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 neutrophilic dense lesions similar to classic cutaneous plaques and nodules that can manifest in nearly any organ system.1–3

• 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 ecrine 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 bronchi (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.

• A 52-year-old edematous nodules 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.

Nasal Endoscopy Lesions

Figure 2: Nasal endoscopy with Sweet’s syndrome involving the sphenoid and ethmoid intranasal structures (ICU Day 2).

• On the bilateral sphenoid surfaces, there are multiple shiny edematous plaques along the nasal mucosa, trachea, and bronchi, with negative stains for bacterial (Gram positive, Gram negative, acid-fast) or fungal forms.

Bronchoscopy Lesions

Figure 3: Bronchoscopy of the bilateral bronchi and proximal bronchioles (ICU Day 8).

• On the luminal bronchial mucosal surface, there are multiple edematous erythematous-to-white plaques (blue arrow) on superficially ulcerated borders (green arrow) with numerous superficial ulcerations (green arrow); the prominent lesion in 2A (yellow arrow) was biopsied and cultured, showing sterile florid neutrophilic-predominant inflammation, felt to be consistent with eSS upon dermatopathology review.

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.4,5 In our case, it appears as an edematous plaque in the bronchus like its appearance on the skin.3

• 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.6,7

References