**CASE REPORT**

Febrile ulceronecrotic Mucha–Habermann disease: a case report and a review of the literature

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This report describes the case of a 76 year old man who suffered from febrile ulceronecrotic Mucha–Habermann disease (FUMHD). Despite this patient's typical clinical and histological findings, the fulminating course led to death. Polymerase chain reaction (PCR) analysis of the skin lesions showed that the infiltrating cells were monoclonal in origin and were from an aberrant clone. FUMHD is a very rare, febrile variant type of pityriasis lichenoides et varioliformis acuta, and is characterised by necrotic cutaneous ulcerations associated with high fever and systemic manifestations. Including this present case, only 18 cases of FUMHD have been reported. FUMHD can occur in both adults and children, although there are several differences between the manifestations of the disease in the two groups. One major difference is prognosis: all cases resulting in fatality are of the adult type, whereas no fatal cases have been reported among children. The aberrant clone detected by PCR may be responsible for host responses, resulting in the severe symptoms observed in this disorder.

Pityriasis lichenoides et varioliformis acuta (PLEVA), which is characterised by erythematous papules with little or no scarring, is most often accompanied by few symptoms other than a low grade fever in a few cases. The patient's general health is usually not affected. It occurs mainly in adolescents and young adults, but it is not uncommon in children. However, there is a very rare variant form of PLEVA called febrile ulceronecrotic Mucha–Habermann disease (FUMHD), which has additional symptoms. FUMHD involves a sudden, severe flare up, characterised by innumerable coalescent necrotic ulcerations associated with high fever. Systemic manifestations include interstitial pneumonitis, abdominal pain, malabsorption, central nervous system involvement, and rheumatological manifestations.

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The aetiology of PLEVA has not been elucidated. Some reports have related the pathogenesis of PLEVA to an infectious agent or the deposition of immune complexes. Furthermore, there are some reports on the relation between PLEVA and T cell clonality.

We report the 18th case of FUMHD, which was analysed by means of the polymerase chain reaction (PCR).

**CASE REPORT**

On 4 January 2000, a 76 year old man presented to hospital after having experienced a one week history of high fever (ranging from 39°C to 40°C), vomiting, and skin eruptions. Directly before these symptoms occurred, he suffered an unknown fever lasting four weeks. He had been treated for idiopathic thrombocytosis for over three years without symptoms.

A physical cutaneous examination revealed scattered erythematous papules with little or no scarring. The lesions were round and varied from 1 to 2 cm in diameter. Some bulla and pustules were covered with haemorrhagic and necrotic crusts.

The biopsy of a cutaneous lesion on his trunk showed a relatively well demarcated lesion: a mild acanthosis and subcorneal small necrotic bullous lesions with pronounced subepidermal lymphocyte infiltrate and haemosiderin pigmentation (fig 1A). The basal cell layer showed nuclei of various sizes with a pronounced subepidermal infiltrate of lymphocytes (haematoxylin and eosin stained; original magnification, ×500).

**Figure 1** (A) Mild acanthosis and subcorneal small necrotic bullous lesions with pronounced subepidermal lymphocyte infiltrate (haematoxylin and eosin stained; original magnification, ×60). (B) The basal cell layer showed nuclei of various sizes with a pronounced subepidermal infiltrate of lymphocytes (haematoxylin and eosin stained; original magnification, ×500).

Abbreviations: FUMHD, febrile ulceronecrotic Mucha–Habermann disease; PCR, polymerase chain reaction; PLEVA, pityriasis lichenoides et varioliformis acuta; TCR, T cell receptor
The PCR conditions and the composition of the reaction mixture with four different primer pairs—V/J1, V/J2, D1/J2, or D2/J2—primers—J1 or J2. Each template was amplified separately from three forward primers—V, D1, or D2—and two reverse enzymes, including lactate dehydrogenase (762 to 1044 IU/litre). The T cell receptor (TCR) template. In this method, the rearranged V-D-J portion of the gene is amplified using several primer pairs selected from formalin fixed, paraffin wax embedded sections as a template. In this method, the rearranged V-D-J portion of the TCRβ gene is amplified using several primer pairs selected from three forward primers—V, D1, or D2—and two reverse primers—J1 or J2. Each template was amplified separately with four different primer pairs—V/J1, V/J2, D1/J2, or D2/J2. The PCR conditions and the composition of the reaction mixtures were the same as described previously. In each experiment, templates from a cutaneous lymphoma and granulation tissue were included as controls for monoclonality and polyclonality, respectively.

The PCR result with the primer pairs D1/J2 and D2/J2 showed a distinct band of the same size (fig 2, lane 3), whereas amplification with V/J1 or V/J2 showed a smear similar to the polyclonal control. Laboratory studies revealed raised C reactive protein (186 mg/litre) and a mild increase in lactate dehydrogenase. Treponema pallidum haemagglutinin test, hepatitis B surface antigen tests, and blood cultures for bacteria and fungi were negative. These clinical and histological findings suggested a diagnosis of FUMHD.

Ten days of treatment with the antibiotics pentocilin and sulperazon resulted in an improvement in the skin lesions and had a slight effect on the patient’s general condition by eliminating fever. However, on January 15, the patient suddenly developed hypovolaemic shock. After receiving a transfusion he recovered, but became increasingly weak, with a high fever. However, there were no relapsing skin eruptions. Chest x ray revealed ground glass opacity, and laboratory studies revealed raised white blood cell counts (9800–16 100/mm³) and liver enzymes, including lactate dehydrogenase (762 to 1044 IU/litre). He was treated with sulperazon and minocycline, but died 10 days later. No necropsy was performed.

**DISCUSSION**

In this patient, the cutaneous eruptions showed erythematos, purplic papular, and vesiculobullous lesions with haemorrhagic and necrotic crusts. These are all typical histological findings of PLEVA. The cutaneous eruptions were not multiple, but were scattered over the entire body. These eruptions, accompanied by a high fever and vomiting, occurred suddenly. As a result of these clinical and histological findings, we diagnosed the patient as suffering from FUMHD. We treated him with antibiotics, which were effective against the cutaneous eruptions and high fever for the initial 10 days. However, he developed hypovolaemic shock, which worsened and, eventually, led to death.

To our knowledge, only 17 cases of FUMHD have been reported previously.¹⁻⁷ Twelve of the patients were children or young adults (under 40 years). Recently (including our case), cases involving increasingly older patients have been reported. Although FUMHD can occur in both adults and children, there are several differences. One of the major differences is prognosis: a fatal outcome has been seen in adults, but there have been no child fatalities.¹ Five of 10 adult patients died at some point between eight days and seven months during follow up treatment. In addition, there were six cases involving patients over 40 years old.¹⁻⁷ These findings strongly suggest that the adult type of FUMHD has a high malignant potential, and this may be related to PLEVA and T cell clonality.¹⁻²

"This disorder may contain an aberrant clone, which has failed to proceed to T-DJ rearrangement after DJ rearrangement"
responsible for host responses, resulting in severe symptoms and poor outcome in the adult type of this disorder. 

There is a clear distinction between the adult and childhood types of FUMHD. However, because so few FUMHD cases have been reported, further investigation is necessary to confirm whether the two types are completely distinct disorders.

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