Abstract
Febrile ulceronecrotic Mucha-Habermann disease (FUMHD), a severe form of pityriasis lichenoides et varioliformis acuta, is an inflammatory dermatosis of unknown etiology manifested by ulcerative and necrotic lesions accompanied by systemic manifestations. The mortality rate of FUMHD is about 15%. It is reported here a case of FUMHD presenting as toxic epidermal necrolysis that resulted in multiple organ failure and death.

Key Words: Febrile ulceronecrotic Mucha-Habermann disease, skin, toxic epidermal necrolysis

Introduction
Febrile ulceronecrotic Mucha-Habermann disease (FUMHD) disease is a rare variant of pityriasis lichenoides et varioliformis acuta (PLEVA). It characterized by ulcerative and necrotic lesions accompanied of systemic findings. The exact pathogenesis of FUMHD still remains unknown, and there is no exact and effective treatment. A total of 69 cases, including the case reported here, have been described to date with 11 reported fatalities. In this report, we present a case of FUMHD who had no response to systemic steroids and immunoglobulin.

Case Report
A 43-year-old man presented to our hospital with a 2-week history of progressive erythematous macules and papules eruption. He had been healthy without any systemic diseases. None in his family had similar symptoms. There was no history of medication use and drug or food allergies. Dermatological examination revealed widespread erythematous macules, papules, and papulovesicles, some with overlying crusts, over his trunk and extremities [Figure 1]. There was no oral mucosal involvement. Laboratory tests revealed normal complete blood count, erythrocyte sedimentation rate, C-reactive protein, and biochemistry values. Serologies for hepatitis B virus, hepatitis C virus, hepatitis D virus, hepatitis E virus, HIV, cytomegalovirus (CMV), rubella, herpes simplex virus, herpes zoster virus, Epstein–Barr (EB) virus as well as rapid plasma reagin, and Treponema pallidum particle agglutination assay tests were negative. The skin biopsy [Figure 2] taken from the abdomen revealed parakeratosis, spongiosis, dense perivascular lymphocytic infiltrate extending throughout the dermis without atypia, and extravasation of erythrocytes. Monoclonal T-cell receptor (TCR) rearrangements were detected in the skin biopsy. Based on the aforementioned clinical and histopathological findings, we made the diagnosis of PLEVA and started oral minocycline hydrochloride 100 mg two times a day; empiric antimicrobial coverage was added, including cefotaxime sodium 2.0 g intravenous, two times a day and start on systemic corticosteroid (methylprednisolone 40 mg/day). However, lesions expanded gradually to involve the entire trunk and extremities. Blisters and pustules also occurred. The eruption was associated with fever (up to 39.3°C)

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How to cite this article: Xing C, Shen H, Xu J, Liu Z, Zhu J, Xu A. A fatal case of febrile ulceronecrotic mucha-habermann disease presenting as toxic epidermal necrolysis. Indian J Dermatol 2017;62:675.

Received: October, 2016. Accepted: August, 2017.
on the 8th day of treatment, together with an alanine transaminase (ALT) value of 91 U/L (reference range, 9–50 U/L), and the skin and blood culture were positive for *Escherichia coli*. Skin examination [Figures 3 and 4] found diffuse erythema, plaques, hemophysislitis, pustules, ulcer on the trunk and extremities, some of the pustules confluent, had Nikolsky’s sign positive; flexural accentuation of the lesions was noticed in the axillae, groin, and neck. The rash presented as toxic epidermal necrolysis. And then, the methylprednisolone was increased to 80 mg daily, the patient continued to decline clinically, and a repeat skin biopsy was performed. A second skin biopsy [Figure 5] revealed an increasingly dense perivascular lymphocytic infiltration, leukocytoclastic vasculitis, and epidermal necrosis. In light of the clinical presentation and the consistent histopathology, the diagnosis of PLEVA fulminans was made. Intravenous immunoglobulin (IVIG) (30 g/day) was initiated and methylprednisolone dose was increased to 120 mg/day for 3 days. Skin and blood culture was positive for *E. coli*; antibiotic regimen was modified accordingly. Systemic vancomycin and gentamicin, imipenem were given. Despite this treatment, he continued to deteriorate. Over the next few days, there was intermittent fever (up to 39.3°C), liver enzymes continue to rise (ALT up to 376 U/L), and leukocyte count decreased (3.1 × 109/L). The patient’s condition continued to deteriorate and finally died of multiple organ failure caused by sepsis.

**Discussion**

FUMHD, first defined by Degos et al. in 1966, is a severe variant of PLEVA. It is characterized by rapid progression of necrotic papules to destructive ulceronecrotic lesions, accompanied by high fever and systemic findings. The period necessary for evolution of PLEVA to FUMHD varies from a few days to a few weeks. A total of 69 cases, including the case reported here, have been described to date with 11 reported fatalities. The mortality rates increased with the age of the patient. There have been only one case of a child fatality reported so far. Fatal outcomes have been attributed to pulmonary thromboembolism, pneumonia, sepsis, hypovolemic shock, cardiac arrest, and thrombosis of superior mesenteric artery. The etiology of this disease is unknown, may be related to infectious antigens (such as EB virus, adenovirus,
CMV). Because there is T-cell infiltration in the skin lesions, some scholars have suggested that FUMHD is also a T-cell proliferative disease, and individual cases can be developed into cutaneous T-cell lymphoma. It is suggested that monoclonality of T-cells might increase the transition of PLEVA to FUMHD and can be considered as an indicator of severity and unfavorable outcome. In our case, the patient’s TCR gene rearrangement was positive; maybe, patients with TCR gene rearrangement positive should be paid enough attention. Although systemic steroids, IVIG is considered to be effective in some reports, our patient did not respond well to these treatment measures. We speculate that large doses of steroids lead to impaired immunity and overwhelming infection ending with sepsis. Both humoral and cellular immunity are involved in the pathogenesis, but IVIG only plays a role of inhibition of humoral immunity by neutralizing antibodies. Methotrexate, among the recovered cases described so far, seems to be the most successful therapy. Because of liver dysfunction, we missed the opportunity to use methotrexate. Early intervention with methotrexate may be particularly useful; however, the treatment of FUMHD is still a challenge, and its optimum treatment remains to be determined. Therefore, more case reports and treatment experience are needed to help establish an ideal approach for its management.

**Financial support and sponsorship**
Nil.

**Conflicts of interest**
There are no conflicts of interest.

**What is new?**
The exact pathogenesis of FUMHD is unknown. Although various treatment options have been tried, treatment efficacy is difficult to determine because of the small number of reported cases. Systemic steroids and IVIG is considered to be effective in some reports, but in this case, our patient did not respond well to these treatment measures. More case reports and treatment experience are needed to help establish an ideal approach for its management.

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