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CLINICAL REPORT

Febrile Ulceronecrotic Mucha-Habermann Disease: A Case Report and Review of the Literature

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Febrile ulceronecrotic Mucha-Habermann disease is a severe variant of *pityriasis lichenoides et varioliformis acuta* characterized by the sudden onset of ulceronecrotic skin lesions and associated with high fever and systemic symptoms. We report here a case of a 20-year-old woman in whom the disease started as *pityriasis lichenoides et varioliformis acuta* and evolved to febrile ulceronecrotic Mucha-Habermann disease. Almost 90% of the body surface was involved, together with high fever and malaise. Steroids alone proved to be an insufficient therapeutic procedure. The remission achieved was attributed to the use of methotrexate. To our knowledge, only 39 cases of febrile ulceronecrotic Mucha-Habermann disease have been reported in the literature to date. *Key words: febrile ulceronecrotic Mucha-Habermann disease; pityriasis lichenoides and varioliformis acuta.*

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Febrile ulceronecrotic Mucha-Habermann disease (FUMHD) is a severe variant of *pityriasis lichenoides et varioliformis acuta* (PLEVA), first described by Degos et al. in 1966 (1). To the best of our knowledge, 39 cases of FUMHD have so far been reported in the literature in English (1–36). We describe here a 20-year-old woman with FUMHD who was treated successfully with methotrexate and we review all cases reported in the literature.

CASE REPORT

The patient was a 20-year-old woman with FUMHD. The disease started as classic PLEVA with the abrupt onset of a polymorphic pruritic skin eruption comprising scattered erythematous, purpuric papules and vesicles on the trunk and extremities. The condition was misdiagnosed as chickenpox and treated with acyclovir, antibiotics and topical steroids by general practitioners.

However, the skin lesions consecutively disseminated. One week later, in October 2006**,** when she was referred to our hospital more than 90% of her body surface was involved, leaving the patient susceptible to bacterial infections (Fig. 1).

No personal or family history of dermatological diseases was reported. She denied receiving any medications, she had no sexual contact and no drug or food allergies.

Physical examination revealed a generalized skin eruption comprising necrotic papules and pustules with thick haemorrhagic crusts, round erosions and ulcers from several millimetres to several centimetres in diameter. The lesions presented in different evolutionary stages. There was extensive and extremely painful skin necrosis with a purulent exudate over flexural sites such as the neck, axilla, antecubital, umbilical and inguinal areas. A small number of pustules and tense bullae were present on the palms and soles. Oral mucosa was also involved, with painful fissures located on the dorsal surface of the tongue. The patient had high fever (39.5°C) with chills, malaise and myalgia.

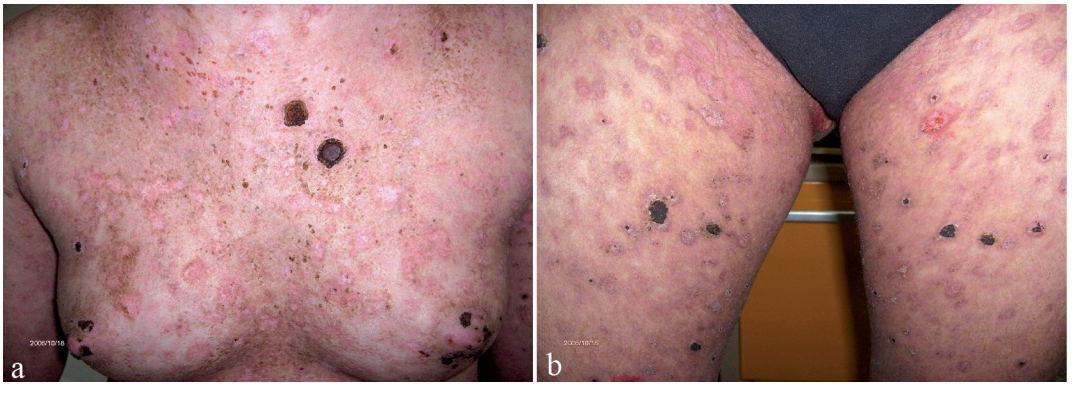

Fig. 1. Disseminated necrotic papules, round erosions and ulcers varying from 1 to 2 cm in diameter.

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Routine laboratory tests showed elevated erythrocyte sedimentation rate, increased levels of C-reactive protein and a normochromic-normocytic anaemia. Serology for hepatitis B virus, hepatitis C virus, HIV, Epstein-Barr virus (EBV), cytomegalovirus, varicella-zoster virus, human herpes virus 6,7,8, as well as *Treponema pallidum* haemagglutination test were negative. Serology for herpes simplex virus was negative for IgM but positive for IgG, suggesting a past infection. Skin cultures detected *Staphylococcus epidermidis* and *Pseudomonas aeruginosa*.

Histology of a cutaneous lesion of her right forearm revealed basket-weave hyperkeratosis with a few nuclear fragments derived from polymorphonuclear cells, focal parakeratosis and preservation of the granular layer (Fig. 2). In the malpigian layer there were areas of spongiosis accompanied by exocytosis of lymphocytes. The dermis showed a perivascular lymphocytic infiltrate without evidence of vasculitis.

Initial treatment with oral corticosteroids (prednisolone 40 mg/day) for 4 days proved insufficient to halt disease progression. Intravenous administration of ticarcillin-clavulanic acid 5.2 g 3 times a day and cloxacillin 1.5 g 4 times a day was based on skin cultures. Despite the above treatment new skin lesions appeared. Intravenous methotrexate was initiated on a weekly regimen of 15 mg together with folic acid with a simultaneous decrease in prednisolone to 20 mg per day. The patient also received supportive care including a high-calorie parenteral diet, as well as meticulous nursing care with topical mupirocin, whirlpool and sodium permanganate baths.

The patient showed a dramatic response within 10 days; she became afebrile and her general condition improved significantly. No new lesions appeared, with progressive re-epithelization of the existing ones. Administration of methotrexate and prednisolone lasted 2 months.

Relapse occurred 7 months after the original eruption and new papulosquamous lesions developed at the lateral sides of the neck, the infra-mammary area and periumbilically with the formation of erosions and ulcers at the axillary and popliteal fossa. Methotrexate per os was restarted, albeit at a lower dosage (10 mg/week), coupled with prednisolone 20 mg/day. The skin condition started to improve after 5 days, so the prednisolone was subsequently tapered off until discontinuation.

At present, almost a year after the patient's initial presentation, she remains on methotrexate 10 mg weekly. Her general condition is excellent apart from the significant hyperpigmentation and the varioliform-atrophic hypopigmented scars at the sites of previous ulcerations (Fig. 3)

DISCUSSION

FUMHD occurs more frequently in children, adolescents or young adults than in older subjects (29). It exhibits a male predominance (male:female ratio 27:14). The mean age of patients in the reported cases is 27.4 years (range 4–82); 30 of the 40 patients reported were younger than 35 years at the time of presentation (Table I).

The disease may either start with a mild form of skin changes consistent with PLEVA that evolve later to large coalescent ulceronecrotic lesions, or it may

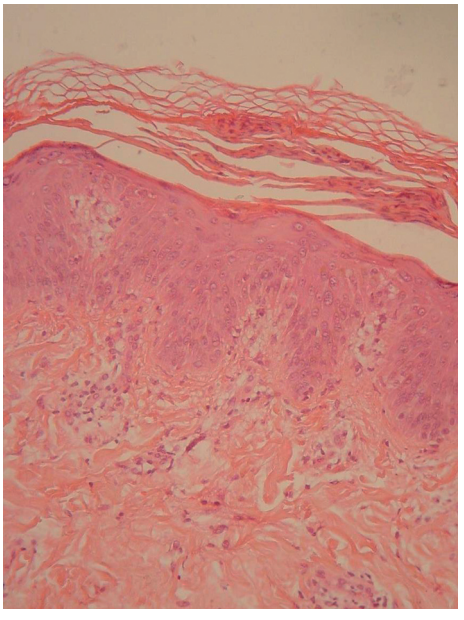

Fig. 2. Focal parakeratosis, areas of spongiosis accompanied by exocytosis of lymphocytes. The dermis shows a perivascular lymphocytic infiltrate without evidence of vasculitis (haematoxylin and eosin ×40).

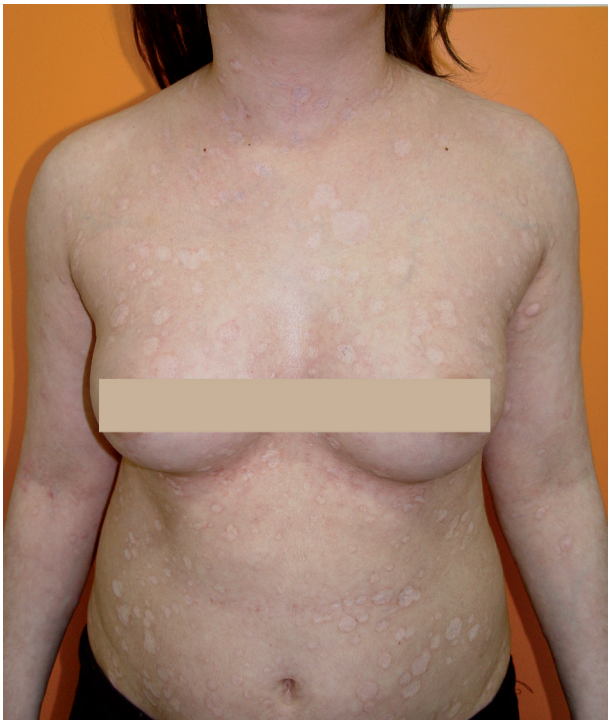

Fig. 3. Significant hyperpigmentation and varioliform – atrophic hypopigmented scars at the sites of previous ulcerations.

have an acute onset with large papulonecrotic and haemorrhagic lesions all over the body (28). The ulcerated lesions often become secondarily infected and tend to heal with hypertrophic scars (23). Oral, genital and conjunctival mucosae can be affected (37). The disease is associated with high fever; up to 40ºC. Associated systemic manifestations in the reported cases include gastrointestinal (1, 6, 15, 26, 27, 31, 34, 36), neurological (1) and pulmonary involvement (4, 12, 31, 32, 34), cardiomyopathy (3, 8), rheumatological manifestations (9, 20), megaloblastic anaemia (6, 12, 26), pancytopaenia (28) and diffuse intravascular coagulation (31).

The reported mortality rate of FUMHD is approximately 20% (9 of 40 patients). There have, so far, been no child fatalities (27). Considering the fact that the mortality rate in children (0–14 years) is zero, this rate in adult raises to 33.3%. A comparison between adult and paediatric cases shows a more favourable outcome in children (24). In fact, fatal cases were confined to persons older than 40 years, until 2004–05 when Cozzio et al. (28) and Aytekin et al. (30) reported cases of 26–27-year-old females with fatal outcome.

In the cases with fatal outcome death was attributed to pulmonary thromboembolism (8, 34), pneumonia (12), cardiac arrest (15), sepsis (18, 28, 30, 34), hypovolemic shock (27) and massive thrombosis of the superior mesenteric artery (34).

The aetiology of PLEVA remains controversial. A hypersensitivity reaction to an infectious agent is suggested to be the main cause (1, 6, 20, 38). Many different infectious agents, such as EBV (22, 24, 39), adenovirus (4), and cytomegalovirus (20) have been implicated in the pathogenesis of the disease, but there has been no consistent finding so far. In a case described by Yanaba et al. (23), a predominantly CD8+ lymphocyte infiltrate around the dermis and into the epidermis might suggest cytotoxic attack of lymphocytes to altered epidermal antigens, perhaps induced by an unknown infectious agent.

Recent studies have demonstrated PLEVA to be a benign disorder of activated T-cell lymphocytes. Dereure et al. (40) concluded that the disease exists within a spectrum of clonal T-cell cutaneous lymphocytic disorders, while Weinberg et al. (41) found that 57% of their specimens demonstrated monoclonal T-cell receptor gene rearrangements. These findings lead to the conclusion that PLEVA is a benign clonal T-cell disorder. Additional evidence to this hypothesis was presented by Magro et al. (42). Cases with fatal course revealed clonal TCR rearrangement, but it remains unclear whether TCR clonality stands for the predominance of a pre-neoplastic lymphocyte clone or if it represents a clonal cytotoxic T-lymphocyte reaction against a supposed intracellular pathogen (33).

Histopathological findings in most of the presented cases include the typical features of PLEVA and different grades of leukocytoclastic vasculitis (1–38)

Several treatment modalities have been described, but no definite therapy can be recommended for all patients at present (23, 32). All reported patients received multiple drugs including systemic steroids, methotrexate, antibiotics, psoralen + ultraviolet A (PUVA), ultraviolet B (UVB), unspecified ultraviolet receptor, acyclovir, immunoglobulins and 4,4-diaminodiphenylsulphone (DDS) (see Table I). The effectiveness of these treatment modalities is difficult to assess as they failed to achieve remission in patients with eventually fatal outcome.

Therapeutic efficacy of systemic steroids is highly controversial. Twenty-seven of 40 patients (including the present case) were treated with steroids, but the only positive effect was reported by Degos et al. (1). Methotrexate has been used in 15 patients with FUMHD (including ours). It induced rapid remissions and was successful in cases that did not respond to other therapeutic measures. Four patients died despite methotrexate therapy, but this might be attributed to its late institution (8, 15, 28).

Successful treatment with a short course of oral cyclosporine in 2 cases of an 8-year-old boy and a girl of the same age has been reported (31, 35), as well as a case that required debridement of necrotic skin and placement of epidermal autografts (23). Treatment with tumour necrosis factor (TNF)-α inhibitors might represent a first-line option in future cases, as elevated levels of serum TNF-α have been reported in FUMHD cases (29). Furthermore, intensive care, treatment of superinfection and maintenance of the patient's general condition is required.

In conclusion, considering the severity of this disease, which can be fatal, it is of great importance to make the diagnosis at an early stage, to commence treatment as soon as possible, to monitor patients closely and, if necessary, to transfer them to intensive care.

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Table 1. *Literature review*

Reference	Age/Sex	Histology	Mucosal involvement	Systemic involvement	Therapy	Outcome
Degos et al. 1966 (1)	36/M	Fibrinoid necrosis, nuclear pyknosis	Unknown	CNS involvement, myalgias, lymphadenopathy	SS, ATB	Cure
Degos et al. 1966 (1)	30/M	Lymphocytic vasculitis, no leucocytoclasia	Unknown	Abdominal pain, generalized lymphadenopathy	ATB	Cure
Burke et al. 1969 (2)	15/M	Leucocytoclasia	+	Lymphadenopathy, anaemia	ATB	Cure
Burke et al. 1969 (2)	12/F	Leucocytoclasia	-	-	SS	Cure
Lowe 1975 (3)	32/M	Perivascular lymphocytic infiltration, no vasculitis	Unknown	Cardiomyopathy	UVR	Cure
Auster et al. 1979 (4)	7/F	Leucocytoclastic vasculitis	Unknown	Pulmonary involvement	ATB	Cure
Cavalieri et al. 1982 (5)	13/M	Perivascular lymphocytic infiltration	Unknown	-	SS, ATB, antivirals, IVIG	Cure
Warshauer et al. 1983 (6)	54/M	Perivascular lymphocytic infiltration, no vasculitis	+	Abdominal pain, megaloblastic anaemia, eosinophilia	MTX ATB, SS, thiabendazol	Cure
Nakamura et al. 1986 (7)	21/M	Lymphocytic vasculitis, little leucocytoclasia	Unknown	-	DDS, ATB	Cure
Hoghton et al. 1989 (8)	49/F	Perivascular lymphomonocytic infiltration	-	Splenomegaly, myocarditis, mesenteric lymphadenopathy, pulmonary embolism	SS, ATB, DDS, MTX, UVR, acyclovir	Fatal
Luberti et al. 1991 (9)	12/M	Leucocytoclastic vasculitis, interface dermatitis	+	Arthritis, sepsis	SS, ATB, acyclovir	Cure
Kasamatsu et al. 1993 (10)	4/M	Leucocytoclastic vasculitis	Unknown	Lymphadenopathy	SS, ATB, DDS, antivirals	Cure
Lopez-Estebaranz et al. 1993 (11)	18/M	Leucocytoclastic vasculitis	-	Liver dysfunction	SS, ATB, MTX, PUVA	Cure
De Cuyper et al. 1994 (12)	82/F	Leucocytoclastic vasculitis	+	Megaloblastic anaemia, pneumonia, malabsorption	SS, ATB, UVB	Fatal
Fink-Puches et al. 1994 (13)	16/M	Leucocytoclastic vasculitis	+	-	SS, ATB, MTX	Cure
Maekawa et al. 1994 (14)	16/M	Perivascular lymphocytic infiltration	-	Eosinophilia	ATB, acyclovir	Cure
Gungor et al. 1996 (15)	59/M	Perivascular lymphocytic infiltration, no vasculitis	Unknown	Abdominal pain, diarrhoea	SS, MTX, ATB	Fatal
Reseghetti et al. 1996 (16)	27/M	Leucocytoclastic vasculitis	Unknown	Staphylococcus aureus bacteremia	SS, PUVA, ATB	Cure
Suarez et al. 1996 (17)	32/M	Perivascular lymphocytic infiltration	+	Sepsis, impaired gait, conjuctival ulcer, liver dysfunction	SS, MTX, ATB	Cure
Puddu et al. 1997 (18)	43/F	Dense lymphocytic and neutrophilic infiltration, necrotic keratinocytes	Unknown	Sepsis	SS, ATB	Fatal
Romani et al. 1998 (19)	12/F	Deep dermal lymphohistiocytic infiltration	Unknown	-	MTX, PUVA, ATB	Cure
Tsai et al. 2001 (20)	45/M	Thrombonecrotic vasculopathy	Unknown	Arthritis	SS, UVB, ATB, acyclovir	Cure
Hsieh et al. 2001 (21)	8/F	Unknown	-	-	ATB, UVB, acyclovir	Cure
Ricci et al. 2001 (22)	10/F	Spongiosis, vacuolar alteration, neutrophilic dermal infiltration	-	-	Acyclovir	Cure
Yanaba et al. 2002 (23)	21/M	Dense perivascular lymphocytic infiltrate	-	Sepsis	SS, DDS, ATB, FFP, IVIG, skin grafting	Cure
Yang et al. 2003 (24)	14/M	Perivascular lymphocytic infiltration, no vasculitis, lichenoid infiltration of upper dermis	+	-	SS, ATB	Cure
Rivera et al. 2003 (25)	33/F	Perivascular-periadnexal lymphocytic infiltration	+	-	SS, MTX, acetylsalicylic acid	Cure
Ito et al. 2003 (26)	12/M	Exocytosis of lymphocytes, vacuolar alteration, perivascular – periadnexal lymphocytic infiltration, no vasculitis	-	Anaemia, abdominal pain, liver dysfunction, lymphadenopathy	SS, MTX	Cure
Miyamoto et al. 2003 (27)	76/M	Lymphocytic perivascular infiltration	-	Hypovolemic shock, emesis	ATB	Fatal
Cozzio et al. 2004 (28)	72/M	Lichenoid mononuclear infiltration, exocytosis of atypical lymphocytes	-	Pancytopenia, sepsis	MTX, IVIG	Fatal
Cozzio et al. 2004 (28)	26/F	Massive dermal infiltrate, exocytosis of CD8+ lymphocytes	-	Sepsis	SS, MTX, PUVA	Fatal
Tsianakas & Hoeger 2005 (29)	9/M	Dense perivascular lymphocytic infiltration, leucocytoclastic vasculitis, exocytosis	-	-	SS, ATB, MTX	Cure

Table I contd.

Aytekin et al. 2005 (30)	27/F	Subcorneal necrotic bullous formation,exocytosis of lymphocytes	-	Sepsis	SS, ATB, acyclovir, IVIG, FFP	Fatal
Herron et al. 2005 (31)	8/F	Dense lymphocytic infiltration, CD30+ lymphocytes in dermis-epidermis	-	ARDS, DIC, GI haemorrhage, sepsis	MTX, CyA, ATB	Cure
Aydingoz et al. 2006 (32)	37/M	Perivascular lymphohistiocytic infiltration, no vasculitis	-	Pulmonary involvement	SS, ATB	Cure
Helmbold et al. 2006 (33)	20/M	Perivascular-interstitial lymphocytic infiltration, epitheliotropism	-	-	SS, ATB, acyclovir	Cure
Malnar et al. 2006 (34)	60/M	Lichenoid lymphocytic infiltration, vacuolar degeneration of DEJ	+	Sepsis, pulmonary thromboembolism, massive thrombosis of superior mesenteric artery (GI gangrene)	SS, ATB	Fatal
Kim et al. 2007 (35)	8/M	Spongiosis , exocytosis of inflammatory cells, focal epidermal haemorrhage	-	-	SS, CyA, ATB	Cure
Pyrpasopoulou et al. 2007 (36)	17/F	Unknown	Unknown	Sepsis, diarrhoea, anaemia	SS, MTX, ATB, IVIG, acyclovir	Cure
Present case 2007	20/F	Spongiosis, exocytosis of lymphocytes, perivascular lymphocytic infiltration, no vasculitis	+	-	SS, MTX, ATB, acyclovir	Cure

M: male; F: female; CNS: central nervous system; SS: systemic steroids; ATB: antibiotics; MTX: methotrexate; CyA: cyclosporin A; IVIG: intravenous immunoglobulin; FFP: fresh frozen plasma; DDS: 4,4-diaminodiphenylsulphone; UVR: ultraviolet radiation; UVB: ultraviolet B; PUVA: psoralen plus ultraviolet A; ARDS: acute respiratory distress syndrome; DIC: diffuse intravascular coagulation; GI: gastrointestinal; DEJ: dermo-epidermal junction

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