Chapter 21
Abdominal Rash and a History of Hirschsprung

Mahboubeh Mansouri

Our patients was a 10-year-old boy, who was consulted due to an eczematous rash in the abdominal region. He had a history of severe constipation since 10 days old. A colostomy had been performed after confirming the diagnosis of Hirschsprung at 6 years old. However soiling and constipation continued after re-anastomosis and pull through surgery. The boy also had multiple episodes of diarrhea and fever, leading to repetitive hospitalizations. As the constipation did not respond to any treatment, an appendicostomy was performed, yet rectorrhagia and bleeding from the site of surgery, nausea and vomiting had persisted then after.

In his past medical history, the boy was diagnosed with asthma. He also had refractory seizures from 3 years old which were irresponsive to a number of anticonvulsive drugs. The boy had developed several episodes of drug allergy reactions as a consequence of antiepileptic drugs as well. All other laboratory findings including autoantibody screening, immunoglobulin levels, stool exams for parasites, as well as all radiological evaluations were normal.

In one colonic mucosal biopsy, ulcerative surfaces and severe infiltration of acute inflammatory cells with eosinophil dominance was reported. His CBC was normal apart from eosinophilia (700/μL) and anemia (Hb: 9 mg/dL). Skin prick test was highly positive for weed and tree pollens and foods like egg yolk, peanut, and soy.

Q1. What is the most probable diagnosis?

A. HIV disease
B. Inflammatory bowel disease
C. Primary immunodeficiency diseases
D. Eosinophilic associated gastrointestinal disorders

M. Mansouri (✉)
Department of Immunology and Allergy, Mofid Children's Hospital, Shahid Beheshti University of Medical Sciences, Tehran, Iran
e-mail: m.mansouri@sbmu.ac.ir

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***Answer**: The correct answer is **D**.*

Eosinophilic gastrointestinal disorders (EGIDs) are defined by abnormal eosinophilic infiltration of different segments of the gastrointestinal tract in the absence of secondary causes, including collagen vascular disease, malignancy, or parasitic infections [1].

Clinical manifestations depend on the involved sites and layers and may include abdominal pain, nausea, vomiting, diarrhea, weight loss, gastrointestinal bleeding, intestinal malabsorption, even ascites or protein loosing enteropathy (PLE) [2]. Anemia with or without apparent gastrointestinal bleeding is an important clinical finding to look for [2].

Q2. All of the below findings are in favor of eosinophilic associated gastrointestinal disorders, except:

A. No response to elimination diets
B. Normal tissue pathology
C. Absence of peripheral eosinophilia
D. Negative skin prick test for food allergens

***Answer**: The correct answer is **A**.*

EGIDs is considered an allergic condition and strongly associates with a higher prevalence of other allergic disorders [1]. Peripheral blood eosinophilia are not necessary for the diagnosis, and negative histological findings cannot exclude the possibility of eosinophilic gastroenteritis. Because eosinophilic infiltration is usually patchy in distribution, biopsy from multiple sites should be obtained.

Q3. What classification of hypersensitivity reactions is the main immunological mechanisms involved in EGID?

A. IgE-mediated reaction (type 1)
B. Antibody-mediated cell cytotoxicity (type 2)
C. Immune complex disease (type 3)
D. Cell-mediated hypersensitivity reactions (type 4)

***Answer**: The correct answer is **D**.*

It is believed that cell-mediated hypersensitivity is the main immunological mechanism involved in EGID, as allergen-specific IgE (sIgE) is usually negative in most patients [2]. There is a strong association of EGID with food allergies, environmental allergies, asthma, and atopic dermatitis [3, 4]. Recently, an empiric diet, preferentially devoid of the six most common food-allergens, milk, soy, egg, wheat, peanuts, tree nuts, shellfish, and fish (six food elimination diet) has been suggested with significant improvement in patients with EGID and efficacy reaching equivalent to topical steroids [5, 6].

Q4. All of the followings are correct treatment, except:

A. Topical budesonide
B. Prednisolone
C. Six foods avoidance
D. Bone marrow transplantation

***Answer**: The correct answer is **D**.*

All the mentioned treatments, except Answer D, are widely accepted in the treatment of EGID [7].

Practical Points

- In contrast to IgE-mediated clinical reaction in food allergy the clinical presentations in Eosinophilic associated gastrointestinal disorders (EGID) are not straight forward and might be misleading
- Clinical symptoms are variable according to the site and layer of infiltration of eosinophils in esophagus, stomach, duodenum or colon or location of eosinophil infiltration in submucosal, muscular or subserosal layers
- The diagnosis of EGID needs a high index of suspicion by the physician
- The diagnosis of EGID should be considered in the presence of any chronic symptoms in the GI tract, with or without atopic diathesis
- Multiple biopsies depending on site of the involvement, must be taken, in spite of normal look mucosa

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