Clinicopathologic Correlates and Differential Diagnoses of Sweet Syndrome in a Pediatric Patient

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BACKGROUND

- Acute febrile neutrophilic dermatosis, or **Sweet syndrome (SS)**, is a rare autoinflammatory condition characterized clinically by painful erythematous papules and plaques, and histologically by a dermal neutrophilic infiltrate and edema.
- We discuss the clinical and histopathological correlates of the disease, differential diagnoses, and key diagnostic features.

CASE PRESENTATION

- A **6-year-old** male presented with a **pruritic, oozing rash** for the past **2.5 years**, which started on the buttock area and extended to the upper and lower extremities, and recently spread to the face.
- **Associated symptoms**: fatigue, weakness, cold intolerance, epistaxis
- **PMHx and FMHx**: Unremarkable
- **Medications**: None
- On **examination**, circinate erythematous plaques were noted, some with infiltration and others with ulceration, extending to the upper and lower extremities, ears, and face (including the nasal mucosa) (**Figure 1**).
- **Labs:** Unremarkable
- **Ddx**: Pemphigus, IgA pemphigus, subcorneal pustular dermatosis, and SS
- **Treatment:** Mometasone 0.1% daily
- **Punch biopsy:** Dense infiltrate of neutrophils with scattered eosinophils extending into the deep dermis, with overlying spongiosis and papillary dermal edema (**Figure 2**). Biopsy was negative for fungi, bacteria, mycobacteria, and EBV.
- **Direct immunofluorescence (IF):** Negative for immunoprotein deposition
- **Indirect IF:** Increased IgA and mildly elevated IgG and BP180 antibodies
- Given the clinical and histopathologic findings, **idiopathic SS versus linear IgA bullous dermatosis** were of highest suspicion, and the patient was started on daily dapsone therapy.
- **Follow-up** three weeks later **revealed lesion improvement** (though not yet at target). Patient denied joint pain, fevers, or medication side effects.

CLINICAL AND HISTOLOGIC FINDINGS

Figure 1. Clinical Findings: Erythematous circinate plaques are shown over the bilateral upper and lower extremities **(A-C)**. An ulcerated plaque is noted over the right elbow **(C)**.

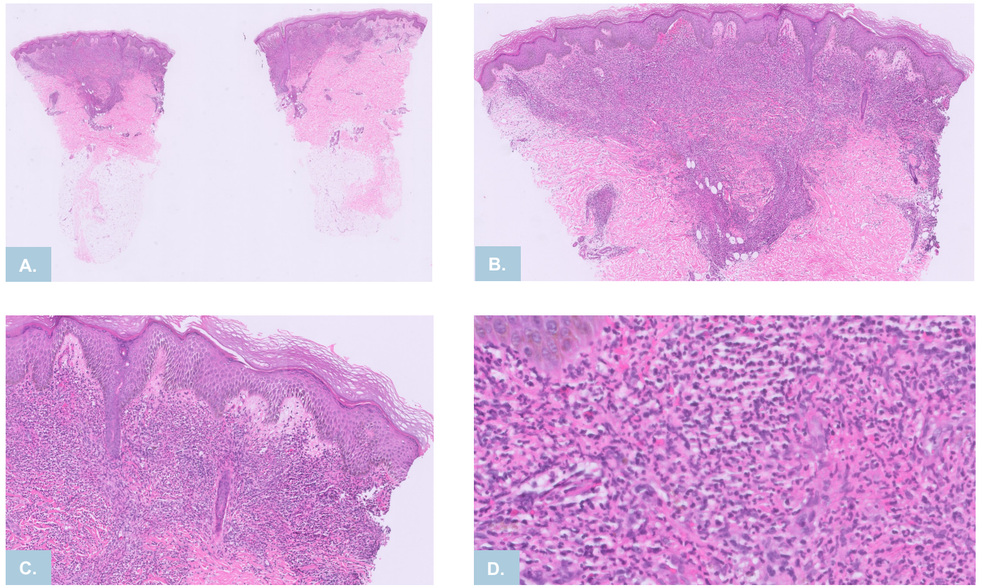

Figure 2. Histologic Findings at Low **(A, B)** and High **(C, D)** Power. Biopsy showed dense neutrophilic infiltrate extending into the deep dermis, with overlying spongiosis and papillary dermal edema.

DISCUSSION

- SS classically presents with a **nonscarring rash**, **fever**, and **generalized** constitutional symptoms. Fever is present 80-90% of the time in classical SS.[1]
- Physical examination reveals **erythematous plaques** that may be diffuse. **Oral** involvement is **rare** in classical SS (2%), while **ophthalmic** symptoms are **common** (17-72%).[1]
- **Biopsy is key** to diagnosis and features **neutrophilic** (as well as lymphocytic) infiltration and **papillary dermal edema** with relative **sparing of the epidermis** and **blood vessels**;[2] scattered eosinophils may be present.[2]
- Though uncommon, several pediatric cases of SS are reported, with prognosis often favorable.
- Cases of pediatric SS have been **associated with respiratory** infections,[3] **HIV**,[4] **rotavirus**,[5] **drugs**, and underlying **autoinflammatory** conditions (e.g., lupus erythematosus)[6] or **malignancy** (e.g., AML).[7]
- Our patient had no identifiable underlying condition, was not taking any medications at the time of presentation, and likely experienced the **idiopathic, classical**, subtype of SS (the **most common subtype** of SS in children).[8]

SUMMARY

- SS is a **rare** neutrophilic dermatosis with **distinct clinical** and **histologic** findings, classically presenting with a nonscarring characteristic rash, fever, and generalized constitutional symptoms.
- Biopsy is key to diagnosis, of which neutrophilic infiltration and papillary dermal edema with relative sparing of the epidermis and blood vessels are hallmarks.
- As with many dermatologic conditions, **correlating clinical and histologic findings** is paramount in diagnosing SS. This case highlights key features of the disease in pediatric patients.

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