



## A Strange Case of Esophageal Dysmotility and Neck Swelling in a Lupus Patient

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### Abstract

Achalasia is a disorder of esophageal dysmotility characterized by impaired lower esophageal sphincter (LES) relaxation, resulting in poor passage of food bolus into the stomach. Gastrointestinal manifestations are frequently observed in systemic rheumatologic conditions, including systemic lupus erythematosus (SLE).

This is a report of a middle-aged female with SLE who presented with sudden-onset dysphagia and anterior neck swelling. Imaging revealed a massively dilated, air-filled esophagus with associated anterior tracheal displacement. Although manometry showed a normal integrated relaxation pressure (IRP) of 12 mmHg—technically falling short of formal achalasia diagnosis per Chicago Classification v4.0—the patient demonstrated 100% failed swallows, absent contractility, and pan-esophageal pressurization.

She underwent pneumatic dilation and later laparoscopic Heller myotomy with Dor fundoplication with significant improvement. This case illustrates an atypical esophageal dysmotility pattern in an SLE patient, highlighting the need to consider sigmoid esophagus with air trapping in patients presenting with concurrent dysphagia and anterior neck swelling.

**Keywords:** Achalasia; Lupus; Dysmotility; Esophagus; Manometry

### Introduction and Background

Achalasia is a rare esophageal motility disorder characterized by impaired LES relaxation and absent peristalsis, which impairs the passage of ingested contents into the stomach. It affects approximately 1.6 per 100,000 individuals annually [1]. Symptoms are often nonspecific, including dysphagia to solids and liquids, retrosternal discomfort, regurgitation, and weight loss. Due to its overlap with more common conditions such as gastroesophageal reflux disease (GERD), achalasia is frequently misdiagnosed [2]. Patients are at increased risk for aspiration-related complications due to esophageal stasis.

The differential diagnosis for achalasia includes neuromuscular dysphagia, esophageal stricture or neoplasm, Schatzki ring, scleroderma, diverticulum, and even cardiac ischemia. Initial workup typically includes endoscopy and barium swallow. High-resolution esophageal manometry (HREM) is the gold standard for diagnosis [3-5].

According to the Chicago Classification v4.0, achalasia is classified into three subtypes [6]:

- Type I (classic): 100% failed swallows, no pressurization
- Type II: failed swallows with pan-esophageal pressurization
- Type III: spastic distal contractions

All subtypes demonstrate impaired LES relaxation.

Gastrointestinal manifestations are common in rheumatologic diseases. In systemic lupus erythematosus (SLE), gastrointestinal involvement may precede diagnosis and can affect any part of the alimentary tract. Dysphagia, oral ulcers, and mesenteric vasculitis are among the better-recognized complications [7,8]. Dysphagia has been reported in up to 11% to 50% of SLE patients [9].

### Case Presentation

A middle-aged Caucasian woman with a history of SLE presented to the emergency department

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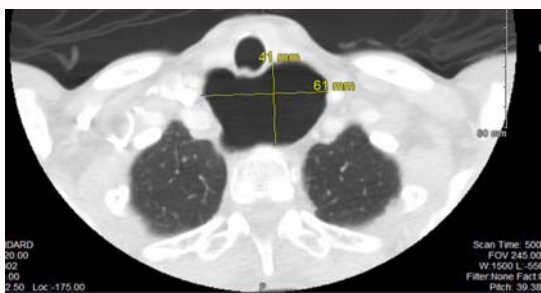
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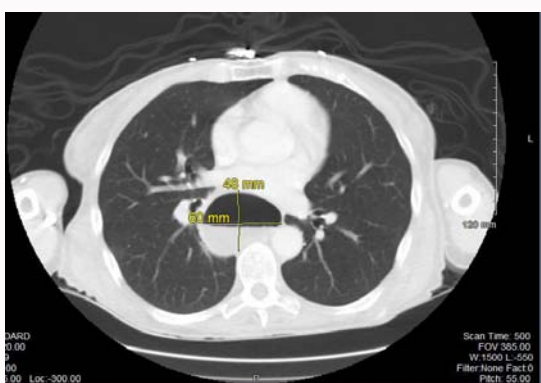
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**Figure 1:** Axial section near the lung apices revealing a very dilated proximal esophagus full of air posterior to the trachea.



**Figure 2:** Axial section below the carina shows that the mid-esophagus is still quite dilated retaining both air and fluid.



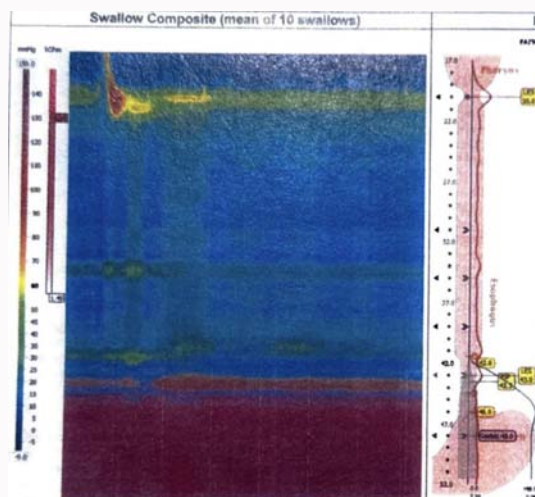
**Figure 3:** Coronal of chest CT scan revealing a diffusely dilated sigmoid esophagus full of air.

with sudden-onset dysphagia and difficulty breathing following dinner. She reported a sensation of food trapped in her throat. Physical examination revealed a large anterior neck swelling with anterior displacement of the trachea. Oropharyngeal examination was unremarkable. After drinking a small amount of water and jumping up and down, the patient noted immediate resolution of the swelling.

She presented again several weeks later with identical symptoms. A CT scan of the neck with contrast revealed a massively dilated, air-filled esophagus extending from the cervical to abdominal



**Figure 4:** Barium swallow fluoroscopy revealing minimal passage of contrast through the LES.



**Figure 4:** Barium swallow fluoroscopy revealing minimal passage of contrast through the LES.

segments [5] (Figures 1-5). She was admitted for further evaluation. A barium esophagram demonstrated minimal contrast passage into the stomach. Esophagogastroduodenoscopy (EGD) showed a dilated, fluid-filled esophagus with resistance at the LES. HREM was subsequently performed.

Manometry demonstrated absent contractility with 100% failed swallows and 60% pan-esophageal pressurization. Her IRP was 12.2 mmHg (within normal limits), and no hiatal hernia was present. Bolus clearance was complete.

She was referred for pneumatic dilation. During endoscopic intubation, a loud rush of air was observed, with immediate decompression of the neck swelling. Dilation was performed to 30 mmHg. Her symptoms briefly improved but recurred within two days. She subsequently underwent a laparoscopic Heller myotomy with Dor fundoplication without complication. Her symptoms improved, and she was discharged after a short observation period. The patient was lost to follow-up.

## Discussion

This patient's HREM did not meet strict criteria for achalasia under Chicago Classification v4.0, given her normal IRP. However, she demonstrated 100% failed swallows, absent contractility, and pan-esophageal pressurization—findings that support a functional obstruction phenotype. Atypical cases like this may still benefit from interventions typically used for classic achalasia [6].

One limitation was the lack of histopathologic evaluation during EGD. In patients with autoimmune disease, biopsy of the esophagus may reveal inflammation, fibrosis, or plexus degeneration. Previous studies have proposed immune-mediated mechanisms in achalasia pathogenesis, including CD4+ T cell infiltration, Th17/Treg imbalance, and elevated cytokines such as IL-17 and TNF- $\alpha$  [10].

This case highlights the underrecognized link between SLE and esophageal dysmotility. Although most literature on autoimmune motility focuses on systemic sclerosis, there is growing evidence of esophageal involvement in SLE [8,9,11]. Air trapping and tracheal compression secondary to esophageal distension is a rare but important presentation [12,13]. Future research should explore manometry-immunopathology correlations and assess outcomes of decompression or surgical therapy in such atypical cases [14,15].

## Conclusion

This case illustrates an unusual presentation of esophageal dysmotility in an SLE patient, marked by sudden dysphagia, neck swelling, and functional outflow obstruction. When faced with concurrent dysphagia and anterior neck swelling, consider sigmoid esophagus with air trapping as a rare but serious differential. Normal IRP does not rule out clinically significant motility disorders. Multidisciplinary evaluation and manometry remain essential for diagnosis and management.

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