

CASE REPORT

Pediatric sweet syndrome

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Abstract

Pediatric Sweet syndrome is a rare dermatosis often triggered by a prodromal illness or infection and characterized histologically by a dense neutrophilic infiltrate. We report a 2-year-old girl with a classic presentation of Sweet syndrome following an acute thumb paronychia, who had a negative history of malignancy or immunodeficiency.

KEYWORDS

acute febrile neutrophilic dermatosis, neutrophilic dermatoses, pathergy

1 | CASE REPORT

A 2-year-old girl presented to the emergency department with an acute, rapidly progressive swelling and tenderness of her left hand and a superficial erosion over her abdomen. On examination, there was erythema with notable swelling of the left thumb as well as edematous papules and a tense hemorrhagic bulla on the left volar wrist (Figure 1). Laboratory studies were significant for an elevated WBC 39.2 K/ μ l (reference: 6.0–17) and 80% neutrophils, ESR 73 mm/h, and CRP 20.1 mg/dl (reference: <10). A bacterial culture of the thumb grew methicillin-resistant *Staphylococcus aureus*, but bedside incision and drainage and broad-spectrum antibiotics did not lead to clinical improvement. Blood and viral cultures and a blood smear were unremarkable, and a CT scan with contrast of the left arm showed extensive subcutaneous edema. Notably, new lesions developed in regions of minor trauma, including intravenous cannula sites and areas of braided hair. The lesions that developed secondary to pathergy allowed a clinical diagnosis of Sweet syndrome to be made. Her

symptoms started to resolve after a 5-day course of prednisolone 0.5 mg/kg/day and wound care.

2 | DISCUSSION

Sweet syndrome (acute febrile neutrophilic dermatosis) is characterized by an acute onset of painful, erythematous, violaceous, and edematous papules. The disorder has been documented in children, although the condition is rare.^{1–3} Adult cases generally occur in women between ages of 30–60.⁴ The mean age for pediatric cases is five, with a male predominance under ages of three.² The pathogenesis of Sweet syndrome remains undetermined, but the presence of fever and leukocytosis suggests a septic process.⁴

Adult Sweet syndrome has three clinical subtypes: classical, malignancy-associated, and drug-induced.⁴ Such classifications have not been explicitly delineated in pediatric Sweet syndrome but are likely similar.² Classically, for both pediatric and adult populations, the disorder is preceded by an infection or illness. Hematologic malignancies

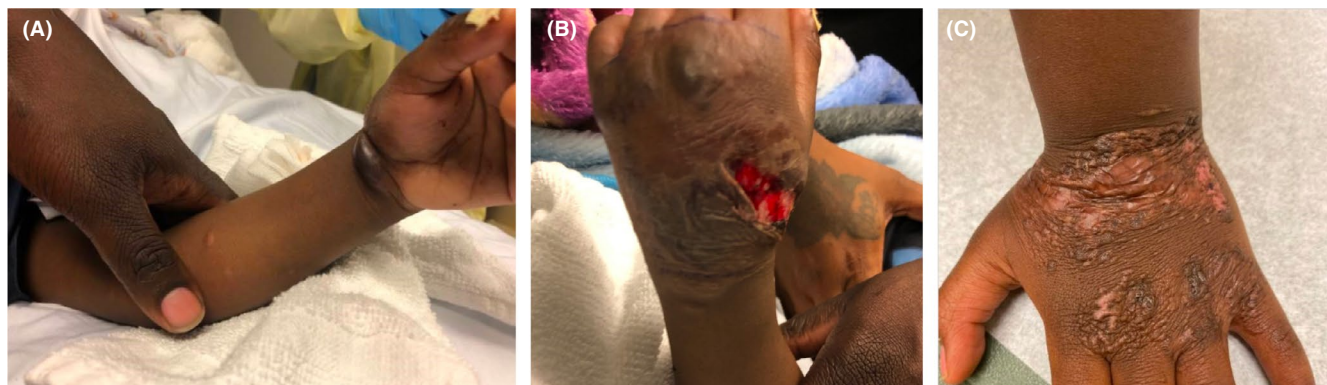


FIGURE 1 Tense hemorrhagic bulla (A) with erythema and erosions of near the left thumb (B) and edematous papules on the dorsal wrist (C)

have been associated with 24% of pediatric cases of Sweet syndrome.² In addition, medications such as granulocyte-stimulating factor and trimethoprim have been linked to syndrome onset.² Our patient likely has a classical presentation triggered by a localized soft-tissue infection, even meeting adult diagnostic criteria.⁴ Additional reports of pediatric Sweet syndrome would be helpful to further define subtypes in this population.

The presence of a widely distributed pustular rash, fever, neutrophilic leukocytosis, and elevated inflammatory markers can cause clinicians to anchor to an infectious cause, such as erysipelas/cellulitis. The lack of clinical improvement after an incision and drainage and several antibiotic courses in our case suggested another diagnosis. Blood smears and imaging ruled out hematologic malignancies and abscesses, respectively. A unique sign of Sweet syndrome is pathergy, which occurs in 20% of cases and increases the likelihood of post-inflammatory scarring.² While a skin biopsy is confirmatory, a diagnosis can be achieved through clinical presentation and laboratory abnormalities. Patients rapidly improve after initiating systemic steroids followed by a tapered dose. Routine echocardiography may be necessary to monitor for cardiac sequelae.³

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We would like to thank the patient's guardian for consent to publish this case report.

CONFLICT OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

AEZ was responsible for writing the patient vignette and manuscript, synthesizing the appropriate literature, and answering editorial comments. CMW, SG, and KMH were

responsible for patient management, reading the appropriate literature, and supplying feedback on the case presentation and manuscript. MSD was responsible for patient management, supplying feedback and comments, and conceived the idea for the manuscript.

ETHICAL APPROVAL

Not applicable.

DATA AVAILABILITY STATEMENT

Data sharing is not applicable to this article as no datasets were generated or analyzed during the current study.

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