Eosinophilic Gastroenteritis: A Case Series from Iran

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OBJECTIVE

Eosinophilic gastroenteritis (EG) is a rare inflammatory disorder of the gastrointestinal (GI) tract. There have been several case series of patients with EG from the western world and East Asia. However, there has not been a report of patients with EG from the Middle East region. The aim of this study is to describe clinical characteristics and treatment response in a series of EG patients from Iran.

METHODS

We retrospectively reviewed charts with a diagnosis of EG from 1997 to 2010 at Shariati Hospital and the private clinics of the authors. Clinical characteristics of the patients were evaluated, and the treatment response and relapse rate were assessed.

RESULTS

Twenty-two patients (9 male) with EG were identified. Mean age of the patients was 45.1±15.5 (range: 27-75) years. Median duration between symptom onset and diagnosis was 12 (range 1- 48) months. Twenty (90%) patients had mucosal involvement, one (5%) had muscular involvement and one (5%) had subserosal involvement.

Patients were followed for a median duration of 36.5 (range 4-123) months. Two patients had spontaneous remission with supportive care. The remaining 20 patients responded well to oral corticosteroid treatments. The relapse rate was 33%. Episodes of relapse were successfully controlled with a repeat course of corticosteroids. Two patients with several relapses required maintenance treatment with azathioprine.

CONCLUSION

The clinical characteristics and treatment responses of EG patients from Iran are similar to reports from other parts of the world. Patients need to undergo close follow up after treatment to detect early signs of relapse.

KEYWORDS

Eosinophilic gastroenteritis; Esophagus; Small intestine; Eosinophils

INTRODUCTION

Eosinophilic gastroenteritis (EG) is a rare inflammatory disorder, characterized by eosinophilic infiltration in one or multiple areas of the gastrointestinal (GI) tract from the esophagus to the rectum.

Clinical presentations depend on the region of the GI tract involved and the depth of bowel wall involvement. Klein et al.¹

ABSTRACT

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Eosinophilic gastroenteritis (EG) is a rare inflammatory disorder, characterized by eosinophilic infiltration in one or multiple areas of the gastrointestinal (GI) tract from the esophagus to the rectum.

Clinical presentations depend on the region of the GI tract involved and the depth of bowel wall involvement. Klein et al.¹
classified EG into three patterns based on the depth of disease process. The three main patterns include predominant mucosal disease, predominant muscle layer disease and predominant subserosal disease.

Reports from different parts of the world provide important information regarding epidemiology, disease characteristics and management. There have been several case series of patients with EG from the western world, Australia and East Asia.

To our knowledge, there has not been report of a series of patients with EG from the Middle East region. The aim of this study was to describe clinical characteristics and treatment response in a series of EG patients with long-term follow up from Iran.

MATERIALS AND METHODS

This study was approved by the Institutional Review Board of Digestive Disease Research Center, Tehran University of Medical Sciences.

Using prospectively updated databases at Shariati Hospital and private outpatient clinics of the authors from January 1997 to December 2010, we identified and reviewed medical records of all patients with the diagnosis of EG. EG was defined by the presence of all the following criteria: 1) GI symptoms attributable to EG, 2) biopsies showing marked eosinophilic infiltration of one or more regions of the GI tract from the esophagus to the colon (>20 eosinophils per high power field) and 3) exclusion of parasitic infection or extraintestinal infiltration of eosinophils.

We collected the following data: demographic characteristics, presenting symptoms, date of symptoms onset, date of diagnosis, duration of follow up, absolute eosinophil count, treatment regimen and number of possible relapses.

Relapse was defined by the presence of one of the following criteria: 1) reappearance of GI symptoms with re-elevation of peripheral blood eosinophilia, 2) reappearance of GI symptoms and restarting immunosuppressive drugs by an experienced gastroenterologist and 3) reappearance of GI symptoms and re-demonstration of EG on biopsy specimen after previous normalization of biopsy specimens.

Patients were divided into the following groups: 1) those with predominantly mucosal disease as defined by infiltration of mucosa with eosinophils, no evidence of intestinal obstruction or eosinophilic ascites; 2) those with predominant disease in muscular layer as defined by the presence of intestinal obstruction and eosinophilic infiltration of muscular layer, and absence of eosinophilic ascites; and 3) those with predominant subserosal disease manifested by eosinophilic infiltration of the GI tract and eosinophilic ascites.

Descriptive and frequency statistical analyses were obtained for each of the variables analyzed. Values were expressed as means and standard deviations, or medians and ranges.

RESULTS

Twenty-two patients (9 male) with EG were identified. Mean age of the patients was 45.1±15.5 (range: 27-75) years. Of the 22 patients, 18 (82%) had small bowel, 11 (50%) had gastric, 5 (23%) had colonic, and 2 (9%) had esophageal involvement (Table 1, Figures 1A-1C). Twenty (90%) patients had mucosal involvement, one (5%) had muscular involvement and one (5%) had subserosal involvement.

| Involved organs in 22 patients with EG. |
|----------------------------------------|
| Involved organ                          | Number (%) |
| Esophagus                               | 1 (5)      |
| Stomach                                 | 0 (0)      |
| Small bowel                             | 5 (23)     |
| Colon                                   | 3 (14)     |
| Esophagus + stomach + small bowel       | 1 (5)      |
| Stomach + small bowel                   | 10 (46)    |
| Small bowel + colon                     | 2 (9)      |
Three (14%) patients had anemia and 4 (18%) had positive fecal occult blood tests.

Peripheral eosinophilia was observed in 18 (82%) patients. The mean eosinophil count in patients with eosinophilia was 4720 ± 4213 (range: 500-10720) per microliter. Seven (32%) patients had personal allergy histories.

The presenting symptoms included abdominal pain in 15 (68%), diarrhea in 6 (27%), pruritus in 6 (27%), weight loss in 6 (27%), nausea/vomiting in 4 (18%), fever in 3 (14%) and dysphagia in 1 (5%) patients.

The median duration between symptom onset and diagnosis was 12 (range 1- 48) months.

Associated autoimmune disorders included celiac disease (1), sarcoidosis (1) and ulcerative colitis (1).

Patients were followed for a median duration of 36.5 (range: 4-123) months. Two patients with mucosal disease had spontaneous remission with fasting and supportive care, however neither developed relapse after a mean follow up of 44 months.

Twenty patients were treated with prednisolone at initial doses of 15 to 50 mg per day, which was tapered over 6 to 8 weeks. The patients responded well to immunosuppressive treatment with prednisolone. One patient with eosinophilic esophagitis and recurrent dysphagia required several courses of endoscopic esophageal dilation in addition to systemic corticosteroids.

Fifteen patients were followed for more than 12 months. Five out of 15 (33%) developed relapse (range: 1 to 4 relapses) after their initial course of corticosteroid therapy. A repeat course of prednisolone resulted in resolution of symptoms in those who experienced relapse. However, 2 patients with several relapses required maintenance treatment with azathioprine to prevent further episodes of relapse. One of the azathioprine treated patients did not relapse after starting azathioprine; but the second patient who had serosal involvement required low dose prednisolone in addition to azathioprine to maintain remission.

DISCUSSION

Our study represents the first report of patients with EG from Iran. First reported in 1937,6 EG is a rare but increasingly recognized inflammatory disorder of the GI tract characterized by the presence of severe eosinophilic infiltration in one or multiple segments of the GI tract. The pathogenesis of EG is unknown, but it may be related to the destruction of intes-
tinal epithelium by the release of eosinophilic major basic protein from the resident eosinophils. Eosinophilic infiltration may involve various depths of the GI tract.

Patients with predominantly mucosal disease present with diarrhea, abdominal pain, vomiting and nutritional deficiency (i.e., iron deficiency anemia). Those with predominantly muscular involvement present with symptoms of gastric outlet obstruction or intestinal obstruction. Patients with involvement of serosal and subserosal layers present with eosinophilic ascites. The majority of our patients had predominantly mucosal involvement.

Although peripheral eosinophilia is an important guide to diagnosis, it was absent in 19% of our patients. The 81% rate of peripheral eosinophilia in our study was similar to previous case series of EG in the western countries or East Asia.

In our patients, 33% had personal histories of allergy and atopia.

In this study, the median duration between symptom onset and diagnosis was 12 (range 1-48) months. On the other hand, endoscopic findings in the majority of our patients were non-specific. Thus, EG could be easily missed in outpatient clinics. Therefore, gastroenterologists need to be vigilant, including EG in the differential diagnosis of patients with GI symptoms who have associated peripheral eosinophilia, a remarkable personal history of allergy, or those whose GI symptoms do not respond to regular therapeutic measures.

Our patients responded well to a 6 to 8 week course of corticosteroid therapy. In this study, the relapse rate after initial corticosteroid therapy was 36%. Episodes of relapse were successfully treated with a repeat course of corticosteroid therapy. One patient with several relapses needed azathioprine to prevent further relapse episodes. Therefore, patients need to be followed in terms of clinical symptoms and peripheral eosinophil counts to detect early signs of relapse.

In conclusion, we report a series of patients with EG with long term follow up from Iran. The clinical symptoms of EG patients are mostly non-specific and may be confused with functional bowel disorders. Therefore, gastroenterologists need to be aware of this treatable condition, and take mucosal biopsies during endoscopies in patients with suspected EG.

CONFLICT OF INTEREST
The authors declare no conflict of interest related to this work.

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