Eosinophilic gastroenteritis in a patient with Ehlers-Danlos syndrome – A rare combination

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Eosinophilic gastroenteritis is a rare disorder. Patients with this condition are probably underdiagnosed or misdiagnosed as having irritable bowel syndrome, particularly when the disease involves the serosal layer of the small bowel. The diagnosis often is made at surgery and is confirmed by serosal biopsy, although it may also be made by mucosal biopsy when that site is affected. A history of allergy is common, and the standard treatment of choice appears to be glucocorticosteroids. Ehlers-Danlos syndrome also rarely affects the gastrointestinal tract. The case of a young woman with both eosinophilic gastroenteritis and Ehlers-Danlos syndrome is discussed.

CASE PRESENTATION

A 33-year-old female was diagnosed at the Mayo Clinic, Rochester Minnesota, as having Ehlers-Danlos syndrome type VIII when she presented with a history for several months of epigastric pain and vomiting. She was treated with glucocorticosteroids. This was followed by a dramatic loss of muscle power, which reverted to normal after discontinuation of the drug.
The patient continued to be symptomatic with epigastric pain, vomiting and nonbloody diarrhea and she lost 16 kg body weight in two years. The patient had no known food allergies, and a good appetite. She had over 100 suspected drug allergies, ranging in severity from skin rash to near fatal anaphylactic shock which she had suffered twice in the past. Pertinent past medical history included duodenal ulcer disease for which antrectomy and vagotomy was performed; she currently denied symptoms suggestive of dumping syndrome.

The patient was adopted and her family history was not available. She has a 10-year-old daughter who also has Ehlers-Danlos syndrome.

Physical examination disclosed a thin young woman in no apparent distress, with normal vital signs. Abdominal examination revealed a midline scar from previous surgery, a tender epigastric region with no organomegaly and normal bowel sounds. Laboratory tests revealed hemoglobin of 12.2 g/dL, leukocyte count of 4800/mm³ with no eosinophilia and a normal platelet count of 250,000/mm³. The SMA-12 was normal. Stools were negative on several occasions for ova and parasites, culture and sensitivity, Clostridium difficile toxin and culture. Serum IgE level was slightly increased to 177 kunits/L, and vitamin E level was normal. Esophagogastroduodenoscopy revealed normal esophageal mucosa and a small gastric remnant with intense redness and prominent friable folds. Similar changes were present in the first and second part of the duodenum. There was no gastric outlet obstruction. Multiple biopsies from the duodenum revealed dense eosinophilic infiltrate into the lamina propria (Figure 1), compatible with eosinophilic gastroenteritis.

The patient was admitted to hospital to institute sodium chromoglycate treatment while under observation for any possible allergic reaction, as well as to institute a trial of an elemental diet. Sodium chromoglycate was given as a 100 mg tablet test dose orally, which was tolerated well; this was subsequently advanced to 200 mg orally qid. While in hospital, the patient had a J-tube placement and enteral diet feeding with Osmolite (Ross Laboratories). The patient was well when discharged on sodium chromoglycate, an antihistamine (diphenhydramine, Benadryl; Parke-Davis, 50 mg qid) and multivitamins. She gained 5 kg of weight over the initial 10 week treatment period on Osmolite (three cans per day). Oral intake of food was slowly reintroduced, but the patient continued to use the elemental diet to supplement calorie, nitrogen and vitamin intake.

**DISCUSSION**

Eosinophilic gastroenteritis and Ehlers-Danlos syndrome are both rare disorders. In searching the literature, no reported association was found between the two disorders. They co-exist likely by chance alone. The present patient had no history of any of the gastrointestinal manifestations of Ehlers-Danlos syndrome described in

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**Figure 1** Eosinophils are present in the small intestinal lamina propria and infiltrate the epithelium (Luna's stain x160)

**Figure 2** Large numbers of eosinophils are present in the submucosa of the small bowel (Luna's stain x160)
Eosinophilic gastroenteritis

...include dysphagia secondary to mucosal eosinophilic infiltrations of the esophagus, impaired growth in children presumably secondary to malabsorption and protein-losing enteropathy, pyloric outlet obstruction (10), repeated bowel obstruction with perforation requiring multiple surgical resection (11), massive eosinophilic ascites due to serosal involvement, biliary and duodenal obstruction (12), and small bowel perforation (13).

Eosinophilic gastroenteritis can be diagnosed by eosinophil infiltrate in the bowel wall (Figure 2). However, mucosal biopsies may sometimes fail to show eosinophilic infiltrate, as the disease can be patchy or involve only the deep layers. Hence, full thickness biopsies may be needed to confirm the diagnosis. The disease may involve any part of the gastrointestinal tract; the common sites are the duodenum and proximal jejunum, followed by the gastric antrum and body. The terminal ileum is an uncommon site of involvement. The present patient had had an anrectomy, but the jejunum was involved with eosinophilic gastroenteritis.

Some 30% of eosinophilic gastroenteritis patients with mucosal disease have malabsorption and protein-losing enteropathy, and a history of allergy is seen in up to 80% of patients. Peripheral eosinophilia is found in 77% of cases, but was not seen in the current case. Serum IgE level and erythrocyte sedimentation rate may be elevated. The present patient had slightly increased IgE levels, and suffered from multiple allergies. The most common finding on barium examination is thickening and/or irregularity of the mucosal folds in the stomach and/or small intestine (14). Recent barium studies were not available for this patient.

The standard treatment of eosinophilic gastroenteritis is glucocorticosteroid therapy, but this was not tolerated by the present patient. Other treatment modalities include elemental diet, but relapse is common (15-17). While on Osmolite first by J-tube and then by mouth, the patient did well, gaining 5 kg in 10 weeks. Sodium chromoglycate has been shown to prevent release of toxic mediators such as serotonin and histamine from mast cell membranes. It has been successfully used in the treatment of allergic conditions such as asthma, milk allergy gastrointestinal allergy in adults (18) and malabsorption secondary to systemic mastocytosis (19). Unfortunately, there are no double-blind controlled trials for evaluation of the benefit of any of the above treatment modalities in eosinophilic gastroenteritis. Sodium chromoglycate has been successful in several case reports of eosinophilic gastroenteritis (20,21). It appeared to be useful in this patient when combined with an elemental diet.

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REFERENCES


