

# Oh, the Places Sweet's Syndrome Will Go: An Advanced Case of Extracutaneous Sweet's Syndrome with Pathologic Sampling and Direct Visualization of Cutaneous, Intranasal, and Bronchial Lesions

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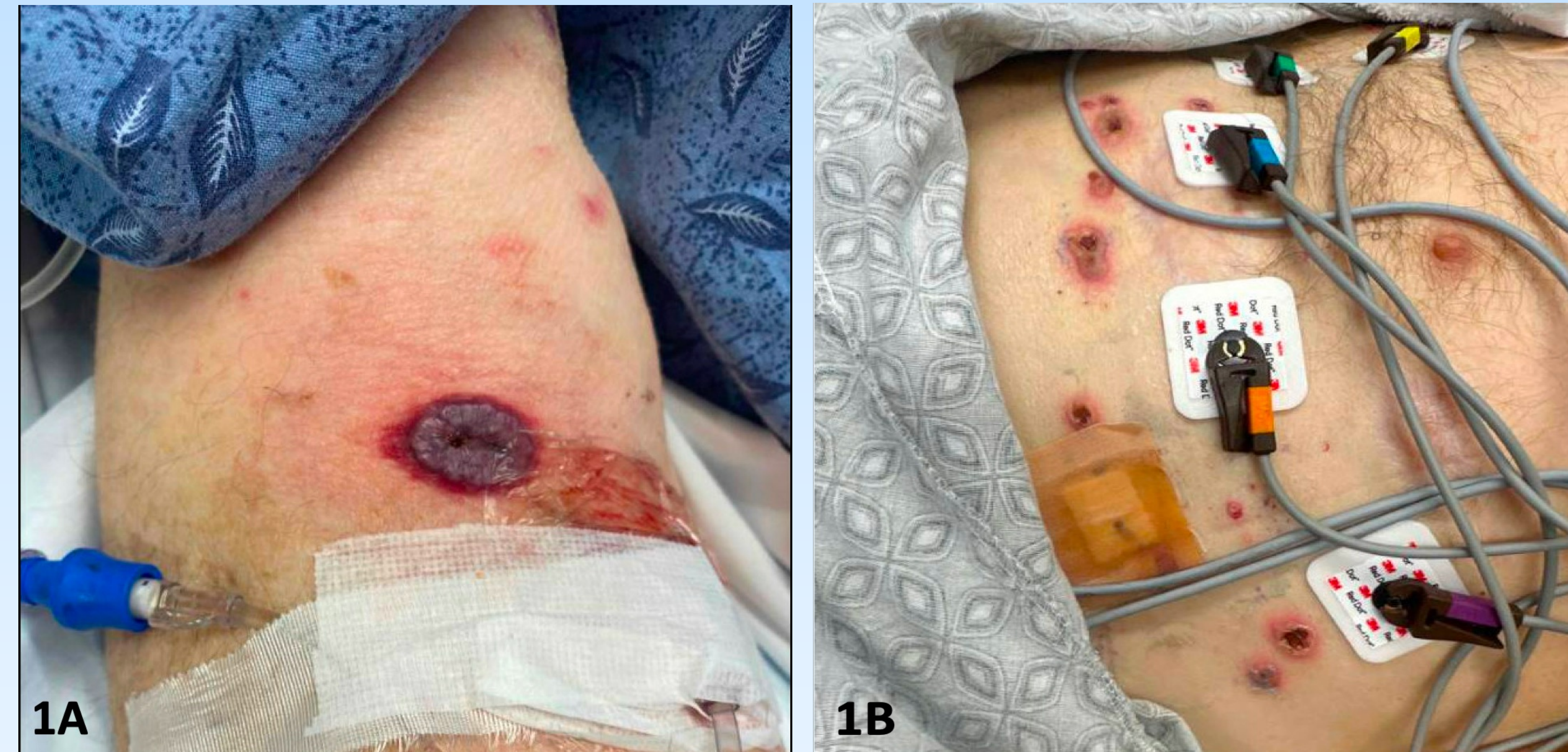
## Introduction

- **Advanced cases of Sweet's syndrome can progress to extracutaneous involvement**, with neutrophil-dense lesions similar to classic cutaneous plaques and nodules that can manifest in nearly any organ system.<sup>1-3</sup>
- Extracutaneous Sweet's syndrome (eSS) lesions are frequently unable to be confirmed by pathology and **have rarely been directly visualized**.

## Clinical Case

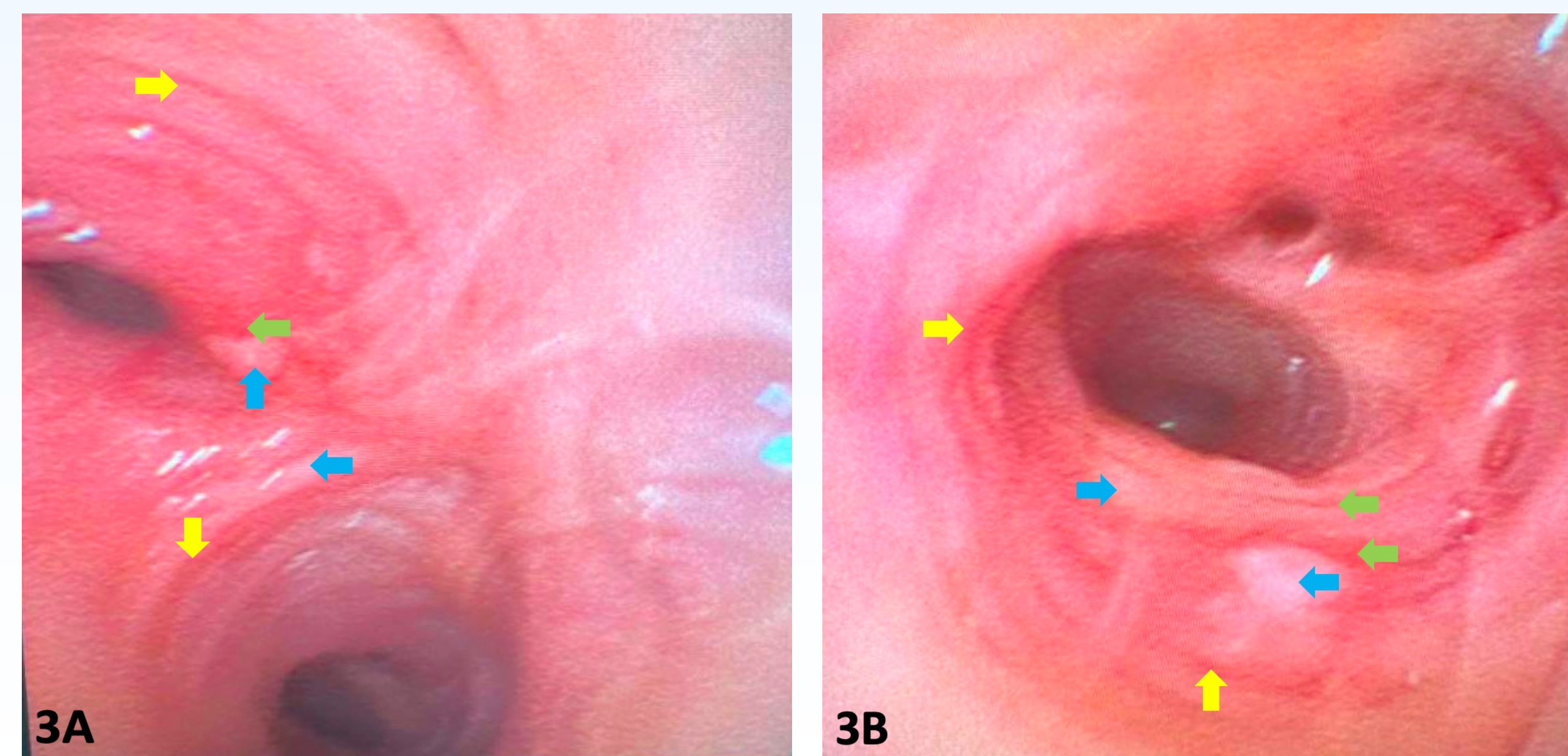
- **Brief Clinical Summary:** 70M with recurrent secondary **acute myeloid leukemia** (transformed from myelodysplastic syndrome, undergoing treatment with venetoclax/decitabine; complicated by neutrophilic eccrine hidradenitis due to cytarabine therapy, and pyoderma gangrenosum) with progressive hypoxia.
- Evaluated by the inpatient dermatology consult service on oncology floor due to episodic fevers and progressively erupting **tender, edematous, erythematous plaques** on the abdomen and extremities, including venipuncture sites (**Figure 1**); biopsy was completed.
- The patient was transferred to the ICU due to hypoxemia, with concern for invasive fungal infection.
- **Nasal endoscopy** visualized edematous ulcerated plaques in the bilateral **sphenoid and ethmoid structures** (**Figure 2**); biopsy was completed.
- Six days later, **bronchoscopy** directly visualized edematous, erythematous plaques in the **bronchus and proximal bronchioles** (**Figure 3**); biopsy on repeat bronchoscopy was unable to be completed due to rapidly decompensating clinical instability.

## Cutaneous (Skin Exam) Lesions



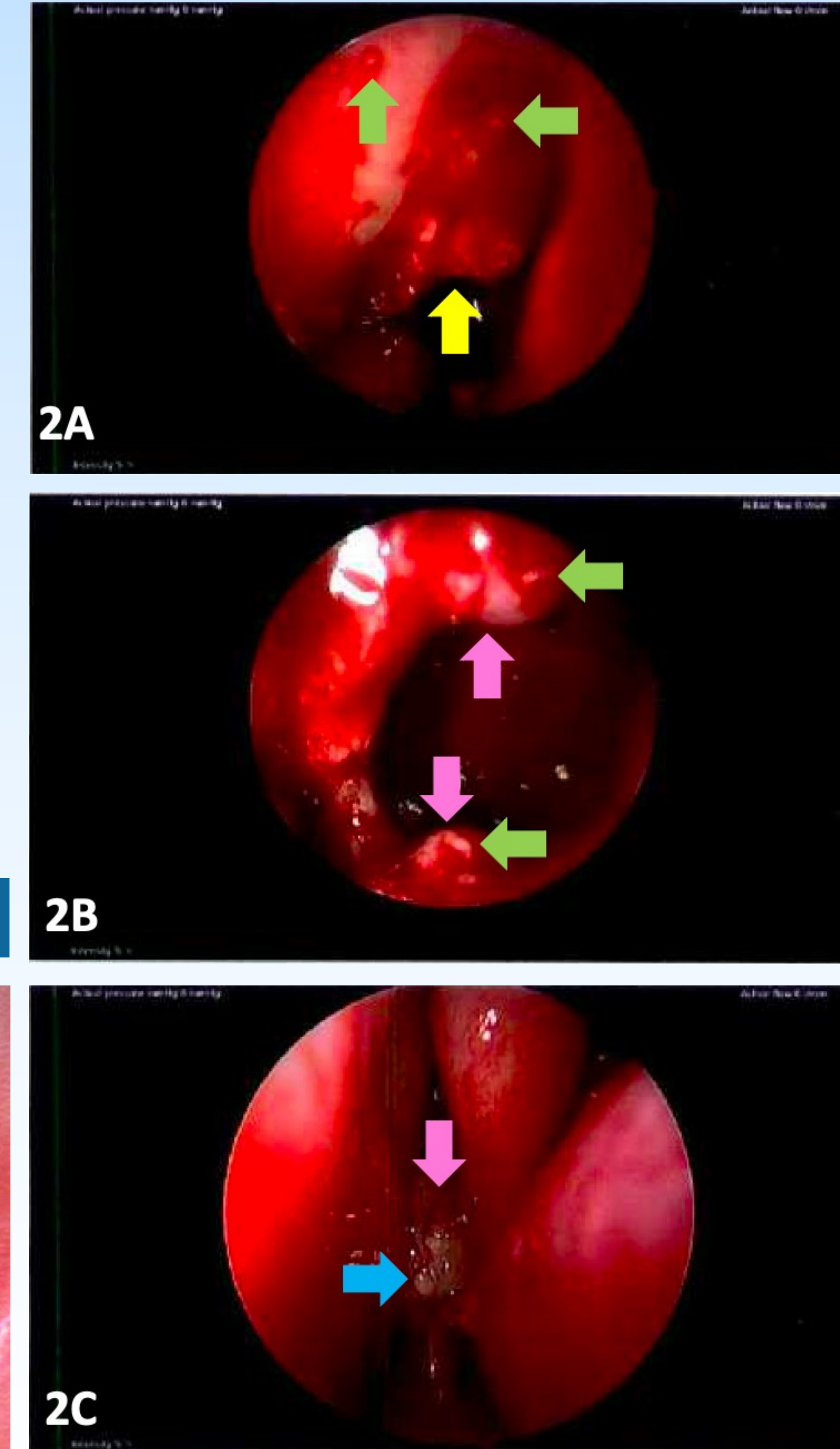
**Figure 1:** Cutaneous Sweet's syndrome lesions at sites of trauma on the upper extremity and abdomen. **1A:** A dusky violaceous edematous nodule with central linear ulceration and ecchymotic border erupted on the right antecubital fossa at site of prior venipuncture; dermatopathology confirmed the presence of a sterile florid infiltrate of neutrophils in the dermis and subcutaneous tissue. **1B:** At sites of prior electrocardiogram lead placement and skin trauma, there are erythematous edematous plaques with significant ulcerations and hemorrhagic crusts.

## Bronchoscopy Lesions



**Figure 3:** Bronchoscopy of the bilateral bronchi and proximal bronchioles (ICU Day 8). **3A, 3B:** On the luminal bronchial mucosal surface, there are multiple edematous erythematous-to-white plaques (blue arrow) on superficially ulcerated borders (green arrow) with concentric erythematous rings (yellow arrow); biopsy unable to be completed due to clinical instability during bronchoscopy, but later determined to be consistent with eSS upon autopsy.

## Nasal Endoscopy Lesions



**Figure 2:** Nasal endoscopy with Sweet's syndrome involving the sphenoid and ethmoid intranasal structures (ICU Day 2). **2A, 2B:** On the bilateral sphenoid surfaces, there are multiple shiny edematous, bleeding nodules (pink arrow) with numerous superficial ulcerations (green arrow); the prominent lesion in 2A (yellow arrow) was biopsied and cultured, showing sterile florid neutrophil-predominant infiltration, felt to be consistent with eSS upon dermatopathology review. **2C:** At the inferior junction of the ethmoid and lacrimal bones, there is a similar appearing brightly erythematous edematous nodule (pink arrow) with overlying vesiculation (blue arrow).

## Clinical Outcome

- The patient ultimately passed away after a transition to palliative care, due to escalating hypoxia and hypotension. Blood cultures, including fungal cultures, remained negative.
- **Autopsy confirmed diffuse coalescent purulent plaques along the nasal mucosa, trachea, and bronchi**, with negative stains for bacterial (Gram positive, Gram negative, acid-fast) or fungal forms.

## Advanced Teaching Pearls

- This case demonstrates the gross clinical appearance of **pathology-confirmed eSS in the nasal sinuses and bronchial airway**.
- Bronchial eSS has rarely been reported.<sup>4,5</sup> In our case, it appears as an edematous plaque in the bronchus like its appearance on the skin.<sup>3</sup>
- Pathology is critical in diagnosing eSS, which **can appear similarly to atypical mycobacterial or invasive fungal infections**, with fevers and ulcerating or dusky edematous plaques in nasal passages and airways.
- eSS likely represents an **“end stage” of disease with a high likelihood of life-threatening clinical decompensation**, from partially obstructing masses of the airway and blood vessels or systemic inflammatory response syndrome with distributive shock.<sup>6,7</sup>

## References

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