

# Esophageal Stricture Recalcitrant to Repeated Dilation in Complex Rheumatological and Bullous Disease

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## INTRODUCTION

Many rheumatological diseases have been well characterized to affect the esophagus. Esophageal involvement in autoimmune bullous disease is rare and almost always occurs with severe cutaneous symptoms. We report a case of severe esophageal strictures recalcitrant to repeated dilatation in a patient with complex rheumatological disease and isolated esophageal bullous disease.

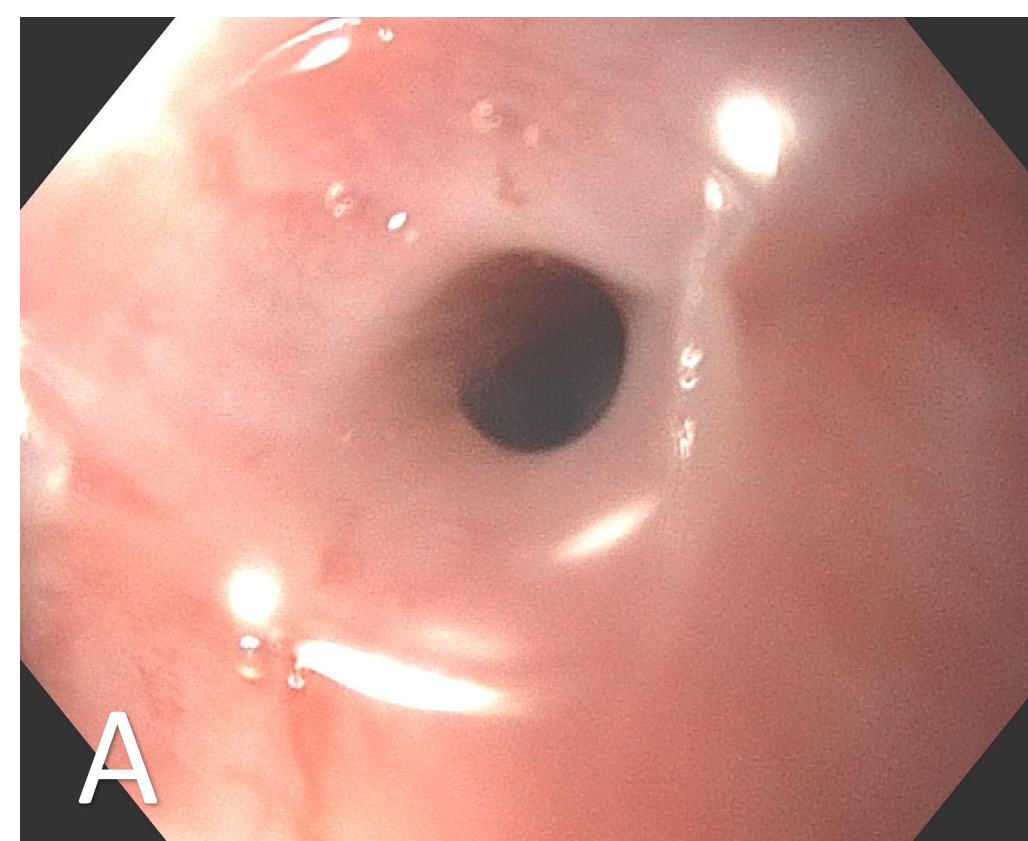
## CASE REPORT

- A 50-year-old female presented to the hospital with complete inability to tolerate oral intake for several weeks.
- Past history included Sjogren's syndrome and esophageal strictures with 11 previous dilatations over two years.
- Physical exam showed poor skin turgor, dry mucous membranes, and skin hyperpigmentation on her hands and forearms.
- Esophagram and EGD showed a proximal esophageal stricture which was dilated using a push style dilator to 14mm. The stricture recurred, requiring repeated dilatation and placement of a percutaneous gastrojejunostomy.
- Once oral intake was tolerated, she was discharged with outpatient follow-up.

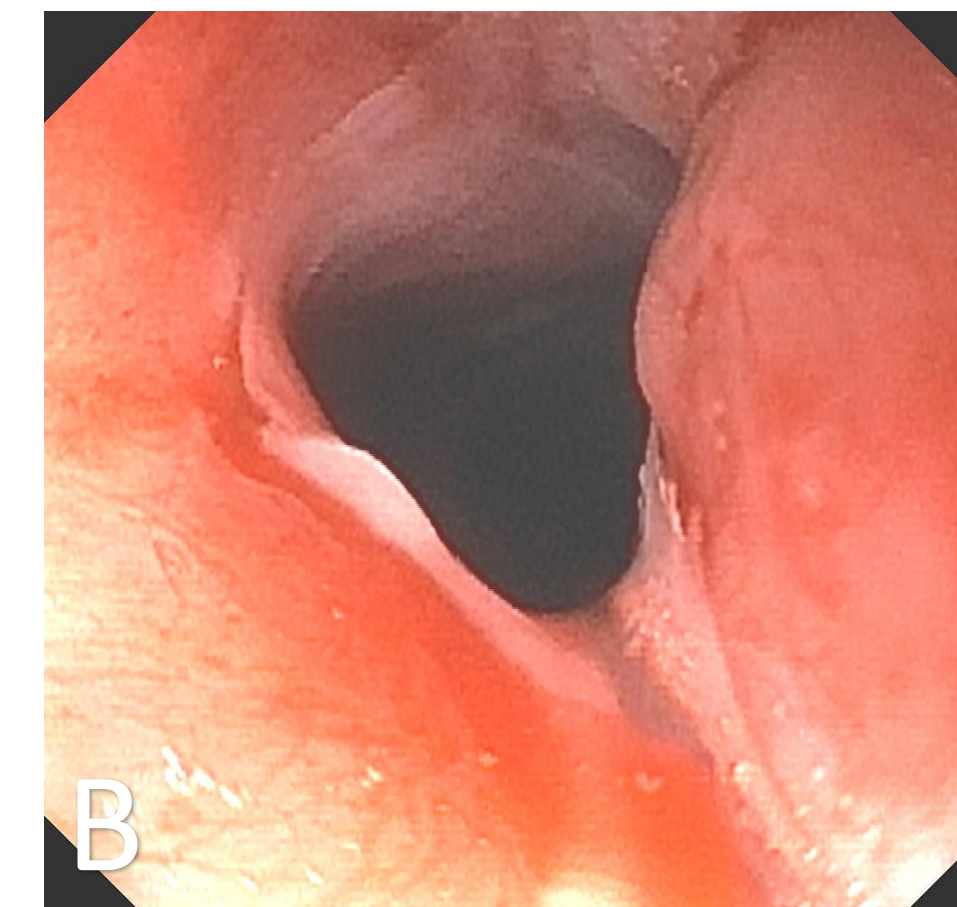
## CASE REPORT

- Subsequent outpatient rheumatological workup showed antibodies to ANA, RNP, SSA, CCP, and Smith indicating mixed connective tissue disease with features of systemic lupus erythematosus, rheumatoid arthritis, and scleroderma.
- She required multiple dilatations over several months and was started on mycophenolate mofetil with some clinical improvement.
- The dysphagia recurred two months later and EGD showed and esophageal stricture measuring 7mm requiring a neonatal upper gastroscope to facilitate balloon dilatation.
- Biopsies examined for immunofluorescence showed continuous linear deposition along the basement membrane zone for IgG and C3.
- Her final differential was atypical esophageal bullous systemic lupus erythematosus or atypical esophageal bullous pemphigoid.
- The patient declined systemic steroids. Her mycophenolate mofetil dose was increased and she received weekly dilatations to prevent Koebnerization and allow time for medical therapy effect.
- After three months of treatment, she had symptom improvement.

## IMAGES



A. Cervical stricture pre-dilatation (7mm)

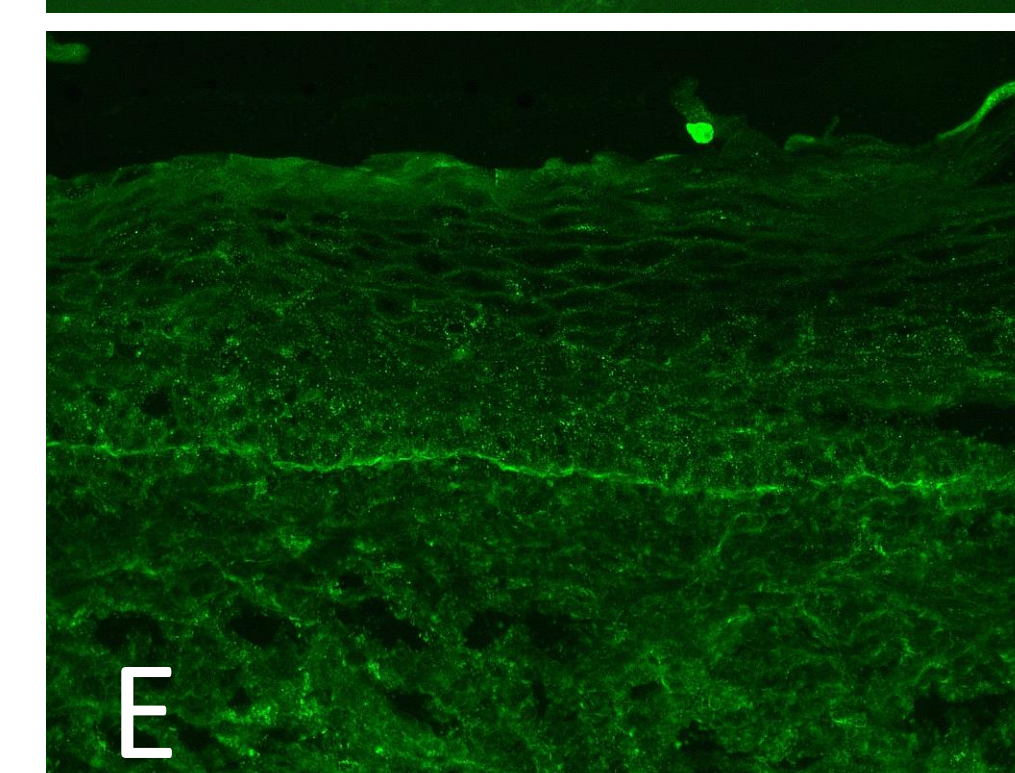
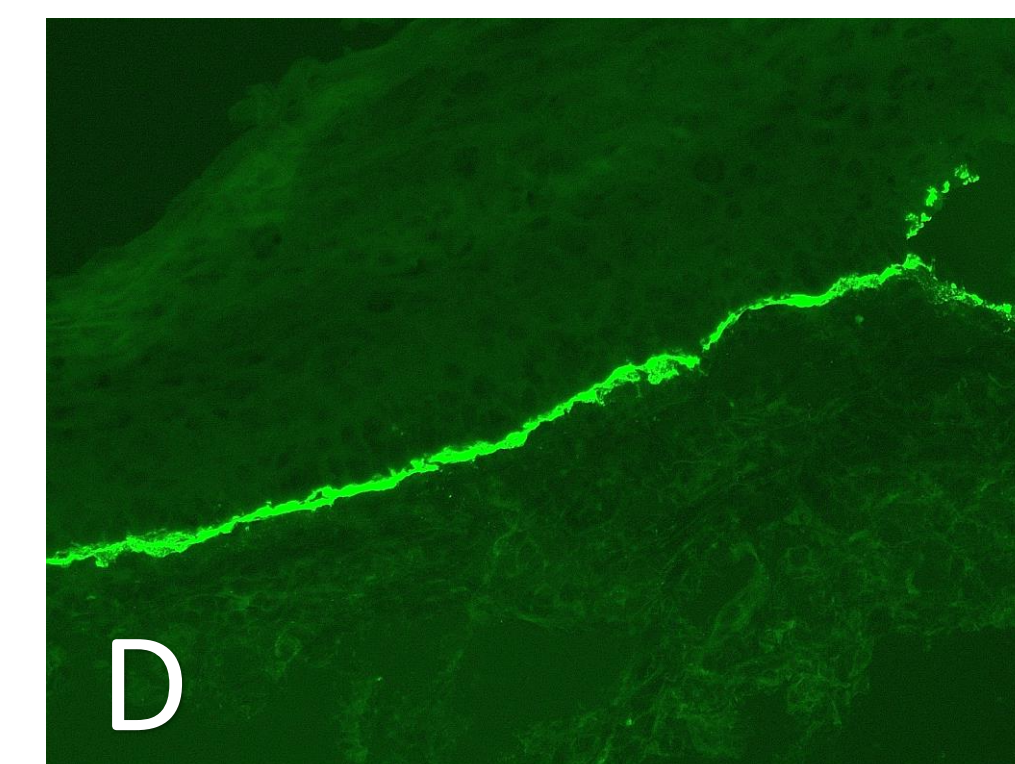


B. Post dilatation (12mm) with push type dilator

C. Esophagram with upper cervical stricture

D. IgG conjugate demonstrating strong and diffuse linear deposition along the mucosal basement membrane zone (200x)

E. C3 conjugate demonstrating strong and diffuse linear deposition along the mucosal basement membrane zone (200x)



## DISCUSSION

- In patients with systemic immune-mediated disorders including rheumatological disease, bullous diseases can be considered as a cause of esophageal stricture.
- Bullous disease that affects the esophagus is rare, especially in the absence of cutaneous symptoms.
- Bullous disease with esophageal involvement can be difficult to treat, especially in this patient who declined corticosteroids. A combination approach using both immune suppression and dilatation was effective.
- Both bullous SLE and bullous pemphigoid can affect the esophagus, and can present with esophagitis dissecans. Other symptoms can include dysphagia, hematemesis, and melena.
- Diagnosis of bullous esophageal disease is primarily made based on biopsy showing IgG with or without IgA, IgM, or C3 at the basement membrane. Antibodies to type VII collagen can be found.

## CONCLUSIONS

- Autoimmune diseases that can affect the esophagus include both rheumatological and bullous disease.
- In patients with recurrent, severe strictures, bullous disease should be a consideration even in the absence of cutaneous findings.

## REFERENCES

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