Eosinophilic Gastroenteritis and Related Eosinophilic Disorders

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KEYWORDS

• Eosinophilic gastroenteritis • Eosinophilic gastritis • Eosinophilia • Food allergy • EGID

KEY POINTS

- Eosinophilic gastroenteritis (EGE) is diagnosed by the presence of gastrointestinal symptoms, biopsies showing predominant eosinophilic infiltration, and the absence of allergic, parasitic, or other diseases that may cause eosinophilia.
- EGE is a rare disease affecting approximately 22 to 28 per 100,000 persons.
- Because EGE may vary by both the site of involvement (stomach, duodenum, jejunum) and the depth of involvement (mucosal, muscularis, or serosal disease), its manifestations are protean.
- Dietary therapy is effective in allergic EGE.
- Systemic and topical corticosteroids are effective treatments for EGE, but are limited by long-term corticosteroid side effects.

CLINICAL PRESENTATION OF EGE

Eosinophilic gastroenteritis (EGE) represents one member within the spectrum of diseases collectively referred to as eosinophilic gastrointestinal disorders (EGIDs), which includes eosinophilic esophagitis (EoE), gastritis, enteritis, and colitis. Although some patients present with EGID limited to the stomach (eosinophilic gastritis, EG) or duodenum (eosinophilic duodenitis), it is often simplest to refer to the combined entity of EGE. EoE as a clinical entity is effectively limited to "solitary EoE"; patients having coexistent EoE and EGE are a small minority. EoE and EGE are closely related disease entities, the relationship of which is discussed below. The diagnosis of EG is confirmed by a characteristic biopsy and/or eosinophilic ascitic fluid in the absence of other causes of gut eosinophilia.

The disease can affect patients of any age, but case series have noted a dominance of presentations starting in the third through fifth decade. As the prevalence of EoE has

Disclosures: The author has no competing financial interests.
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Gastroenterol Clin N Am 43 (2014) 317–327
http://dx.doi.org/10.1016/j.gtc.2014.02.013 **gastro.theclinics.com**
0889-8553/14/$ – see front matter Published by Elsevier Inc.

increased and there is an overall greater appreciation of EGID, it is likely that a second peak of incidence in the first decade of life will become better appreciated. As with EoE, there is a clear male predominance. An electronic survey sent to North American Allergists and Pediatric Gastroenterologists indicate prevalence for EGE of 22 to 28 per 100,000 persons.[1] Although no large longitudinal study has been performed, EGE is largely understood to be a chronic disease with few remissions after the first year.

The clinical features of EGE are protean and are related to the organs, tissue layers affected, and the intensity of eosinophilic inflammation.[2–6] Some patients present with dominant gastric or duodenal disease, whereas others have involvement of both organs. Dominant gastric disease often presents with nausea, vomiting, and early satiety. In contrast, dominant duodenal disease may present with malabsorption and protein-losing enteropathy. Both forms of EGE often have crampy abdominal pain and bloating as additional features. Because jejunal and ileal biopsies are not routinely obtained on endoscopy, it is not known how much these gut segments contribute to disease. Patients can variably present with either diarrhea or constipation.[7]

In addition to the varying distribution of eosinophils along the length of the gastrointestinal (GI) tract, multiple reports have cited EGE subtypes based on differing depth of eosinophilic infiltration.[3,8] The 3 well-described subtypes include dominant involvement of the mucosal, muscularis, and subserosal layers, respectively. Whether these actually represent different diseases or simply different presentations of the same disease is not known. The prevalence of each subtype is unknown because of reporting and referral biases. For example, surgical series report a predominance of muscularis disease with obstruction, whereas medical series primarily describe patients with mucosal involvement. Serosal disease is associated with eosinophilic ascites, but it is not known whether this reflects isolated serosal involvement or simply intense transmural eosinophilic inflammation.

In addition to the common presentations noted above, EGE can present with a variety of unusual manifestations. Patients may have gastric ulcer disease as a feature of their EGE.[9] Typically these ulcers do not respond well to proton pump inhibitor therapy, but do respond to either topical or systemic corticosteroids. There is a case report of ulcer disease responding to an elemental diet.[10]

In contrast to EoE, stricture formation is not a common feature of EGE. That said, a subset of perhaps 5% to 10% of EGE patients do have clinically significant strictures at some point. Such cases will typically present as an acute bowel obstruction with nausea, vomiting, crampy abdominal pain, and bloating.[11] Such bowel obstructions appear to be a combination of both mechanical obstruction due to stricture and functional obstruction due to inflammation, edema, and decreased GI motility. Most of the time these obstructions are reversible with corticosteroid treatment, suggesting that in many cases there is a functional component that can be reversed with treatment.[12] As such, clinically stable EGE patients presenting with bowel obstruction should generally first be treated with parenteral corticosteroid therapy, such as methyl prednisolone 1 to 2 mg/kg/d, and carefully observed.

EGE can occasionally involve the hepatobiliary tree. Pancreatitis is the best described hepatobiliary complication of EGE.[13] It is not clear if this is due to eosinophilic infiltration of the pancreas, gall bladder, or hepatobiliary ducts or to a secondary cause. In addition, EGE can rarely present with eosinophilic cholangitis.[14,15]

PATHOGENESIS OF EGE

The many similarities between EGE and EoE suggest they share a common pathogenesis. Shared features include tissue eosinophilic inflammation, coexisting allergic