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EOSINOPHILIC GASTROENTERITIS PRESENTING AS PROTEIN – LOSING ENTEROPATHY*
(Case Report)

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SUMMARY: Karademir S, Akçayöz A, Bek K, Tanyel FC, Sungur M, Çelik İ, Ural C, (Dr. Sami Ulus Children's Hospital, Ankara, Turkey). Eosinophilic gastroenteritis presenting as protein-losing enteropathy. Case report. Turk J Pediatr 1995; 37: 45-50.

Eosinophilic gastroenteropathy is an uncommon, idiopathic disease in children that is characterized by eosinophilic inflammation of the intestine. Predominant involvement of the mucosa is associated with diarrhea and less commonly gastrointestinal protein and fat malabsorption. A seven-year-old female was diagnosed with eosinophilic gastroenteritis. This condition was proven by biopsies attained through an endoscope. The most common symptoms were abdominal pain, diarrhea and edema. The patient had no eosinophilia. Her serum immunoglobulin E level was increased (1590 mg/dl). Barium studies revealed mucosal thickening of the antrum, distal jejunum and proximal ileum and prominent mucosal folds of the colon. Ultrasound examination revealed thickening of the colonic wall. The patient was treated with prednisolone (2 mg/kg/day). The symptoms subsided and serum immunoglobulin E decreased to 500 mg/dl 45 days later. The patient is being followed with a small maintenance dose of prednisolone with no relapse. *Key words: eosinophilic gastroenteritis, protein-losing enteropathy.*

Eosinophilic gastroenteropathy (EG) is an uncommon, chronic, relapsing disorder in which eosinophils constitute the predominant cell type in the inflammatory infiltrate of the gastrointestinal (GI) tract[1-3]. Its etiology remains unknown, and whether eosinophils have a role in tissue damage is uncertain[4].

The clinical manifestations, usually recurrent and protean, relate to the site and depth of GI tract involvement[5,6]. Recurrent abdominal pain and diarrhea are the most common presenting symptoms. usually in association with anorexia, nausea or vomiting[3,7]. Protein loss, gastrointestinal bleeding and malabsorption may lead to significant growth delay in the pediatric age-group.

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In this case report we describe a seven-year-old girl who presented with protein-losing enteropathy and was diagnosed with eosinophilic gastroenteropathy.

Case Report

A seven-year-old-girl was admitted to the hospital in April 1993 with a one-year history of upper quadrant abdominal pain and distention accompanied by diarrhea and edema. She had no history of food intolerance, asthma, allergic rhinitis or atopy.

On physical examination, her weight was 15 kg (below the 3rd percentile) and her height was 103 cm (below the 3rd percentile). The blood pressure was 110/60 mm Hg, pulse rate was 100 beats/min, and body temperature was 37 °C. The head and abdominal circumferences were 50 cm and 65 cm, respectively. She had periorbital and lower extremity edema. Her abdomen was distended without ascites (Fig. 1). The liver was palpable 2 cm below the right costal margin, while the spleen was not palpable. Rectal examination was unremarkable, except for a watery brown stool.

On admission, laboratory studies revealed a hemoglobin level of 7.8 g/dl and white blood cell count of 10,000/mm^3 (58% neutrophils, 36% lymphocytes, 4%

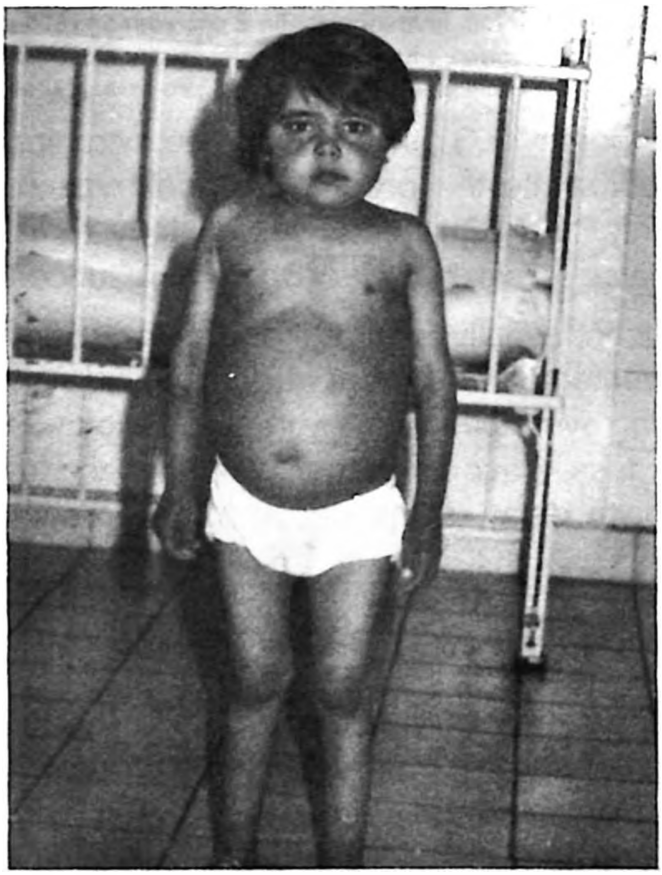

Fig. 1: The photograph taken on admission shows periorbital edema and grossly distended obdomen.

monocytes, 2% eosinophils). Urinalysis was normal. Blood electrolytes, liver and renal function tests, and coagulation studies were within normal limits. Total serum protein and albumin levels were 3.1 g/dl and 1.8 g/dl, respectively. The erythrocyte sedimentation rate was 10 mm/h. Pharynx, urine, stool and blood cultures were all negative. The stool examination was negative for blood, ova and parasites. Stool pH was in the normal range and no sugar was detected in stool samples. Serum iron and iron-binding capacity, vitamin B_{12} and folic acid levels were within normal limits. In immunoelectrophoresis of the serum, the immunoglobulin levels were as follows: IgG 109 mg/dl (normal range: 638 to 1783 mg/dl), IgM 31 mg/dl (normal range: 24 to 98 mg/dl), IgA 24.2 mg/dl (normal range: 89 to 559 mg/dl), and IgE 1590 mg/dl (normal range: 0 to 15 mg/dl). Blood D-xylose concentration was 8.4 mg/dl (normally above 20 mg/dl) after an oral load. The test for HB_sAg was negative. A tuberculin skin test done with 5 TU was negative. The total eosinophil count was 120/mm^3 (normal range: 40 to 400/mm^3).

Ultrasound examination revealed thickening of the colonic wall (8-9 mm), mainly in the rectosigmoid and left flexural regions, and luminal fluid. An upper gastrointestinal x-ray series showed mucosal thickening of the antrum, distal jejunum and proximal ileum. Prominent mucosal folds were noted in the colon on barium enema. A panendoscopy demonstrated erythematous mucosa 7 to 8 cm distal to the splenic flexura.

Histological examination of the specimen from the stomach, duodenum, jejunum and colon showed eosinophilic enteritis predominantly in the mucosa and lamina propria (Fig. 2).

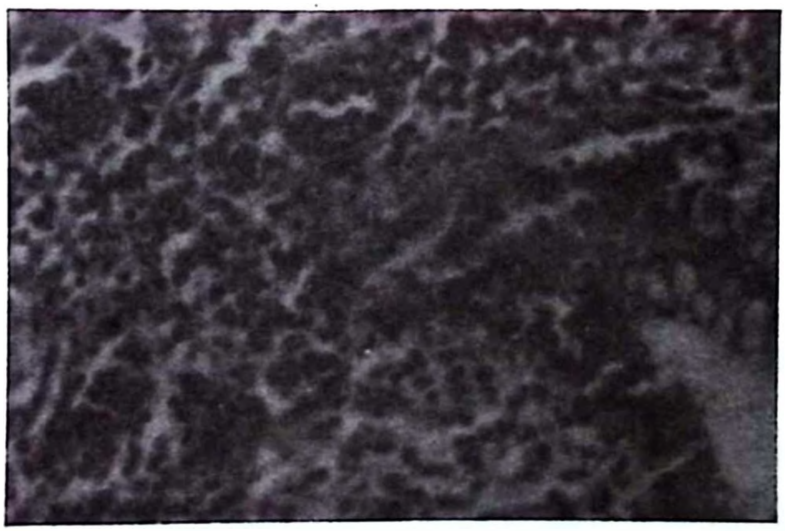

Fig. 2: Colon: lamina propria showing eosinophilic infiltration between the mucosal glands. (H + E, x 400).

During the first two months of hospitalization, the patient was given human albumin 18 times because of severe hypoalbuminemia and edema. She had

findings of intestinal obstruction (vomiting and abdominal distention) five times during this period. After the establishment of the definitive diagnosis of eosinophilic gastroenteritis with typical biopsy findings, the patient was treated with oral prednisolone (2 mg/kg/day). She responded promptly to the therpy. On the tenth day, serum total protein and albumin levels rose to 6.1 g/dl and 3.2 g/dl, respectively. Peripheral edema disappeared. The level of IgE decreased to 500 mg/dl on the 45th day. On the sixth day of the prednisolone therapy, the dose was decreased to 1 mg/kg/day and the patient was discharged. On the follow-up examination performed two months later, her clinical status was good. The abdominal circumference was found to be decreased to 57 cm, with normal levels of serum protein and albumin. The dose of prednisolone was decreased to 5 mg every other day.

Discussion

Eosinophilic gastroenteritis was first described by Kaijser'in 1937[8]. Subsequently, patterns of clinical presentation were correlated with the extent of the eosinophilic infiltration by Ureles et al.[9] and the depth of the eosinophilic reaction by Klein et al.[6] Over 250 cases have been reported in the literature[6-8]. Approximately 15 to 20 percent of the reported cases are in children, and several cases have been documented in early infancy[1,3].

Klein et al.[6] have classified EG according to the layer of gastrointestinal tract involved. Three groups have been described: one with mucosal involvement leading to malabsorption and gastrointestinal protein loss; a second characterized by muscle layer involvement, often leading to pyloric obstruction; and a third with serosal surface infiltration with eosinophils, frequently presenting with ascites. These forms may coexist[5].

In several studies, the most common symptoms were abdominal pain (77-100%), nausea vomiting (48-62.5%), diarrhea (42-62.5%), weight loss (17%), abdominal distention (42%), bloody stool (18%), constipation (5%), ascites (5%) and protein malabsorption (1%). Rarely, fat malabsorption, dysphagia, jaundice, peripheral edema, perianal disease and intestinal perforation could be seen[5-7,10]. In our patient, abdominal pain, vomiting and protein-losing enteropathy dominated the clinical picture. These findings suggested mucosal involvement. Although the biopsy specimen did not include the muscular layer, the findings of intermittent intestinal obstruction suggested involvement of the muscular layer.

Eosinophilic infiltration can be seen over the whole gastrointestinal tract, the most common site being the stomach[7]. With the exception of the esophagus, the frequency of involvement decreases distally through the gastrointestinal tract. Naylor[7] defined it as follows: stomach 43%, ileum 33%, jejunum 31%, duodenum

19%, colon and rectum 11%. The involvement of gallbladder, liver, esophagus, spleen, pancreas or appendix was seen rarely. In our patient, we demonstrated eosinophilic infiltration in the stomach, duodenum, jejunum and predominantly in the colon.

The etiology of EG still remains unknown. The association of systemic allergic symptoms, elevated serum IgE values, and symptomatic response to corticosteroid therapy in some patients suggest that immediate hypersensitivity may be implicated in the pathogenesis of the disease[2].

Hypereosinophilia in peripheral blood is not always present in patients with EG (20-90%) as in our patient[5]. Food sensitivity, especially to milk, appears to play a role in half of these patients[6,7]. Torpier et al.[4] demonstrated that selective release of eosinophil major basic protein from the granule core of eosinophils damaged intestinal tissue. Keshavarzion et al.[11] observed that activated eosinophils were present in tissue biopsies using a monoclonal antibody to eosinophil cationic protein. The cause and meaning of elevated serum IgE in patients with EG are not clear[12,13]. Also, in our patient the level of IgE was elevated although she had no peripheral eosinophilia or allergic symptoms.

Steroids remain the mainstay of therapy for eosinophilic gastroenteritis, with good symptomatic responses being reported[6,7,14]. Similar results were attained in our patient. The value of oral sodium cromoglycate is controversial[13,15] and was not used in our patient.

In conclusion, EG is a rare disease and can be difficult to diagnose. Panendoscopic study may be needed in some patients with unexplained gastrointestinal symptoms.

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