

Inflammation and infection

Genital Sweet's Syndrome in a patient with Acute Myelogenous Leukemia

Daniel Ensley, Grant H. Evans^{*}

Brooke Army Medical Center, 3551 Roger Brooke Dr, Ft Sam Houston, TX, 78234, USA



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ABSTRACT

A 71 year old male presented to the emergency room for evaluation of acute erythema, edema and pain of the penis and scrotum. There was initial concern for Fournier's gangrene, however labs were unremarkable and vital signs were stable. He did not improve with antibiotics. Biopsy results showed neutrophil infiltration consistent with Sweet's Syndrome. He was started on corticosteroids and discharged home in stable condition. In a hemodynamically stable patient not responding to antibiotic therapy, close observation is prudent until the tissue biopsy results.

Introduction

For better or worse, urologists are often called upon to evaluate cutaneous genital lesions, simply because they exist on the genital skin. Given this, urologists should have a reasonable grasp of dermatologic conditions that can present with genital manifestations. To the best of our knowledge, we present the first documented case of Sweet's Syndrome occurring in the male genitalia.

Case presentation

A 71 year old male presented to the Emergency Department (ED) for urological evaluation of acute erythema, edema, and pain of the penis and scrotum. He was sent to the ED by his dermatologist, who had just performed a biopsy of the genital lesions, and was concerned for a possible infectious etiology such as Fournier's Gangrene. The rash had acutely worsened in the past 7 days and there was now severe pain in the penis with any type of movement, passive or active. He had been treated presumptively for balanitis with topical anti-fungals for the preceding 2 weeks. According to the patient this had not helped. Of note, the patient had recently been admitted for a rash on his right upper extremity consistent with Sweet's Syndrome and had initiated an oral steroid regimen.

His initial evaluation in the ED included physical examination and basic laboratory studies. His vital signs were normal and stable. On examination, the patient was a cachectic man who was alert, calm, and in mild distress secondary to genital pain. The penis and glans were swollen, and there were purpuric lesions along the shaft and mild

erythema extending from the base of the penis up the escutcheon and down to the anterior scrotum (Fig. 1). There was a small, 4 mm hemostatic biopsy site along the dorsum of the penis base. There was no crepitus on palpation, but discomfort was associated with passive manipulation of the penis. There was no drainage per the urethral meatus. Testicular examination was grossly normal. There was a purpuric rash on his right upper extremity and an area of swelling on the left leg in the pre-tibial area that was firm and not tender. The remainder of the exam was unremarkable. His serum sodium was low at 127, while the remainder of metabolic and hematologic labs were normal. Urine, blood, and wound cultures were all pending at the time of evaluation. There was subcutaneous air noted near the biopsy site on CT.

His past medical history was notable for Myelodysplastic Syndrome (MDS) that had progressed to Acute Myelogenous Leukemia (AML), rheumatoid arthritis and COPD. He had no recent sexual contacts and no known history of STIs. He had no known drug, food, or contact allergies.

The patient was started on broad spectrum antibiotics for a presumptive infectious etiology and he was admitted for observation. A foley catheter was placed due to the degree of edema and concern for urinary retention. Despite no change in his physical exam, he continued to remain afebrile without leukocytosis and with normalization of his chemistry derangements. Oral prednisone was increased to 50mg daily with taper and the skin eruption resolved after discharge.

The 4 mm suprapubic punch biopsy final report described superficial, deep and interstitial infiltration of neutrophils, lymphocytes, and somewhat atypical mononuclear cells with hyperchromasia and occasional prominent nucleoli (Fig. 2). The final interpretation was favored to be Sweet's Syndrome due to the patient's known high-grade myeloid

^{*} Corresponding author.

E-mail address: grant.h.evans.mil@mail.mil (G.H. Evans).

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Fig. 1. Gross appearance of penile lesions.

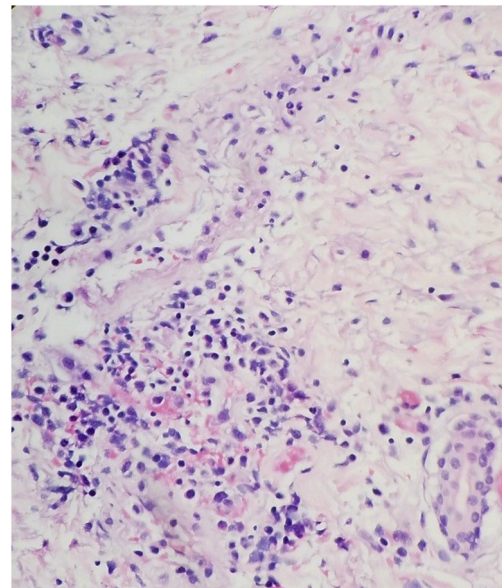


Fig. 2. Microscopic appearance of punch biopsy.

neoplasm.

Discussion

In any male with painful swelling and erythema of the external genitalia, the differential diagnosis must always include potentially life-threatening conditions, such as Fournier’s Gangrene. Once this has been ruled out, other more common conditions, such as trauma and less severe infectious etiologies should be considered. A full physical exam and thorough history (to include sexual history, current and previous partners, prior STI exposure, dysuria, penile drainage, condom usage, antibiotic use, hygiene, allergies and immunosuppressive medication or diseases) should be obtained. The timeline of onset of symptoms and how quickly the swelling and erythema have spread are also very important.

Histiocytoid Sweet’s Syndrome is classically a paraneoplastic process associated with AML, and can be the heralding sign of a malignancy or its recurrence.¹ Often it is accompanied by a flulike prodrome and appears as multiple, firm, tender, erythematous papules distributed on the head, neck and extremities. Diagnosis requires 2 major and 2 of 4 minor criteria (Fig. 3).² Treatment consists of systemic steroid administration, which is associated with a rapid improvement of signs and symptoms. Oral potassium iodide or colchicine may also be used if the clinical picture does not permit corticosteroid use. Surgical debridement is associated with increased morbidity in Sweet’s Syndrome.³ In fact, surgical intervention can elicit a non-specific inflammatory response known as pathergy which can be resistant to healing.^{4,5}

Conclusion

Any patient with subjective fevers, pain and a spreading rash of the external genitalia should be evaluated for the possibility of Fournier’s Gangrene. Diagnosis is clinical and often cannot be delayed for imaging. However, in this patient, past medical history is important context with a prior Sweet’s Syndrome eruption and history of MDS and AML. In a hemodynamically stable patient not responding to antibiotic therapy, close observation is prudent until the tissue biopsy results.

Declaration of competing interest

No competing financial interests exist. The view(s) expressed herein

Major Criteria:	Minor Criteria:
Sudden onset of painful erythematous nodules or plaques	Fever >38 degrees C
Dense neutrophilic infiltrate without evidence of leukocytoclastic vasculitis	Association with: malignancy, inflammatory disease, pregnancy, upper respiratory infection, gastrointestinal tract infection, inoculation
	Abnormal Lab values: ESR >20 mm/h, ELevated CRP, WBC >8,000, >70% neutrophils
	Responds to systemic corticosteroids or potassium iodide

Fig. 3. Major and minor criteria for diagnosis of Sweet’s Syndrome.

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