciences, Universiti Putra Malaysia, 43400, Serdang, Selangor Darul Ehsan, Malaysia.
²Department of Medicine, Faculty of Medicine, Universiti Kebangsaan Malaysia Medical Centre, Jalan Yaacob Latif, Bandar Tun Razak, 56000 Cheras, Kuala Lumpur, Malaysia.

ABSTRACT

Eosinophilic gastroenteritis, an inflammatory disease of unknown etiology, commonly involves the stomach and small intestine with eosinophilic infiltration. Here, we report an unusual case of eosinophilic gastroenteritis involving the entire digestive tract as a manifestation of hypereosinophilic syndrome (HES). A 22-year-old woman presented to us with diarrhoea, pleural effusion, ascites and...

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Address for correspondence and reprint requests: Assoc Prof Dr. Rajalingham Sakthiswary, Department of Medicine, Faculty of Medicine, Universiti Kebangsaan Malaysia Medical Centre, Jalan Yaacob Latif, Bandar Tun Razak, 56000 Cheras, Kuala Lumpur, Malaysia. Tel: +603-91456097 Fax: +603-91456679 Email: sakthis5@hotmail.com
marked peripheral eosinophilia. Stool specimens were negative for parasites, ova, bacteria, and fungi. Endoscopic studies showed pangastritis and duodenitis. Biopsy specimens of the oesophagus, stomach, duodenum, ileum, and colon demonstrated eosinophilic infiltration. A diagnosis of hypereosinophilic syndrome with eosinophilic gastroenteritis involving the entire digestive tract was made. Hence, she was treated with prednisolone. Symptoms and peripheral eosinophilia rapidly resolved with treatment, and radiological investigations revealed resolution of effusion. This case illustrates the wide spectrum of clinical manifestation of the disease, whereby it involves the entire digestive tract and it also emphasizes the diagnostic yields of endoscopic biopsies.

Keywords: hypereosinophilic syndrome, eosinophilic gastroenteritis, endoscopic biopsies, corticosteroids

**INTRODUCTION**

Eosinophilic gastroenteritis (EG) is a rare manifestation of HES with a frequency of around 25% (Rothenberg 2004). The clinical features of EG varies based on the region and depth of the gastrointestinal tract (GIT) affected by the eosinophilic infiltration (Rothenberg 2004; Shifflet et al. 2009; Freeman 2008; Conus & Simon 2008). The common symptoms include abdominal pain, diarrhea and vomiting. The definitive diagnosis of EG requires histological evidence of eosinophilic infiltration of >20 eosinophils/high-powered field in the lamina propria (Rothenberg 2004; Shifflet et al. 2009; Liacouras 2007). The main aim of the treatment is to reduce the eosinophil counts. In general, the majority of patients respond well to steroid therapy (Conus & Simon 2008; Liacouras 2007; Chen et al. 2003). However, relapses are common and such patients may require long term immunosuppressive agents (Chen et al. 2003).

**CASE REPORT**

A 22-year-old lady with a past medical history of well controlled eczema and asthma presented with a history of diarrhoea, vomiting and abdominal pain for one week. The initial blood investigations showed leucocytosis with marked eosinophilia (TWC of 41.2 with eosinophil of 69%) (Figure 1). She was treated initially as acute gastroenteritis with intravenous fluid and antibiotics but her symptoms persisted. Over the span of few days, she developed dyspnoea. Her chest radiograph and CT scan of her chest and abdomen revealed bilateral pleural effusion and ascites. She also developed purpuric rashes over the dorsum of her hands associated with joint pain.

Her blood, stool and urine cultures were negative. Stool specimens were also negative for ova and cysts. In view of her persistent diarrhoea, patient underwent an upper and lower gastrointestinal endoscopy. Her upper endoscopy showed pangastritis and
duodenitis. The gastric and duodenal biopsies showed significant eosinophil infiltration (>20 per high power field) each (Figure 2). Her colonoscopy revealed mild ileitis and pancolitis predominantly involving the descending and sigmoid colon. The biopsies taken throughout the colon and terminal ileum also demonstrated presence of mild to moderate infiltration of the lamina propria by eosinophils of >30 per high power field (Figure 3 and 4). A skin biopsy revealed scattered eosinophils at the periadnexal and perivascular area. Bone marrow trephine exhibited increased number of eosinophils and its precursors. No malignant cells were observed.

Hence, the provisional diagnosis was idiopathic hypereosinophilic syndrome with eosinophilic gastroenteritis, cutaneous involvement and serositis. The patient was started on prednisolone 0.5mg/kg and the dose was tapered slowly over the next four weeks. She was discharged well and six weeks after treatment, she was asymptomatic and her serositis had resolved. Her total white count normalized and the percentage of eosinophils reduced to 9%.

**DISCUSSION**

Idiopathic hypereosinophilic syndrome (HES) constitutes a rare and heterogeneous group of disorders...
with unknown prevalence (Roufosse et al. 2007). The term HES was first introduced by Hardy and Anderson in 1968 and in 1975, Chusid et al. established four diagnostic criteria for idiopathic HES (Roufosse et al. 2007; Kahn et al. 2008; Klion 2005) namely the presence of i) blood eosinophilia >1500/mm³ for at least six months, ii) absence of an underlying cause of eosinophilia despite extensive evaluation, and iii) presence of end organ damage or dysfunction related to the eosinophilia. HES has a predilection for the skin, heart and nervous system but almost any tissue or organ can be affected. Of note, the cardiovascular and hematological systems account for more than 50% of the disease manifestation (Roufosse et al. 2007). The onset of HES is generally between the age of 20-50 years with male preponderance (Freeman 2008; Klion 2005).

About 25% of HES patients have eosinophilic infiltration of the gastrointestinal tract which may involve almost any region from the esophagus to rectum (Freeman 2008). Although the eosinophilic infiltration in EG can involve any part of the gastrointestinal tract, this disease is characterized by predominant involvement of the gastric and small intestinal mucosa (Rothenberg

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**Figure 3:** Colonoscopy showed mild ileitis with pancolitis, predominantly involving the descending, sigmoid colon with no loss of vascular pattern

**Figure 4:** Histopathological examination of rectum and terminal ileum showed presence of mild to moderate eosinophilic infiltration of lamina propria of > 30 per high power field
EG as the initial manifestation of HES with such extensive involvement of the entire digestive tract is extremely rare.

The exact etiology of EG is unknown but up to 40% cases were associated with underlying allergies or atopy, namely food allergies, eczema, allergic rhinitis and asthma (Chen et al. 2003; Nair et al. 2011). The elevated Ig E levels in the sera of affected patients implies that atopy maybe involved in the pathogenesis of EG (Rothenberg 2004; Chen et al. 2003). In the present case, the patient had asthma and eczema.

Iron deficiency associated with blood loss as well as protein-losing enteropathy are known complications of this disease entity. If muscular layers are involved, intestinal obstruction may ensue, while serosal involvement may be associated with ascites (Rithenberg 2004; Freeman 2008; Chen et al. 2003). The endoscopic appearance of EG is nonspecific. The abnormal findings include erythematous mucosa, friable, nodular, and ulcerative changes. In a retrospective study by Chen et al. (2003) involving 15 patients with EG, 10 of the patients had only nonspecific gastritis or colitis while two patients had shallow gastritis and duodenal ulcers (Chen et al. 2003).

Definitive diagnosis requires histological evidence of eosinophilic infiltration. Oeosinophilic infiltration is usually patchy in distribution and may be present in otherwise normal, non-inflamed bowel wall (Rothenberg 2004; Chen et al. 2003). Because of this, multiple biopsies should be taken to improve diagnostic yield. For the purpose of deriving the diagnosis of EG, the majority of studies have used a definition of >20 eosinophils/ high-powered field in the lamina propria (Rothenberg 2004; Shifflet et al. 2009; Liacouras 2007).

The evidence for the management of EG is largely based on case studies and retrospective studies (Liacouras 2007; Chen et al. 2003). Presently, there is no randomized trial on its treatment because of the rarity of the disease (Liacouras 2007). The main aim of the management of EG is to reduce the eosinophil counts. In general, patients respond rapidly to steroids (Conus & Simon 2008; Liacouras 2007; Chen et al. 2003). However, the optimal duration of the steroid therapy is unknown but Chen et al. (2003) demonstrated that short courses of steroid therapy followed by repeated courses for relapses gave promising results in treating EG (Chen et al. 2003). Our patient was treated with prednisolone 0.5 mg per kg for two weeks which was tapered after two weeks. This regime resulted in marked symptomatic and radiological improvement. For patients who have frequent relapses of EG, long term immunosuppressive agents or low dose corticosteroids is strongly recommended (Chen et al. 2003). Few authors have described the use of sodium cromoglycate (a stabilizer of mast cell membranes) or montelukast (a selective, competitive leukotriene receptor antagonist) as steroid-sparing agents (Schwartz et al. 2001).

REFERENCES