

DERMATOPATHOLOGY UNIT

UPP | Department of Dermatology

UPMC Dermatopathology “Case of the Month” Presentations

UPP - Department of Dermatology, Dermatopathology Unit

5230 Centre Avenue (412) 623-2614

Pittsburgh, PA 15232 (412) 682-6450 FAX

Ellen Roh, Matthew Zirwas, John McSorley, Hina Sheikh, Drazen Jukic

MAY 2006 CASES OF THE MONTH

CLINICAL FINDINGS

CLINICAL HISTORY

Clinical history:

DM presented to the emergency department and was subsequently admitted for a painful leg rash and guaiac-positive stool. He reported a sore throat prior to his admission, and described also currently having blurry vision, malaise, abdominal pain, joint pain and sores on his lips and genitals. He denied any new medications, IV drug use, high risk sexual activity, or history of similar oral/genital ulcers prior to this illness. An EGD showed multiple erosions and inflammation in his stomach and duodenum. His colonoscopy was negative, and imaging was significant only for some lymphadenopathy. He was started on prednisone, pain medication and pantoprazole prior to a dermatology consult.

Physical exam:

His vital signs were significant for a slight fever (38.7 deg), and in general, he appeared to be an uncomfortable but undistressed male. There were no apparent conjunctival lesions. His lower lip was crusted, but he had no lesions in his oropharynx. His legs had multiple palpable petechiae and a pustule on his left ankle (see figure 1), and there was a tender, firm cord on his left forearm at a previous IV site (see figure 2). There were multiple shallow ulcers on his penis, scrotum and perineum, but no penile discharge (see figure 3). Only his cervical lymph nodes were palpable.

Laboratory tests:

A CBC was notable for an elevated WBC of 20.4. Liver function tests, electrolytes, renal function tests and a fasting lipid profile were within normal limits. Hepatitis B and C, HIV, RPR, RF and HSV were all negative. A throat culture was positive for *Streptococcus pyogenes*. Histopathology: Four punch biopsies were performed from his left forearm, groin and left ankle (second for immunofluorescence). Both the left forearm and groin showed a necrotizing pustular vasculitis with concomitant neutrophilic dermal infiltrate (see figures 4 & 5). A leukocytoclastic vasculitis without emboli or thrombi was seen in the left ankle (see figure 6), with trace deposits of C3 and fibrin around the dermal blood vessels on direct immunofluorescence.

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Figures & Images

1. Click on the *Figure number* you wish to review.
2. Click on the *image* to enlarge



FIG 1



FIG 2



FIG 3

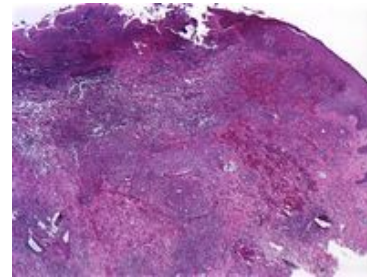


FIG 4 A

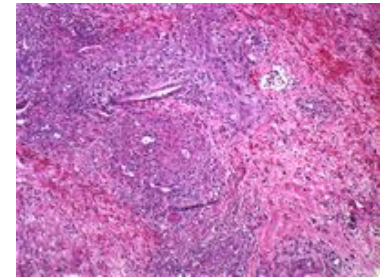


FIG 4 B

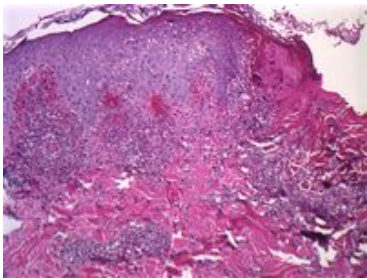


FIG 5 A

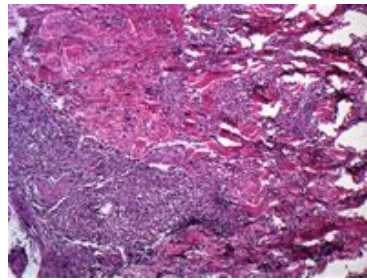


FIG 5 B

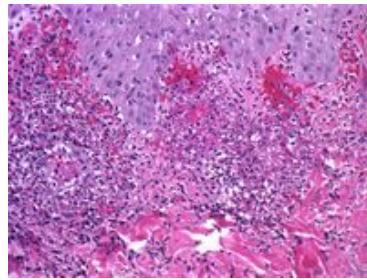


FIG 5 C

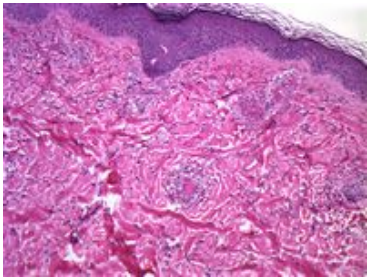
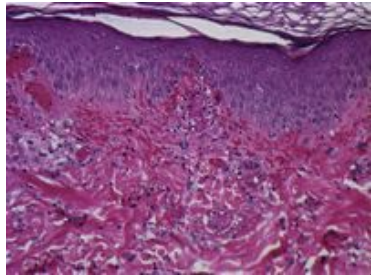


FIG 6 A



6 B

FIG

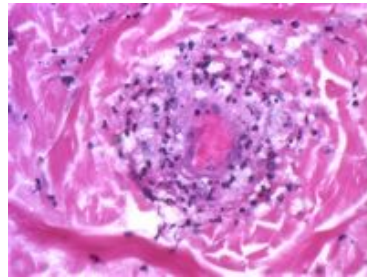


FIG 6 C

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DISCUSSION & DIAGNOSIS

Diagnosis

Behçet's disease

Discussion

Behçet's disease is a rare, multisystem inflammatory disorder of unknown origin. Young adults are most commonly affected, with an increased prevalence in the Middle East and Japan and with the HLA-B51 haplotype. The disease is presumed to be an autoimmune disease, and the primary lesion is vasculitic. It is classically associated with the triad of oral and/or genital ulcers, uveitis and arthritis, but the official diagnostic criteria established by the International Study Group for Behçet's disease require the presence of recurrent oral ulceration and at least two of the following: recurrent genital ulceration, ophthalmic lesions (including anterior or posterior uveitis, hypopyon or retinal vasculitis), skin lesions (including erythema nodosum, pseudofolliculitis or papulopustular lesions), and pathergy (a pustular response of the skin to an intradermal needle prick). However, all organs are susceptible, and it is not uncommon for patients to also develop arthritis, phlebitis, thrombosis of the superior vena cava, ulcers along the gastrointestinal tract or even nervous system involvement.

The diagnosis of Behçet's disease is one of exclusion. Laboratory tests are often nonspecific, and only reflect the inflammatory state. The c-reactive protein, erythrocyte sedimentation rate, leukocyte count and complement components all may be elevated, and up to one-third of patients with Behçet's disease are found to have factor V Leiden deficiency mutations. Histology is also nonspecific. Vasculitis of the involved organ is the most important finding, but one may also find in skin biopsies erythema nodosum, folliculitis, dermal inflammation or fibrosis.

The prognosis of most cases of Behçet's disease is good, as the disease often "burns out" after several years of cycling. Although the recurrent ulceration, arthralgias and skin lesions can be debilitating, the most severe morbidity is with ophthalmic inflammation and resulting visual loss. Mortality is low, but can still occur with CNS and vascular involvement. Hence, treatment is dependent on a patient's organ involvement. Topical anesthetics and corticosteroids can be helpful for symptomatic ulcer relief, but patients are often also treated with systemic immunosuppressive agents, such as systemic corticosteroids, methotrexate, thalidomide or other immunomodulators.

Though this was our patient's first reported episode of oral ulcers, the exclusion of other causes and his other clinical findings of genital ulcers, skin lesions and pathergy (at his IV site) were consistent with Behçet's disease. An ophthalmology exam revealed no ocular pathology, and as the rheumatology consult agreed that the patient's presentation was very suggestive of Behçet's disease, he was started on azathioprine and prednisone, with much clinical improvement soon afterwards.

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