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Febrile ulceronecrotic Mucha–Habermann disease associated with herpes simplex virus type 2

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Febrile ulceronecrotic Mucha–Habermann disease is a rare severe variant of pityriasis lichenoides et varioliformis acuta, a disease within the pityriasis lichenoides spectrum of disorders. It remains uncertain whether these disorders are preneoplastic or reactive against infectious or other antigenic stimuli. Febrile ulceronecrotic Mucha-Habermann disease varies in severity and may be accompanied by a range of systemic symptoms. We present a case associated with herpes simplex virus infection. In this case, no significant T cell clone was identified. Clonality may be a prognostic marker, but reports with T-cell receptor polymerase chain reaction results are limited to eight previous cases, and further reports are required. Awareness of the condition is important because of its fulminant potential. (J Am Acad Dermatol 2009;60:149-52.)

Pityriasis lichenoides is an uncommon inflammatory skin disease in a spectrum from pityriasis lichenoides chronica (PLC) to pityriasis lichenoides et varioliformis acuta (PLEVA). Febrile ulceronecrotic Mucha–Habermann disease (FUMHD) is a rare, potentially fatal variant of PLEVA that is frequently associated with systemic symptoms. Viral and bacterial infections have been associated with several cases.[1]

Reported cases range from fulminant to milder febrile disease. In milder cases, large ulceronecrotic lesions and fever distinguish the condition from typical PLEVA.[2-9]

We describe a case of FUMHD presenting with fever, rigors, malaise, generalized rash with ulceration, and mild liver enzyme elevation. Skin biopsies showed a lichenoid reaction pattern consistent with PLEVA. No significant clonality was found on T-cell receptor polymerase chain reaction (PCR). Herpes simplex virus (HSV) type 2 was cultured from genital lesions, and a skin biopsy from the thigh was positive

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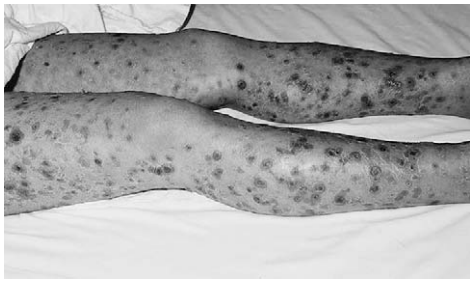

Fig 1. Multiple annular hyperkeratotoic lesions on the legs at presentation. These lesions were also distributed on the arms and trunk, and several had central necrotic ulceration.

Fig 2. Skin biopsy of a lesion on the thigh at presentation showed parakeratosis and acanthosis with severe basal layer vacuolar change and apoptotic keratinocytes. An intense perivascular and band-like lymphocytic infiltrate is present with lymphocytic exocytosis. (Hematoxylin–eosin stain; original magnification: ×40.)

on PCR for HSV DNA, an association not previously described.

CASE REPORT

A 24-year-old south Asian male presented with a 1-month history of rash and a 1-week history of malaise, fever, and rigors. He had axillary lymphadenopathy and generalized annular hyperkeratotic plaques 5 to 30 mm in diameter (Fig 1), many with central hemorrhagic and necrotic ulceration. There were small ulcers on the penis. He reported recurrent genital ulcers 2 years earlier, but had not previously reported a generalized rash. He denied taking any medications recently.

He was admitted for intravenous flucloxacillin 1 g, 6-hourly, topical steroid wraps, and potassium permanganate baths. He had temperatures up to 39°C for 7 days, and then the fever began to settle. He was discharged on oral flucloxacillin and topical steroids. Extensive crops of skin lesions continued for 2 months, developing into 10- to 20-mm necrotic ulcers. These healed slowly, leaving atrophic, hypopigmented scars.

Blood tests showed mild elevation of liver enzymes, normal renal function, and full blood count. Epstein–Barr virus (EBV), hepatitis A, hepatitis B surface antigen, and antihepatitis C virus (HCV) serology, syphilis screen, antinuclear anticytoplasmic antibodies, and blood cultures were negative. C1q binding activity, determined by enzyme-linked immunosorbent assay, was raised at 57 μg/mL (normal range, <50.0). Skin swabs cultured *Staphylococcus aureus* sensitive to flucloxacillin. A viral culture of established genital ulcers was negative, but a repeat culture from new vesicles grew HSV type 2. Repeat HIV and syphilis serology was negative 6 weeks after presentation.

Skin biopsies showed a florid lichenoid pattern (Fig 2). Epidermal acanthosis, parakeratosis, diffuse lymphocytic exocytosis, marked spongiosis, focal necrosis, basal vacuolar degeneration, and apoptotic keratinocytes were present. Perivascular infiltrates of lymphocytes with occasional eosinophils were seen in the superficial dermis and periadnexal and perivascular lymphocytic infiltrates in the reticular dermis. Direct immunofluorescence, mycobacterial, and fungal tissue cultures were negative. The infiltrate was $CD3^+$ (T cells) and was positive for the cytotoxic markers beta-F1 (alpha-beta T-cell receptor) and Tia-1 (T-cell restricted intracellular antigen). CD30 and CD56 were negative. A faint monoclonal band was present on PCR for the T-cell receptor gamma gene. A low-level presence of HSV DNA by PCR was detected from skin biopsy tissue from the thigh.

He received acyclovir 400 mg five times a day for 5 days, then a prophylactic dose of 400 mg twice a day. No new skin lesions developed during 6 months of follow-up.

DISCUSSION

FUMHD is a rare, potentially fatal condition frequently associated with systemic symptoms. Prognosis and severity may be related to both age and the presence of T-cell receptor gene clonality. Fatalities have been reported in nine adults between 26 and 82 years of age.[10-17] Severe disease has occurred in paediatric cases,[2,18,19] but no deaths have been reported. Several cases have had milder disease lasting a number of weeks, in which the development of large ulceronecrotic lesions and fever distinguish the condition from typical PLEVA.[2-9]

The results of investigations of clonality have been reported in eight cases.[4,9,14,15,18,20,21] From