Acute Laryngeal Edema as a Presenting Manifestation of Systemic Lupus Erythematosus: A Case Report and Systematic Review

Hudson M. Surber, MD, MPH[1]; Madeline P. Smith, MD[2]; Samuel D. Cloyd, MD[1]; Brooke Taylor, MD[2]; Robert E. Gurevich, MD[2]; Laura Pelaez, MD[1]

[1]Department of Otolaryngology–Head & Neck Surgery, Louisiana State University · [2]Tulane University

AAO–HNSF Annual Meeting 2026 · **Case Report**

Introduction

Acute laryngeal edema is a rare but potentially life-threatening manifestation of systemic lupus erythematosus (SLE) and may, in exceptional cases, be its initial presentation. We report a case of acute laryngeal edema in newly diagnosed SLE with a systematic review of lupus-associated upper airway involvement.

Hypothesis

Acute upper airway edema is a rare but life-threatening initial or early manifestation of SLE. Early recognition is essential, as delays in care can be fatal.

Methods

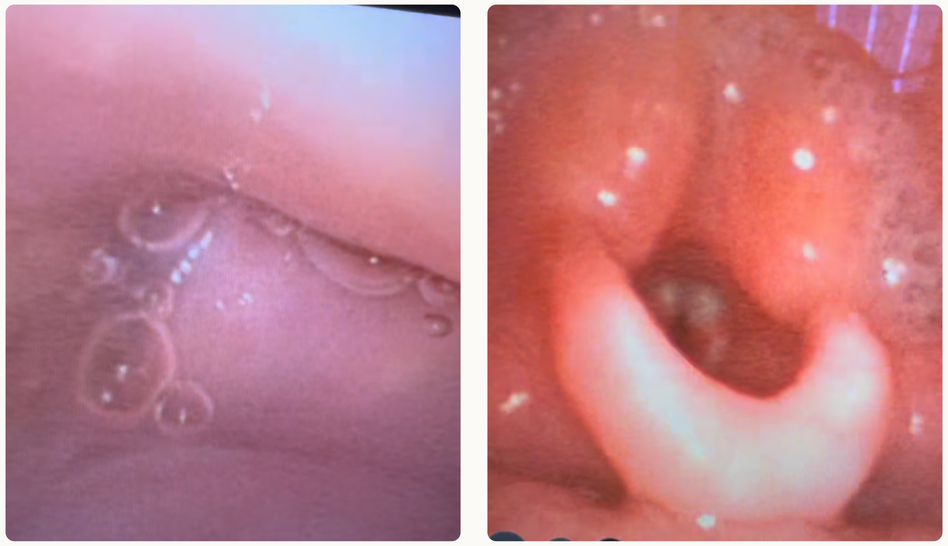
- **Design:** single-patient case report with a systematic review of published cases
- **Setting:** acute care hospital, 2025
- **Interventions:** airway evaluation and management, corticosteroids, treatment of possible infectious causes, multidisciplinary input
- **Outcomes:** resolution of edema, need for airway intervention, response to immunosuppression, short-term course
- **Review:** descriptive synthesis of published cases: presentation, mechanisms, management, and outcomes

Clinical Pearl

Consider SLE in unexplained acute laryngeal edema, and evaluate the airway early in newly diagnosed SLE with airway symptoms.

Case Presentation

An 18-year-old female, diagnosed with SLE one week earlier, was transferred from Mississippi to University Medical Center New Orleans with acute-onset malar rash and acute hypoxic respiratory failure. Tracheal tugging was noted on initial evaluation, and flexible laryngoscopy was performed (Fig. 1).



A. Supraglottic view of the larynx at presentation. **B.** Same view after final successful extubation.

Figure 1. Flexible laryngoscopy before and after acute treatment.

Clinical Course

1 — Airway

Awake fiberoptic transnasal intubation in the OR, well tolerated

2 — Treatment

Multiple rounds of high-dose IV corticosteroids while intubated; rheumatology consulted

3 — Extubation

Failed three attempts, each requiring reintubation; succeeded on the fourth

Had the fourth extubation failed, tracheostomy would likely have been required.

Literature Review

Key published series · systematic review in progress

97 patients with laryngeal SLE in the largest review[1]

28% of those patients had laryngeal edema[1]

4/158 SLE admissions with life-threatening upper airway disease[2]

Laryngeal SLE ranges from mucosal edema and ulceration to cricoarytenoid arthritis, vocal fold paralysis, subglottic stenosis, and necrotizing vasculitis.[1] Proposed mechanisms include immune-complex vasculitis and acquired C1-inhibitor–mediated angioedema.[5] Most cases respond to corticosteroids,[1] yet fatal laryngeal edema has been reported,[3] obstruction can be the sole sign of a flare,[4] and intubation during active SLE may itself trigger airway complications.[2]

Conclusions

- Acute laryngeal edema can be an initial or early manifestation of SLE.
- It is rare, life-threatening, and poorly described in the literature.
- Early recognition and airway management are critical; delays can be fatal.
- Otolaryngologists play a key role in recognizing airway manifestations of systemic disease.

References

1. Teitel AD, et al. Semin Arthritis Rheum. 1992;22(3):203-214.
2. Martin L, et al. J Rheumatol. 1992;19(8):1186-1190.
3. Scarpelli DG, et al. N Engl J Med. 1959;261:691-694.
4. Karim A, et al. Chest. 2002;121:990-993.
5. Cacoub P, et al. Arthritis Rheum. 2001;44(8):1836-1840.