

SHORT COMMUNICATION

Febrile Ulceronecrotic Mucha-Habermann Disease in a Young Boy: A Case Report and Review of the Literature

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Accepted Nov 4, 2013; Epub ahead of print Jan 28, 2014

Pityriasis lichenoides et variiformis acuta (PLEVA), also known as Mucha-Habermann disease, is an uncommon, idiopathic, acquired dermatosis characterised by erythematous and scaly papules that are often accompanied by haemorrhagic and papulonecrotic lesions. The patient's general health is usually not affected. Febrile ulceronecrotic Mucha-Habermann disease (FUMHD) is a rare variant of PLEVA that was first described by Degos et al. in 1966 (1). FUMHD is more destructive and is associated with high fever, systemic symptoms and the development of large coalescent skin necroses which may lead to death. Here we report a young boy with FUMHD successfully treated with systemic corticosteroid.

CASE REPORT

A 7-year-old Japanese boy presented to our clinic in October 2008 with scattered eruption on the trunk and limbs. Approximately 20 days before consultation, he noticed an egg-sized red macule on his lower back that was preceded by a sore throat and nasal congestion. Next, scattered red eruptions suddenly appeared on his trunk and limbs within 2 weeks and were accompanied by a persistent high fever. The patient's family and past history were unremarkable. Physical examination revealed numerous discrete, bean-sized, oval-shaped erythemas and papules with central ulcers and necrotic crusts distributed over the patient's face, neck, trunk, and extremities (Fig. 1A). The oral and genital mucosae were not affected. Laboratory examinations including haemogram, biochemistry studies, C-reactive protein analysis, urine analysis, and chest X-ray were all within normal limits. A fungal test and skin bacterial cultures of the lesions were negative. Histological examination of an erythema on the forearm revealed hyperkeratosis, acanthosis, necrotic keratinocytes, and degeneration of the basal layer. Numerous mononuclear cells were infiltrated in the epidermis. There were superficial and deep patchy lichenoid lymphohistiocytic infiltrations that obscured the dermo-epidermal interface in the dermis. There were no lymphocytes with atypia (Fig. 1B). Based on the clinico-pathological findings, the diagnosis of FUMHD was established.

Oral clarithromycin and potassium iodide treatment were initiated, but the skin lesions and high fever did not improve. Three days later, oral prednisolone (20 mg/

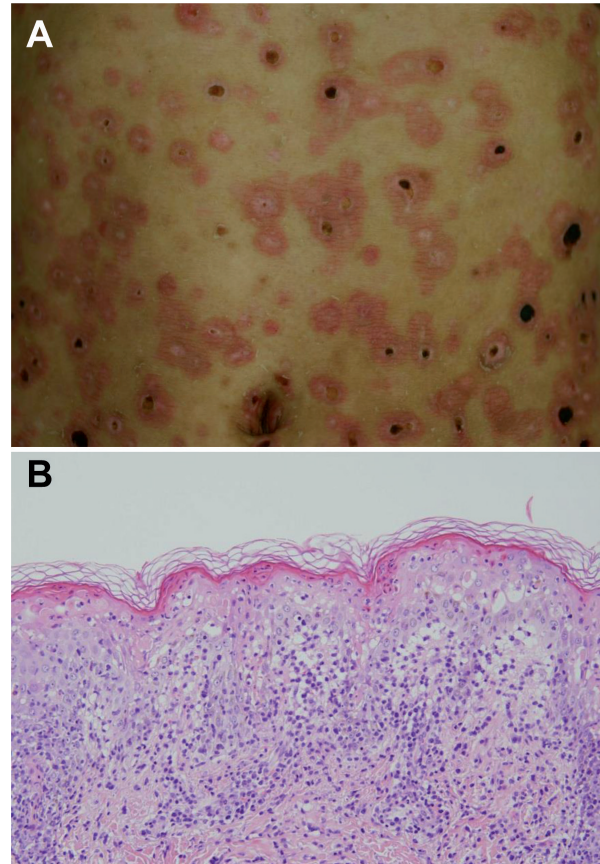


Fig. 1. Multiple erythemas and papules with ulcers and necrotic crusts on the trunk (A). Patchy lichenoid lymphohistiocytic infiltrate obscuring the dermo-epidermal interface (H&E \times 400) (B).

day) was initiated. Most of the necrotic papules disappeared within a week, and the necrotic lesions were healed with scar formation within 3 weeks. The dose of prednisolone was tapered and ended within 2 weeks. No recurrence was observed, and the patient was in good health at a follow-up, approximately 5 years later.

DISCUSSION

FUMHD usually begins as typical PLEVA, as was observed in our case, and rapidly evolves to an ulceronecrotic form with haemorrhagic crusts in 2 to 6 weeks. Oral, genital, and conjunctival mucosae can be affected. Associated symptoms include high fever (up to 40°C), malaise, myalgia, arthralgia, gastrointestinal and cen-

tral nervous system symptoms, interstitial pneumonitis, and lymphocytic myocarditis. A number of fatal outcomes have been reported. To the best of our knowledge, 61 cases of FUMHD have been reported in the literature (2–8). We summarise the reported cases of FUMHD in Fig. 2. Male cases predominated (male:female 43:18). Roughly half of the cases (30 cases) were under 20 years old. We found that the mortality rate of FUMHD is as high as 15% in the cohort as a whole. However, no fatal cases in children have been reported. Interestingly, mortality rates increased with the age of the patients. Fatal cases were confined to older persons, and all cases that occurred in patients over the age of 80 years were fatal. This finding suggests that timely intervention in patients of younger ages may result in a favourable outcome.

Most previously reported cases have been treated with erythromycin and systemic corticosteroids (5). There are other reported treatments, including UV-irradiation, methotrexate, cyclosporine, dapsone, acyclovir and in-

travenous immunoglobulins (IVIg) (2–8). In retrospective analysis, 6 out of 15 cases were successfully treated with systemic corticosteroids (2). Analogously, our patient obtained remarkable improvement after the use of systemic corticosteroids. IVIg appears to be a second-line therapy when corticosteroids are not effective.

The authors declare no conflicts of interest.

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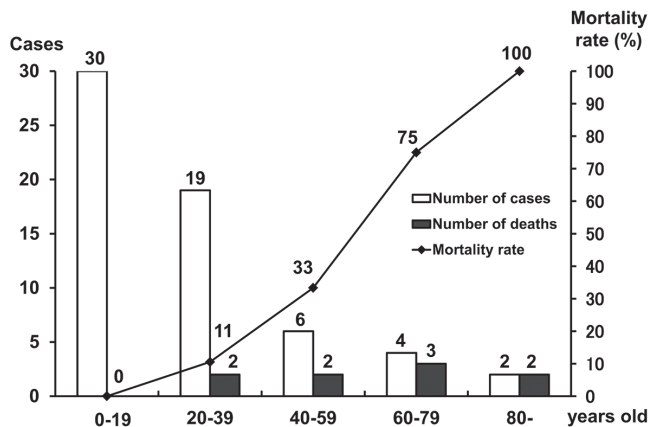


Fig. 2. Number of reported cases in febrile ulceronecrotic Mucha-Habermann disease and mortality rate.