

Lymphocytic esophagitis mimicking eosinophilic esophagitis

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Abstract

A 74-year-old male with a history of dysphagia for 3 years presented with acute food impaction. Endoscopy showed a tight distal stricture with coarse rings at the middle third of the esophagus. Biopsies taken from the middle third of the esophagus showed marked infiltration of the intraepithelial lymphocytes mainly in a peripapillary distribution. The immunostains showed presence of CD3 and CD5 lymphocytes (T cell markers) in the epithelium. Lymphocytic esophagitis is a histologic phenotype of esophagitis diagnosed by marked esophageal lymphocytosis mostly in a peripapillary distribution with no or only rare intraepithelial granulocytes and presenting similar to eosinophilic esophagitis with dysphagia and esophageal rings.

Keywords lymphocytic esophagitis, eosinophilic esophagitis, dysphagia

Ann Gastroenterol 2012; 25 (4): 355-357

Introduction

A novel histologic subset of chronic esophagitis, called lymphocytic esophagitis, has been recently reported. Lymphocytic esophagitis is a histologic phenotype of esophagitis characterized by high numbers of intraepithelial lymphocytes gathered mainly in a peripapillary distribution and by none or occasional intraepithelial granulocytes. Rubio *et al* was the first to describe lymphocytic esophagitis as an independent entity. In his study he found high numbers of intraepithelial lymphocytes (IELs) in the peripapillary fields in esophageal biopsy specimens of patients with lymphocytic esophagitis [1]. Much lower numbers of IELs were found in the interpapillary fields. This contrasted with the distribution of IELs in reflux, radiation, and *Candida albicans* esophagitis in his study; in such cases the number of interpapillary IELs highly exceeded that recorded in peripapillary areas. The true causes for this apparently site-related chronic mucosal inflammation remain unknown. We present a case of a 74 year old man having

recurrent dysphagia who presented with acute food impaction with endoscopy showing rings in the esophagus with high number of intraepithelial lymphocytes in the peripapillary fields in the biopsy. Thus the clinical and endoscopic findings were mimicking that of eosinophilic esophagitis.

Case report

A 74-year-old male with a history of 3 years duration of dysphagia presented with acute food impaction. The patient had a history of lymphoma and esophagitis with stricture

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Conflicts of Interest: None

Funding Sources: Thomas Jefferson University Hospital Department of Gastroenterology and Hepatology

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Acknowledgement: The histopathology images were graciously provided by Juan Palazzo, MD, and Ping Gong, MD, Department of Pathology, Thomas Jefferson University Hospital, Philadelphia

Received 28 April 2012; accepted 17 May 2012

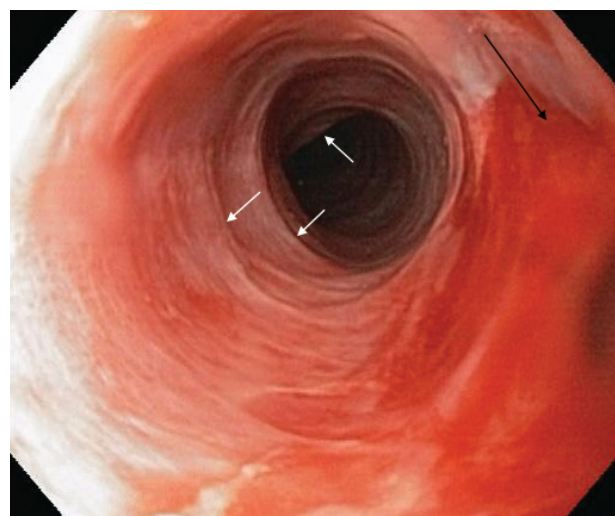


Figure 1 Upper endoscopy showing rings in the middle third of the esophagus (white arrow). Black arrow shows mucosal split at the lower end during endoscopy

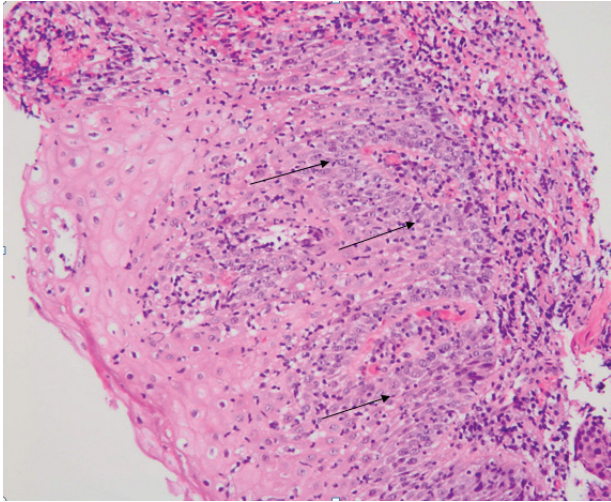


Figure 2 HE stains showing infiltration of large number of intraepithelial lymphocytes in the peripapillary area without or rare granulocytes

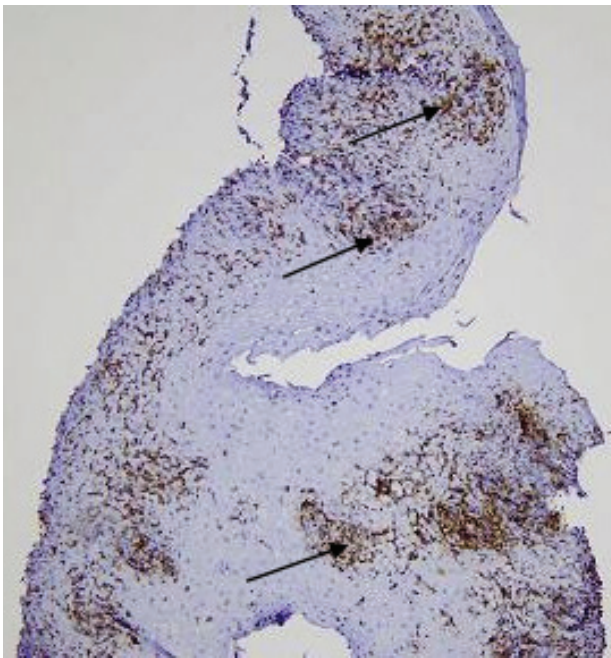


Figure 3 Immunostains showing presence of CD3 cells (T cells)

without the symptoms of gastro-esophageal reflux disease (GERD). Following endoscopic removal of the impacted meat he had severe chest pain with evidence of perforation of the esophagus with pneumomediastinum on chest CT scan which resolved uneventfully. A later endoscopy showed a tight distal stricture and coarse rings at the middle third of the esophagus (Fig. 1). A mucosal split occurred during endoscopy (Fig. 1, black arrow). Biopsies taken from the

middle third of the esophagus showed marked infiltration of exclusive intraepithelial lymphocytes in a peripapillary distribution (Fig. 2). There was no eosinophilic component, thus ruling out eosinophilic esophagitis.

The immunostains showed presence of CD3 and CD5 lymphocytes (T cell markers) in the epithelium (Fig. 3). The specimen was negative for CD 20 lymphocytes (B cell marker). The esophageal manometry showed diffuse esophageal spasm with partial relaxation of the lower esophageal sphincter. The patient had prior chemotherapy for non-Hodgkin lymphoma present in his nostril and leg. The lymphoma was monoclonal B-cell CD20 positive. Thus, the lymphocytes (CD3 and CD5) present in the esophageal epithelium are distinct from the lymphoma (CD20). Four year follow up with 5 endoscopies shows persistence of the endoscopic and biopsy findings. The patient was dilated each time and received botox twice.

Discussion

Lymphocytic esophagitis is a histologic phenotype of esophagitis characterized by high numbers of intraepithelial lymphocytes gathered mainly in a peripapillary distribution and by none or occasional intraepithelial granulocytes [1,2]. The minimum number of lymphocytes per high power field necessary to establish the diagnosis of lymphocytic esophagitis has not been defined. Intraepithelial lymphocytosis of the esophagus has been studied to a limited extent in the context of specific diseases mainly in the pathology literature. Rubio *et al* was the first to describe lymphocytic esophagitis as an independent entity [1]. He concluded that the cause of this apparently site related chronic mucosal inflammation remains unknown. The presence of a high number of intraepithelial lymphocytes especially peripapillary in location along with the absence or presence of occasional granulocytes is an important criterion distinguishing this condition from other causes of esophagitis. In a recent pathology database from a group of gastrointestinal pathologists, lymphocytic esophagitis was seen in 119 patients out of 129,252 patients while 3745 patients had eosinophilic esophagitis and 40,665 had normal mucosa [3]. Dysphagia was as common in the patients having lymphocytic esophagitis as those in eosinophilic esophagitis (53% vs. 63%, not significant). Endoscopic findings of rings in the esophagus were seen in lymphocytic esophagitis and eosinophilic esophagitis [3].

In the present patient, endoscopic features, rings, mucosal fragility, mucosal splitting and perforation are similar to those in eosinophilic esophagitis. We believe that lymphocytic esophagitis is an emerging condition. It may be under-reported by pathologists and under-recognized by gastroenterologists and confused with eosinophilic esophagitis. It may be considered in the evaluation of chronic dysphagia. There are no published studies on therapy for lymphocytic esophagitis.

References

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