

Case Report

Eosinophilic Esophagitis Presenting as Complete Esophageal Desquamation: An Unusual Case of Chest Pain

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The diagnosis of eosinophilic esophagitis has been steadily increasing with the increase usage of endoscopy as a diagnostic tool. Here we present a case of complete esophageal desquamation visualized on endoscopy without any evidence of caustic ingestion or any other potential disease process that would cause a similar presentation. The diagnosis of eosinophilic esophagitis was established by significantly increased intraepithelial eosinophils, eosinophilic micro-abscess and partially detached squamous epithelium on the esophageal biopsy. There was complete resolution of symptoms with standard therapy for eosinophilic esophagitis. To the best our knowledge, this is the first reported case in the English literature of eosinophilic esophagitis that presents as complete esophageal desquamation.

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Key Words: eosinophilic esophagitis, desquamative esophagitis

INTRODUCTION

The incidence of Eosinophilic Esophagitis (EoE) has been steadily increasing since it was first described in the late 1970's. It is defined as "a chronic, immune/antigen-mediated, esophageal disease characterized clinically by symptoms related to esophageal dysfunction and histologically by eosinophil-predominant inflammation."¹ Clinically these patients present with a variety of symptoms including dysphagia, heart burn/chest pain and food impactions.² They would usually undergo an esophagogastroduodenoscopy (EGD) as the next step in their workup as they do not respond to empiric treatment with proton pump inhibitors (PPI). Typically on endoscopy one might see one or multiple of the following morphological features which include rings, linear furrowing, white papules, strictures, attenuation of the subepithelial vascular pattern and a small caliber esophagus. These endoscopic findings alone are not sufficient to make a diagnosis of EoE. To confirm a diagnosis of EoE, biopsies must be taken and show at least 15 eosinophils per high power field. We describe a case with symptoms and histological confirmation of EoE which presents a unique endoscopic finding mimicking desquamative esophagus. The findings are consistent with the endoscopic appearance of Esophagitis dissecans superficialis (EDS).

CASE REPORT

The patient is a 43 year old female with a history of depression who presented for epigastric/low chest pain and

reduced appetite which started 3 days prior to her reduced presentation. The pain was described as a dull discomfort which was non-radiating, but worsened upon eating and deep inspiration. She complained of odynophagia without any complaints of dysphagia. She had a similar presentation 4 months earlier which lasted 4-5 days and spontaneously resolved without any intervention. She additionally denied nausea, vomiting and melena. However, she did endorse a decrease in appetite. Her physical exam was unremarkable. Due to the severity of her symptoms and with a decrease in appetite an EGD was performed. The esophagus showed a complete desquamation of the esophageal mucosa as well as linear furrowing and rings. (**Figure 1**) Additionally, her biopsy obtained from proximal, mid and distal esophagi showed eosinophilic abscesses (**Figure 2**) and 15 eosinophils/High power field (HPF, 400x) on average, ranging from 2 eosinophils/HPF to 40 eosinophils/HPF (**Figures 3, Figure 4**) which confirmed the diagnosis of Eosinophilic Esophagitis. The biopsy material was grossly white-tan, and soft. The patient was started on 4 mg of methylprednisolone which improved the complaints of odynophagia. The patient was eventually transitioned to swallowed fluticasone for maintenance therapy. At her 2-month follow-up, the patient responded well to the treatments, with no symptoms or signs, and unremarkable endoscopic findings.

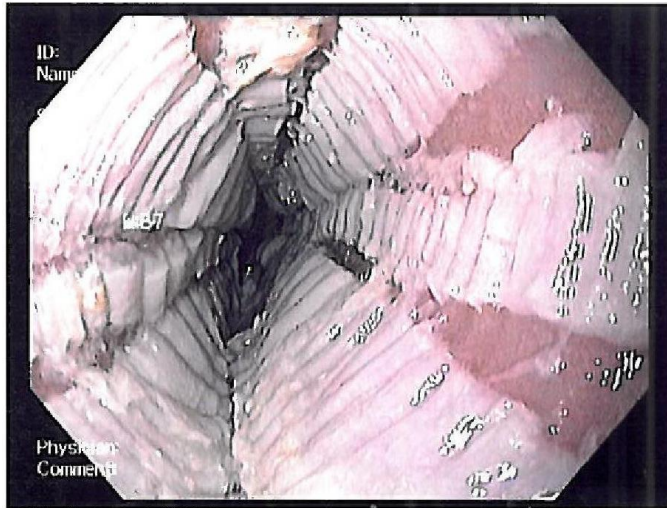
DISCUSSION

Although the diagnosis of EoE is fairly common the endoscopic appearance of a completely desquamative process is indeed a new and undocumented presentation of EoE. Upon literature review we found that the clinical presentation and endoscopic findings to be consistent with Esophagitis

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dissecans superficialis (EDS), an endoscopic finding that is described as sloughing of large fragment of esophageal squamous mucosa.³ Although the pathogenesis of EDS is unknown it has been associated with certain medications (Bisphosphonates, NSAIDs and Potassium Chloride), irritants ranging from hot beverages to corrosive irritants,



Figures 1. Endoscopic view of the esophagus showing complete desquamation of the esophageal mucosa as well as linear furrowing and rings.

Finally certain dermatological conditions with esophageal involvement must be excluded that have similar morphological appearance on EGD such as Pemphigus vulgaris^{4,5} and Lichen planus.^{6,7} Eosinophilic micro-abscess and the presence of intraepithelial eosinophilia in the mid esophagus are consistent with EoE. Clinically the patient did not have systemic pemphigus and endoscopically the lesions involved the whole esophagus while pemphigus is typically more focal in nature.⁸ Additionally pemphigus endoscopically and pathologically also shows bullae and

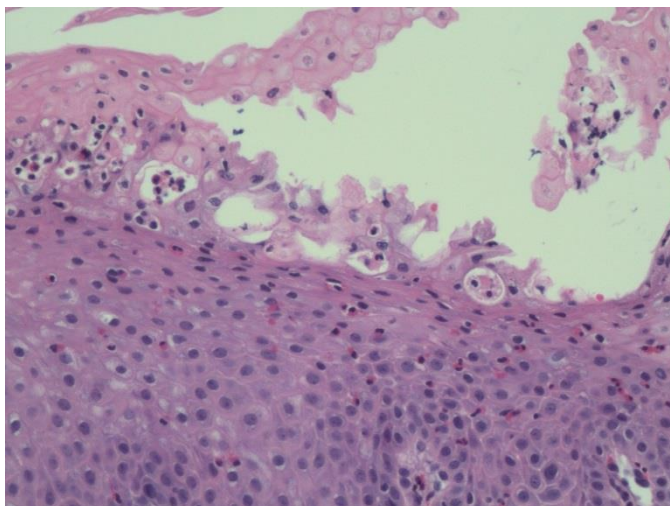


Figure 3. Esophageal biopsy at 200 x with H&E stain, showing desquamation, focal eosinophilic abscess and increased eosinophils in the mid esophagus.

collagen vascular disorders and celiac disease. In the case presented the patient was not on any medications that may cause EDS or was there any history of ingestion of any potential irritants. Additionally she did not have any history suggestive of collagen vascular disorder or celiac disease.

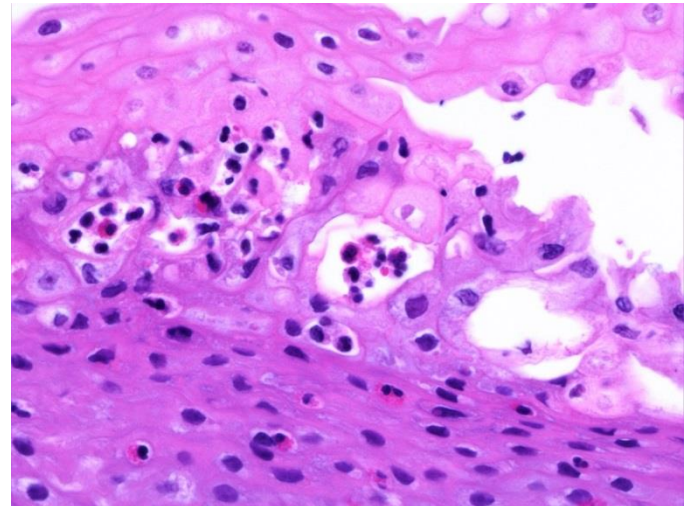


Figure 2. Eosinophilic micro-abscess in the biopsy (400x).

exfoliated erosions.⁵ On the other hand, lichen planus has characteristic band-like or lichenoid lymphocytic infiltrate and elongated rete pegs, which this case also lacks.⁵ These disease processes with esophageal involvement have been described as being a separate entity from EDS. However, a recent study reported that these dermatologic disorders and others with esophageal involvement may be associated with EDS.⁹ Ultimately the patient presented here did not have any dermatological complaint or symptoms that would lead to any diagnosis with a dermatological cause.

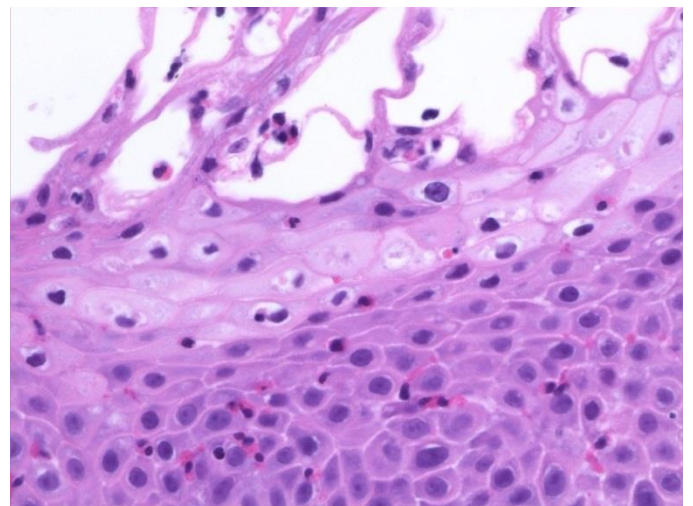


Figure 4. Esophageal biopsy at 400x with H&E stain showing 2-40 eosinophils/HPF (Note: the micrograph only illustrates the cropped central-area of the high power field, with only 21 of the 40 eosinophils shown here).

In conclusion, this case to our best knowledge is the first documented EoE with EDS endoscopic presentation. Although EDS has been reported in association with certain medications and esophageal strictures, the definitive etiology is still unknown. The significance of this case report is that clinicians should keep in mind to include the diagnosis of EoE in the differential diagnoses of EDS. Given the increasing diagnosis of EoE and the more frequent use of EGD as a tool for diagnosing numerous upper gastrointestinal pathologies, our case may represent the first of many future presentations of EDS with a confirmed diagnosis of EoE. Our patient had an excellent response to steroid treatment. Our report also suggests that consideration may be given to the cases with similar endoscopic characteristics and biopsies, ideally from proximal, mid and distal esophagi, must be taken for pathological confirmation.

CONFLICT OF INTEREST

The authors have no conflict of interest to disclose.

REFERENCES

1. Liacouras CA, Furtua GT, Hirano I, et al. Eosinophilic esophagitis: updated consensus recommendations for children and adults. *J Allergy Clin Immunol*. 2011;128(1):3-20.
2. Shahzad G, Mustacchia P, Frieri M. Role of mucosal inflammation in eosinophilic esophagitis: Review of the literature. *ISRN Gastroenterology*. 2011;2011:468073.
3. Matunaga Y, Goto A, Fujii K, et al. Desquamative esophagitis due to pemphigus vulgaris. *Endoscopy*. 2010;2(7):252-256.
4. Fukuchi M, Otake S, Naitoh H, et al. A case of exfoliative esophagitis with pemphigus vulgaris. *Dis Esophagus*. 2011;24(3):23-25.
5. Chandan VS, Murray JA, Abraham SC. Esophageal lichen planus. *Arch Pathol Lab Med*. 2008;132(6):1026-1029.
6. Westbrook R, Riley S. Esophageal Lichen Planus: Case Report and Literature Review. *Dysphagia*. 2008;23(3):331-334.
7. Khamays I, Eliakim R. Esophageal pemphigus vulgaris: a rare manifestation revisited. *Gastroenterol Hepatol (NY)*. 2008;4(1):71-72.
8. Kanbay M, Selcuk H, Gur G, et al. Involvement of the esophagus in a patient with pemphigus vulgaris who was on immunosuppressive therapy. *J Natl Med Assoc*. 2006;98(8):1369-1370.
9. Hokama A, Yamamoto Y, Taira K, et al. Esophagitis dissecans superficialis and autoimmune bullous dermatoses: A review. *World J Gastrointest Endosc*. 2010;2(7):252-256.