

Eosinophilic Esophagitis among Children at King Hussein Medical Center

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ABSTRACT

Objective: Eosinophilic esophagitis is a clinicopathologic entity characterized by esophageal symptoms in association with a dense eosinophilic infiltrate, the aim of this study is to describe the clinical presentation, laboratory, endoscopic and histopathologic results in children diagnosed with eosinophilic esophagitis.

Methods: This is a retrospective review of the medical records for 38 children with histologic diagnosis of eosinophilic esophagitis performed at King Hussein Medical Center, during the period between January 2001 to February 2011, with the cut off biopsy diagnosis ≥ 20 /HPF eosinophilic infiltrate. Patients were excluded if gastric or duodenal biopsies showed prominent eosinophilic infiltrate. Patients medical records were reviewed regarding age at presentation, gender, clinical presentation, associated disease. Laboratory, endoscopic and histologic results were also reviewed.

Results: A total 38 patients with histological diagnosis of eosinophilic esophagitis were included in this study. Thirty-one (82%) were males and 7 (18%) were females. Their ages ranged between 2-14 years. Mean age 8 years.

The most commonly clinical presentation of eosinophilic esophagitis was vomiting which occurred among 26 children (68%), however, rash was presented in only one child (2%). Peripheral eosinophilia $>(0.5 \times 10^9/L)$ was found in 45%. High serum IgE level (>100 IU/ml) was found in 39%. Positive radio allergeo sorbent testing in 39%. The most frequent endoscopic findings were loss of normal vascularity found in 19 (50%) children, however, white exudates were found in 2(5%) children. Mean eosinophil count was 70 (20-120), eosinophilic degranulation, and basal zone hyperplasia reported in 18 children (47%).

Conclusion: Eosinophilic esophagitis in Jordan displays similar clinical, endoscopic and pathologic features to those described in other countries. Endoscopic and histologic feature remain the gold standard for diagnosis of eosinophilic esophagitis

Key word: clinical presentation, Eosinophilic esophagitis, Endoscopic and histopathologic features.

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Introduction

Eosinophilic esophagitis (EE) is a chronic inflammatory condition that has emerged as a

major cause of esophageal disease over the past decade.⁽¹⁾ It is defined as mucosal inflammation of the esophagus with eosinophils, a specific type

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of leukocyte important to the allergic immune response.⁽²⁾ EE is highly associated with atopic disease and emerging evidence suggests a primary role for food antigen sensitization in the disease etiology.⁽³⁾ It has been well described in the pediatric population, but recently has been increasingly recognized in adult.⁽⁴⁾

Children and adolescents with EE commonly present with signs and symptoms similar to Gastroesophageal Reflux Disease (GERD), which include feeding problems, vomiting, and abdominal pain.^(5,6) However, these patients respond poorly to acid suppression and promotility agents that are the main stays of acid reflux therapy.^(6,7) Because there are no definitive clinical and laboratory findings for EE, endoscopic and histologic evaluation of the esophageal mucosa are currently the only methods for diagnosing EE.⁽²⁾ Management options include dietary modification, pharmacological therapy, and endoscopic dilatation.⁽⁸⁾

This study was conducted to describe the clinical presentation, laboratory, endoscopic and histopathologic features of children diagnosed with EE in King Hussein Medical Center over the last 10 years (2001-2011).

Methods

This is a retrospective review of the medical records for 38 children with histologic diagnosis of EE performed at King Hussein Medical Center (KHMC) during the period between January 2001 to February 2011. Generally during esophagogastroduodenoscopy (EGD) multiple biopsies obtained from the proximal, mid and distal esophagus, stomach and duodenum, those biopsies were taken from the 1-3 esophageal level (distal, mid and proximal) were fixed in 10% formalin, processed routinely, and stained with hematoxylin and eosin. Those esophageal biopsies were reviewed by histopathologist and the eosinophils counted where they appeared most numerous in the biopsy. We consider a cut off ≥ 20 eosinophils per high power field (HPF) as diagnostic of EE, patients were excluded if gastric or duodenal biopsies showed a prominent eosinophilic infiltrate.

Patients medical records were reviewed regarding age at presentation, gender, associated disease, clinical presentation, allergic history.

Their laboratory profile including peripheral eosinophilia, food panel test (radio allerge sorbent testing, RAST), serum IgE level. Their endoscopic and pathologic result were also described. Simple descriptive statistics using frequency and percentage were used to describe the study variables.

Results

A total of 38 children were diagnosed as EE. Thirty-one (82%) was males and 7 (18%) were females. Their ages ranged between 2-14 years. Mean age 8 years.

Table I shows clinical presentation and results among the study group. The commonest clinical presentation was vomiting which occurred among 26 (68%) children followed by dysphagia 25 (66%), failure to thrive 12 (32%), abdominal pain 10 (26%), wheezes 9 (32%), acute food impaction 5 (13%), rash 1 (2%) respectively. Three patients known to have celiac disease including one patient with Insulin Dependent Diabetes Mellitus (IDDM), 9 (23%) patients known to have Bronchial asthma on regular treatment, 2 patients with epilepsy on anti epileptic drugs, and one patient with a severe atopic dermatitis and hyper IgE. Peripheral eosinophilia $> (0.5 \times 10^9/L)$ was found in 45%, high serum IgE level (>100 IU/ml) was found in 39%. Positive RAST test in 39% (allergy to a single or multiple food including nut, soya, peanut, eggs, cow milk, wheat, maize, lamb, casein).

Table II shows endoscopic and histopathologic features among the study group. The most frequent endoscopic findings were loss of normal vascularity in 19 (50%) children, esophageal erythema and increased friability in 16 (42%), linear furrow 14 (37%), multiple rings 5 (13%), white exudate 2 (5%), which often presumed to have Candida esophagitis, but this was excluded by negative microbiologic test, esophageal narrowing seen in 3 children (7%) respectively. Seven children (18%) had normal endoscopy.

Mean eosinophil count was 70 (20-120), eosinophilic degranulation reported in 18 children (47%), also basal zone hyperplasia seen in 18 (47%) children, microabscess (considered as an aggregate of > 4 eosinophils) 2 (7%) children, epithelial fibrosis; 1(2%) and Eosinophilic infiltrate seen in Fig. 1.

Table I: Clinical presentation and results of the study group

Clinical presentation*	Number of patients	%*
Vomiting	26	68
Dysphagia	25	66
Failure to thrive	12	32
Abdominal pain	10	26
Wheezes	9	23
Acute bolus obstruction	5	13
Rash	1	2

*Totals do not add to 100% as some patients may have more than one symptom.

Table II: Endoscopic, histologic features among the study group.

Features	Number of patients	%
Loss of normal vascularity	19	50
Esophageal erythema and increased friability	16	42
Linear furrow	14	37
Multiple rings	5	13
Esophageal narrowing	3	7
White exudate	2	5
Normal endoscopy	7	18
Eosinophilic degranulation	18	47
Basal zone hyperplasia	18	47
Microabcess	2	7
Epithelial fibrosis	1	2

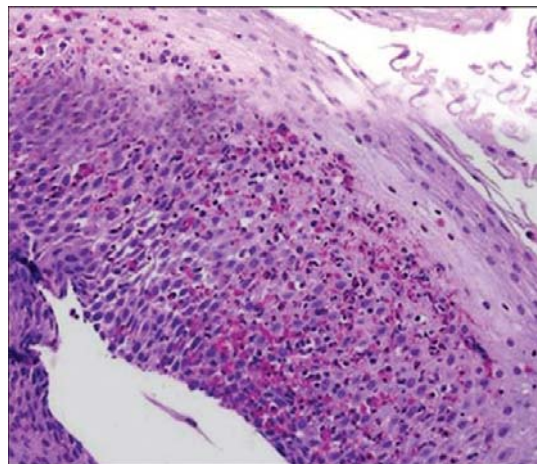


Fig. 1: The esophageal infiltration with eosinophils and eosinophil granules, which was present among the study group

The treatment of EE used in our patients was specific food avoidance, 2 weeks treatment with systemic steroid (oral prednisolon) in a dose of 1mg/kg/day. Then 6 months treatment with swallowed inhaled corticosteroid.

Discussion

EE is of growing interest for pediatricians and allergists.⁽⁹⁾ EE has been reported throughout the life span, from patients under 1 year of age to those in their ninth decade.⁽¹⁰⁾ Few studies have been conducted in our Arab area.^(11,12) Al-Hussian *et al.*,⁽¹¹⁾ reported a case showed Saudi child with esophageal trachealization as a feature

of EE. EE is defined by infiltration of the esophagus with eosinophils without infiltration in other part of gastrointestinal tract, it should be diagnosed when clinical symptoms of esophageal dysfunction are present, ≥ 15 eosinophils in one high power field [HPF] are detected in one or more esophageal biopsies, and high dose PPI fails to ameliorate the symptoms or normal PH monitoring of the distal esophagus.⁽¹⁾ As reported in other series from the different countries,⁽¹²⁻¹⁵⁾ we found a male predominance for EE. Majority of those patients were atopic, ranging 33-70% in different studies^(12,14,16-18) which approximate our study result.

Previous reports show that EE is primarily diagnosed in school-age children, predominantly adolescent male,⁽¹⁹⁾ result of this study suggest that EE should be considered in the differential diagnosis of children as early as two years of age presenting with vomiting and poor weight gain. Symptoms of EE in infants and young children usually mimic gastro esophageal reflux disease (GERD) but are unresponsive to antireflux management and responsive to dietary restriction. In older children, dysphagia and acute food impaction is the usual presenting symptoms, in our study vomiting and dysphagia almost have the same equal presentation occurring in (68%,66%) respectively, followed by failure to thrive and abdominal pain, however, other study⁽¹²⁾ showed that failure to thrive and abdominal pain were the commonest clinical presentation accounting for 86%,53% respectively.

Of interest three of our patients with EE was known to have celiac disease based upon small bowel biopsies, laboratory findings, and was on gluten free diet, which was mentioned as case report with IgE mediated allergy.⁽²⁰⁾ On endoscopy, the typical findings of EE include linear furrow, esophageal erythema, the inflamed epithelium looks dull and thickened, and the usual submucosal vascular pattern is obscured from endoscopist's view. Mucosal rings may be present in long standing disease, but are not common in young children,⁽²¹⁾ it appeared in 5 (13%) children in our study, while other study^(12,13) reported it in 46%, 12% respectively. White exudate and esophageal stricture also present in our study, however the esophagus might appear endoscopically normal in up to 20% of cases,^(1,22) which approximate our study results, however, other studies^(12,13) reported it in 13%,12% respectively.

The number of intra epithelial eosinophils in esophageal biopsies specimens is the main diagnostic criteria of EE, other common histopathologic features observed in our study included eosinophilic degranulation which was found in 47% compared to other studies^(12,13) reported it in 86%, 50% respectively, basal zone hyperplasia found in 47% and eosinophil micro abscess which mainly located at the laminal edge of the epithelium was found in 7% compared to other studies^(12,13) which reported it in 73%,

100% respectively. Studies on adult and children demonstrate fibrosis of the lamina propria in EE patients which may be a distinguishing feature of EE,^(13,23,24) it was demonstrated in only one case in our study.

Conclusion

EE in Jordan displays similar clinical, endoscopic and pathologic features to those described in other countries. Endoscopic and histologic feature remain the gold standard for diagnosis of EE.

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