

Esophagitis Dissecans Superficialis - A Rare Cause of Dysphagia

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DOI: <http://dx.doi.org/10.21276/ijcmsr.2019.4.4.43>

How to cite this article: Abhijith Menon, Shivaprasad B, Uday B Nayak. Esophagitis dissecans superficialis - a rare cause of dysphagia. *International Journal of Contemporary Medicine Surgery and Radiology*. 2019;4(4):D182-D184.

A B S T R A C T

Introduction: Esophagitis dissecans superficialis [EDS] or sloughing esophagitis is a rare benign condition of the esophagus, characterised by desquamation of superficial epithelium of the esophageal lining. Only less than 200 cases of EDS are reported all over the world. We present a case of EDS, who was admitted with complaint of difficulty in swallowing since 3 months.

Case report: A 46 years old male patient was admitted with complaints of dysphagia since 3 months. Esophagogastroduodenoscopy with biopsy was performed and he was diagnosed to have esophagitis dissecans superficialis. Patient improved with proton pump inhibitors and oral steroids. Thymoma diagnosed along with it was surgically resected.

Conclusion: Esophagitis dissecans superficialis is a rare and under diagnosed entity which should be considered during evaluation of dysphagia. It usually resolves without lasting esophageal pathology with treatment. Oral steroids should be added in patients who are refractory to treatment with proton pump inhibitors alone.

Keywords: Esophagitis Dissecans Superficialis, Sloughing Esophagitis, Desquamated Esophageal Mucosa, Thymoma, Steroids

INTRODUCTION

Esophagitis dissecans superficialis [EDS] or sloughing esophagitis is a rare endoscopic finding characterized by sloughing of large fragments of the esophageal squamous mucosa that may be coughed up or vomited.¹ Etiology is unclear, considered idiopathic, but found incidentally with conditions like thymoma during evaluation. It is also found in association with bullous diseases like pemphigus vulgaris² and use of drugs like bisphosphonates. Most common site is distal esophagus.³ Most common presentation is chronic dysphagia due to localised esophageal strictures.⁴ Asymptomatic cases have also been reported.

CASE REPORT

A 46 year-old man with no known comorbidities was admitted in the hospital with complaint of difficulty in swallowing since 3 months. It started initially with solid foods, but later he found difficulty in swallowing liquids also. It was accompanied by retrosternal chest pain after swallowing food which lasted for 5 to 10 minutes. History of weight loss of around 10 kgs was present. He was evaluated in the nearby hospital and treated symptomatically. But symptoms did not subside. General physical examination and systemic examination did not reveal any significant findings. Blood investigations were normal. Barium swallow was done which showed esophageal diverticula at the level of T1 vertebrae. Oesophagogastroduodenoscopy was done

which revealed friable esophageal mucosa with mucosal peeling on mild trauma. Esophagus had plaque like whitish material with vertical and circumferential fissures or cracks. Biopsy was taken from the esophagus. Histopathological examination showed strips of stratified squamous epithelium with parakeratosis and hyperplastic basal layer. There were multiple transverse splits at different levels in the acanthotic layer. Parakeratotic layer was separated from the underlying acanthotic layer. It was suggestive of esophagitis dissecans superficialis.

Treatment was started with proton pump inhibitors. Pantoprazole 40 mg once daily was started and he was advised to review after 3 weeks. But the symptoms did not subside. So oral steroid was started. Prednisolone was given 1 mg/kg [50 mg OD] along with PPIs for 2 weeks which was later tapered and stopped. His symptoms improved dramatically with treatment and follow-up esophagogastroduodenoscopy after 3 months showed complete normalisation of esophageal mucosa.

Later the patient was readmitted with similar complaints after 8 months. Prednisolone was restarted and patient became symptomatically better again. Esophagogastroduodenoscopy and esophageal dilatation was done as stricture was present in the distal esophagus. In view of recurrent cough, chest x-ray and later CECT-Chest was done which was suggestive of thymoma. It was surgically resected later.

Fig-1 shows erythematous esophagus with easy peeling of

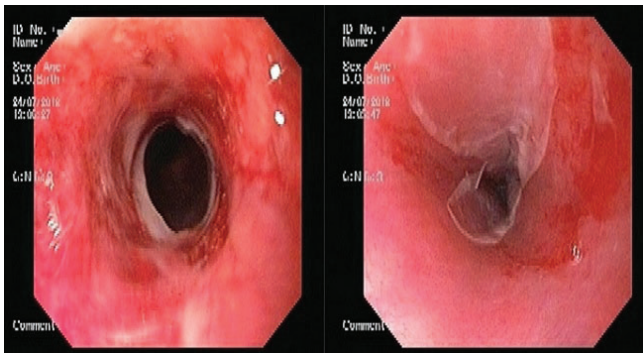


Figure-1: Esophagoduodenoscopy

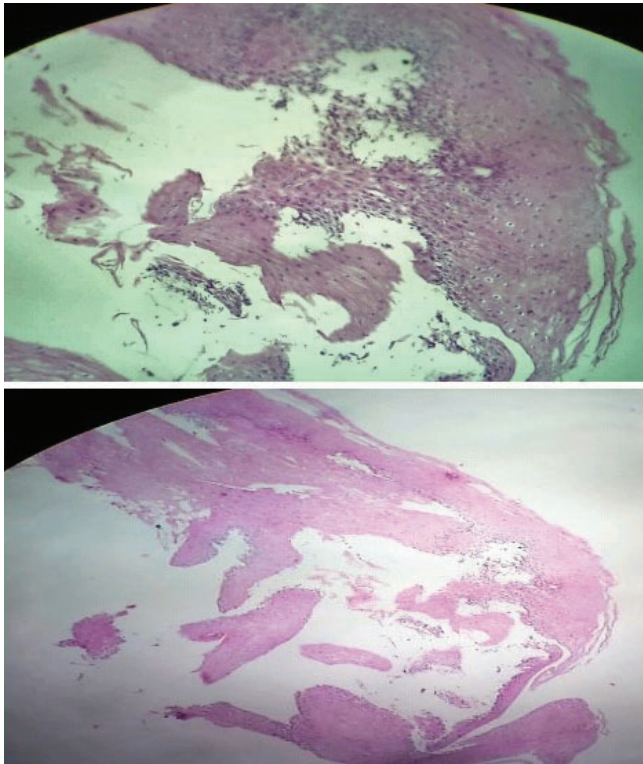


Figure-2: Histopathological examination

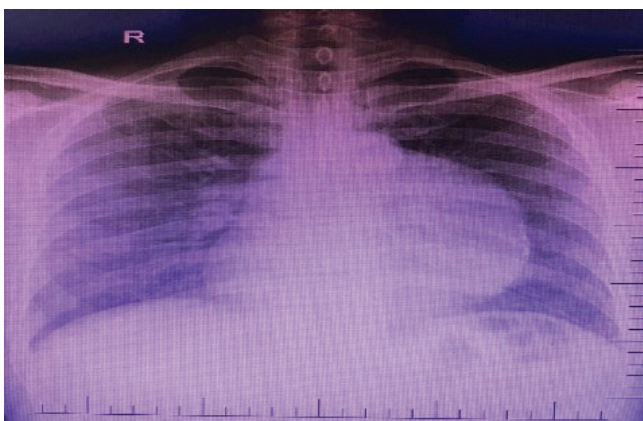


Figure-3: Chest X-ray

mucosa, seen starting from the post cricoid region with plaque like whitish material. Rest of the esophagus also showed mucosa peeling off easily. Fig-2 shows strips of stratified squamous epithelium with parakeratosis and hyperplastic basal layer. Multiple transverse splits at different levels in the

acanthotic layer. Fig-3 shows chest X-Ray of same patient after 8 months showing mediastinal widening. CECT Chest was done later which was suggestive of thymoma

DISCUSSION

Esophagitis dissecans superficialis [EDS] or sloughing esophagitis was first reported in 1892 by Rosenberg.⁵ It is a rare benign condition characterised by desquamation of superficial epithelium of the esophageal lining. There will be strips of detached and sloughed squamous tissue with normal underlying mucosa, associated with minimal or no bleeding. Males and females are equally affected. Only less than 200 cases of EDS are reported all over the world.

Most common site is distal esophagus³ [42%]. Etiology is unclear, considered idiopathic. Based on the reported cases, EDS is thought to be associated with use of drugs like NSAIDs, bisphosphonates, CNS depressants⁶, consumption of hot beverages, heavy smoking, mediastinal radiation, endoscopic therapy [sclerotherapy, esophageal dilatation and band ligation], bullous diseases^{2,8} and thymoma.⁹ As it is associated with bullous diseases like pemphigus vulgaris^{2,8}, some consider it as an autoimmune condition. Ponsot et al study in 1996 showed that many patients of EDS presented with thymoma⁹, which is present in our patient also.

Clinically it might be found incidentally with other conditions like thymoma, bullous diseases etc during evaluation. Most common presentation is chronic dysphagia [32%] due to localised esophageal strictures⁴, followed by vomiting out of casts of esophageal mucosa [23%] and abdominal pain [23%]. Chronic odynophagia localised to the upper sternum, weight loss and hematemesis may be also seen

Diagnosis is usually by UGI Scopy followed by esophageal biopsy and histopathological examination to confirm the diagnosis. In a recent review article, Hart et al proposed three endoscopic criteria for sloughing esophagitis: strips of sloughed mucosa measuring greater than 2 cm, normal underlying mucosa, and lack of ulceration in adjacent tissue.¹⁰ Despite the dramatic appearance of EDS, complete resolution of mucosal injury is usually achieved without any long lasting clinical consequences through combination of oral steroids and proton pump inhibitors. A combination of acid suppression and the discontinuation of precipitating medications has been reported to result in the healing of some cases of EDS.¹ In some cases, esophageal dilatation is done as strictures are present.

CONCLUSION

Esophagitis dissecans superficialis is a rare and under diagnosed entity which should be considered during evaluation of dysphagia. It usually resolves without lasting esophageal pathology with treatment. Any precipitating factors if present, should be avoided. Oral steroids should be added in patients who are refractory to treatment with proton pump inhibitors alone. Clinical examination and relevant investigations should be performed to rule out associated conditions like bullous diseases, thymoma etc

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Source of Support: Nil; **Conflict of Interest:** None

Submitted: 22-11-2019; **Accepted:** 02-12-2019; **Published online:** 26-12-2019