Case Report

A case report of febrile ulceronecrotic Mucha-Habermann disease

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Background: Febrile ulceronecrotic Mucha-Habermann disease (FUMHD) is a rare and severe variant of pityriasis lichenoides et varioliformis acuta, characterized by a rapid onset of painful, necrotic skin lesions and systemic symptoms. The diagnosis of FUMHD is complex, hinging on the clinical presentation, histopathological findings, and exclusion of other severe dermatoses. The key diagnostic criteria include sudden development of ulceronecrotic papules and plaques, fever, and evidence of systemic disease. Due to the rarity of FUMHD, there is no consensus on optimal treatment, reflecting a significant gap in the dermatological practice.

Case Description: This report details a multimodal approach tailored to our 13-year-old patient, incorporating systemic corticosteroids, immunosuppressive therapy, and intensive supportive care. The strategy was designed to address the acute and aggressive nature of the disease while mitigating potential systemic complications. The report emphasizes on the intricate, multi-layered care required to manage FUMHD, illustrating the challenges and considerations in treating this complex condition. It underscored the necessity of a personalized, comprehensive care plan that extends beyond medical intervention to include psychological and social support. The outcome of our patient was encouraging, with a marked reduction in cutaneous manifestations and improvement in systemic symptoms.

Conclusions: It was found that prevention and care of skin injuries and complications, as well as the protection of patient's mental state during the development of the disease, are very important. Therefore, early diagnosis, prompt treatment, close monitoring of infection indicators, and specialized care are essential to improve the prognosis of patients with FUMHD.

Keywords: Febrile ulceronecrotic Mucha-Habermann disease (FUMHD); children; nursing; case report

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Introduction

Pityriasis lichenoides et varioliformis acuta (PLEVA) is a rare erythematous scaly skin disease of uncertain origin (1). It predominantly affects children and adolescents and its etiology remains elusive. The primary manifestations consist of inflammatory papules and papulovesicle of varying durations, accompanied by few systemic symptoms, and minimal scarring after resolution of the skin lesions (2). Febrile ulceronecrotic Mucha-Habermann disease (FUMHD), categorized as a severe variant, is extremely uncommon (3). It progresses rapidly, leading to extensive skin lesions primarily characterized by necrotic ulcers. Patients may experience high fever, headache, abdominal pain, anemia, hypoalbuminemia, and systemic

Figure 1 Large area of burst and erosion on the whole body, hip with effusion.

manifestations such as abnormal liver function, posing a potential risk of death. Due to its rarity, effective treatments for this disease have not been established (4). Diagnostic criteria for FUMHD primarily rely on histological examination, revealing characteristic features such as lymphocytic infiltration and epidermal necrosis. The rarity of FUMHD limits evidence from randomized controlled trials, resulting in an unclear consensus on treatment options.

The etiology of FUMHD is unknown, but it may be associated with exogenous antigens, including infectious agents [such as Epstein-Barr (EB) virus, adenovirus, cytomegalovirus], and drug reactions (5). Here, we present a case report of a patient with FUMHD admitted to our hospital. We present this article in accordance with the CARE reporting checklist (available at https://tp.amegroups.com/article/view/10.21037/tp-23-520/rc).

Highlight box

Key findings

- Prevention and care of skin injuries and complications, as well as protection of patient's mental state during the development of the disease are very important.

What is known and what is new?

- The clinical symptoms of febrile ulceronecrotic Mucha-Habermann disease (FUMHD) have distinct manifestations.
- Specialized care is essential to improve the prognosis of patients with FUMHD.

What is the implication, and what should change now?

- Early diagnosis, prompt treatment, close monitoring of infection indicators, and specialized care are essential to improve the prognosis of patients with FUMHD.

Case presentation

Clinical presentation and clinical course

The previously healthy 13-year-old boy experienced initial symptoms, including scattered erythema and papules, 19 days prior to presentation without any causes. This was accompanied by partial ulceration, scabbing, and mild itching (*Figure 1*). Oral administration of loratadine and levocetirizine did not result in significant improvement. Thirteen days prior to presentation, the patient experienced recurrent fever, peaking at a temperature of 40.2 ℃. Blood tests revealed normal white blood cell count, with monocyte count of 15.7%, positive immunoglobulin G (IgG) antibodies for EB virus, as well as elevated levels of cytomegalovirus and herpes simplex virus IgG. Lung computed tomography (CT) scans showed no abnormalities. Ten days before flushing, there were signs of partial erosion, scabbing, desquamation, and pain on the trunk and limbs. Additionally, perioral and periocular erythema accompanied by repeated high fever were observed. Intravenous treatment with vidarabine and methylprednisolone for 3 days failed to effectively control the patient's condition. Subsequent intravenous administration of gamma globulin for 5 days did not alleviate the fever, which fluctuated between 37.0 and 40.3 ℃. Large areas of skin flushing, desquamation, ulceration, erosion, and exudation were observed on the armpit, elbow fossa, popliteal fossa, waist, and hip. Some areas exhibited black necrotic scabs alongside new rashes. In this case, the patient did not require inotropic support and was subjected to mechanical or non-invasive ventilation, reflecting the severity and management needs of his condition. The patient successfully survived and was discharged from the Pediatric Intensive Care Unit (PICU) after 120 days, with a total hospital stay of 240 days. All procedures performed in this study were in accordance with

Figure 2 Febrile ulceronecrotic Mucha-Habermann disease. Generalized ulceronecrotic papules and plaques covering the whole trunk.

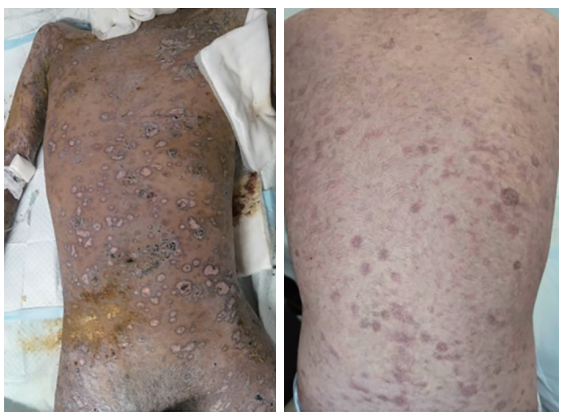

Figure 3 Decrustation and healing of most of the whole body skin.

the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient's legal guardian for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal. Post-treatment, the patient showed significant recovery of skin function, with no relapses observed during the follow-up period. This outcome underscores the importance of tailored, responsive care in managing complex cases of FUMHD (*Figure 2*).

Diagnostic tests

Extensive erosion was noted in the bilateral armpits, elbow pits, popliteal pits, and buttocks, along with obvious pain. However, no oral mucosa ulceration or erosion was observed. After consulting with multiple disciplines, the patient was transferred to the PICU due to the severity and poor control of the condition, which manifested through recurrent high fever, elevated inflammatory markers, hypoalbuminemia, hyponatremia, and abnormal liver function. During the PICU stay, repeated signs of pseudomonas aeruginosa infection were observed from skin secretion cultures, catheter cultures, and blood cultures. *Baumanniella* infection was also identified through catheter cultures and blood cultures. Meropenem and polymyxin were administered sequentially to combat the infections. The diagnosis of FUMHD in our patient was established through a comprehensive evaluation, including a detailed histopathological examination which confirmed the presence of specific histological features. This biopsy-proven approach underscores the rigor of our diagnostic process and aligns with the recognized criteria for FUMHD treatment.

Medical treatment

Additionally, fungal infection was detected through blood cultures. Following consultations with the pharmacy and laboratory specialists, antifungal therapy involving fluconazole, voriconazole, caspofungin, and flucytosine was initiated. Anti-inflammatory treatment with methylprednisolone and ulinastatin was also initiated. Fentanyl was used for pain management, while dexmedetomidine was administered for sedation. Supportive treatments included albumin, red blood cell suspension, and gamma globulin. The patient underwent therapeutic plasma exchange (TPE) four times and received continuous blood purification (CBP) for 8 days, resulting in a significant decrease in inflammatory markers. The levels of interleukin (IL)-8 decreased from 304 to 122 pg/mL, IL-1β dropped from 9.05 to <5 pg/mL, and IL-2 receptor decreased from 5,000 to 2,141 U/mL. After 23 days of treatment, most of the skin on the boy's body had healed and the crusts had disappeared (*Figure 3*) and the patient was transferred to a general ward for further care.

Discussion

Skin care

The patient was presented with necrotic black scabs on the head, face, trunk, and limbs, as well as chapped and

desquamated skin wounds on the back. Bilateral armpits, cubital fossa, groin, popliteal fossa, buttocks, and waist showed erosive surfaces with a small amount of exudation, with severe lesions on the groin and armpits requiring careful nursing. The following measures were taken:

(I) Skin cleaning: sterile press-type scrub pads were used twice a day to wet skin lesions with saline water, cleanse natural loss of necrotic tissue and wound exudate. Chlorhexidine acetate solution was poured into a watering can and sprayed on a large area of the body to disinfect skin and mucosal wounds.

(II) Exposure therapy: disposable mattresses were placed under the body and a brace was used on the bed. As the systemic skin lesions were serious and clothing was not conducive for healing, bedding was placed on the brace to keep the body warm while exposing the skin lesions completely to facilitate recovery.

(III) Timely removal of necrotic scabs: autolytic combined with conservative sharp debridement (6) was used to reduce scab and accelerate epidermal cell migration. Prandtl liquid wound dressing (7) with sterile pads was used for wet compress for 10–15 minutes, and then sterile tweezers were used to gently remove scab skin along edges once a day. Silver ions after debridement dressing and vaseline gauze were used to cover the wound to maintain an appropriate wound temperature and humidity to dissolve the effusion protein enzyme and necrotic tissue.

(IV) Joint skin protection: in areas prone to bleeding and cracking, such as the elbow, popliteal fossa, and ankle joints, Yunnan Baiyao powder was sprinkled on the bleeding and cracked sites to stop the bleeding. Vaseline gauze was attached to keep the skin moist and to prevent secondary damage caused by cracking, with replacement every 3 hours.

Delirium care

Intensive Care Unit (ICU)-acquired delirium is a severe complication of critical illness characterized by acute brain dysfunction, including inattention, disturbance of consciousness, and changes in cognitive function (8). To detect and prevent delirium in a timely manner, the Richmond Agitation-Sedation Scale (RASS) and Cornell Assessment of Pediatric Delirium (CAPD) were administered to the patient every 12 hours (9-11). Several studies have indicated that delirium can be caused by various medications, although the exact reasons remain unclear (9-11). Delirium can lead to adverse effects such as increased mortality, prolonged hospital stay, and negative impacts on growth and development (12,13). Evidence suggests a higher mortality rate in older FUMHD patients, highlighting the need for age-adapted treatment strategies. Effective monitoring, identification of the causes, and timely prevention and control of delirium by ICU nurses have been shown to improve patient prognosis (14).

On the second day of admission to the PICU, the patient scored 10 on the CAPD scale and 1 on the RASS scale, indicating high activity of delirium. Following the concept of chronic wound care proposed by Shi *et al.* (15), the patient received comfort therapy based on the early Comfort using Analgesia, minimal Sedatives and maximal Humane care (eCASH) approach, which involved early and comfortable application of analgesia, minimal sedation, and maximum humanistic care. Our approach to managing FUMHD draws notable parallels to the optimal care strategies for burn injuries, as outlined by Bittner *et al.* (16). Similar to the multidisciplinary and intensive care required for severe burn victims, FUMHD treatment necessitates comprehensive management strategies focusing on wound care, infection prevention, and systemic support. This comparison underscores the importance of applying principles from burn care, such as meticulous wound management and vigilant monitoring for complications, to enhance treatment outcomes for FUMHD patients.

Pain management

Daily pain assessment: the location, nature, and intensity of pain were actively assessed, and timely interventions were implemented. Pain assessment was conducted twice daily, as well as before and after dressing change.

Appropriate use of analgesics: in cases of severe pain during routine skin care, fentanyl was used for analgesia following the guidelines for herpes zoster pain. Oral analgesic drugs were administered to assist with the pain management. The dose of fentanyl was increased during dressing changes to alleviate severe pain. Lidocaine injection, which provides local anesthesia and pain relief, was added to chlorhexidine acetate solution. Approximately 0.1–0.2 g of lidocaine injection per 50 mL solution was mixed and sprayed over a large area of the body (*Figure 4*).

Minimal sedation

To maintain moderate sedation and ensure smooth

Figure 4 Erythema, blisters, papules, necrosis, crusting, and scales visible on the surface of some skin lesions.

treatment and procedures, intravenous dexmedetomidine injection was administered for maintenance over a period of 15 days. Light sedation was maintained with a RASS score of 0–1. The patient's vital signs were closely monitored, and the dosage of sedative drugs was adjusted along the course of disease progression until withdrawal.

Maximize humanistic care

Early exercise was encouraged; however, due to systemic skin lesions, exercise, especially those involving the joint, was affected by skin crusts, and early treatment of skin lesions remain crucial. The patient and his family lacked knowledge about FUMHD, and the prolonged treatment process placed a significant psychological burden on them. Psychological counseling and effective communication were necessary. When the patient, a junior high school boy, expressed concerns about scarring and pain experienced during skin care and activities, responsible nurses played a role in reducing the psychological burden. This was achieved through daily inquiries about the patient's concerns, providing disease-related knowledge to the patient and his family, instilling accurate information regarding the disease, and presenting successful treatment cases to promote patient recovery.

Catheter care

Proper fixation of the central venous catheter is critical for the smooth progress of intravenous fluid infusion and continuous renal replacement therapy (CRRT) treatment in children with extensive skin damage. Each shift evaluated the puncture site, catheter fixation, and pressure changes during CRRT treatment. The central vein was fixed with suture and gauze dressing (17), with the puncture site disinfected and dressings changed every day to prevent catheter-related infection and catheter-related bloodstream infection. We used chlorhexidine acetate cotton balls for disinfection and sterilized gauze for fixation.

Protective isolation

The patient had a *Pseudomonas aeruginosa* and *Acinetobacter baumannii* infection with fungal infection, requiring strict adherence to aseptic operation principles and protective isolation measures. The patient was placed in a single ward, and nurses provided concentrated treatment to reduce ward visits. The wards were sterilized with an air sterilizer twice daily for one hour each time, with weekly air culture. All objects in the ward were wiped with disinfectant wipes three times daily, and clothes, sheets, and covers were sterilized and reused.

Diet nursing

Although the patient had ulceration and scabbing around the mouth, there was no oral mucosa erosion. The patient was encouraged to consume high-calorie liquid or semi-liquid foods, avoid spicy foods and seafood, eat small meals, drink more water, and maintain accurate intake and output records. Oral care was performed three times daily, using compound chlorhexidine mouthwash. The patient's calorie requirements were estimated using physiological requirements and calories calculation, and parenteral nutrition was used as per the doctor's advice.

Conclusions

In 2022, Reich *et al.* (18) reported a rare case of acute fever and ulceration in smallpox pityriasis moss samples, referred to as febrile pityriasis ulcers acute necrotizing smallpox samples. This condition has been reported in less than a hundred cases worldwide, with a current mortality rate of 13% (19), which increases with age. In managing this disease, it is crucial to focus on preventing and caring for skin lesions and complications, as well as safeguarding the patient's well-being. These measures can help shorten

the treatment duration and alleviate the patient's pain. Throughout the treatment in the PICU, the patient's memory remained intact, enabling them to independently manage the diet and activities. The patient actively cooperated with doctors and nurses, displaying no signs of mental or neurological dysfunction. Our case of FUMHD provides valuable insights into the disease's clinical variability and response to treatment, thereby enriching the limited literature. By detailing the diagnostic challenges, treatment strategies, and patient outcomes, we contributed to a deeper understanding of FUMHD, offering a reference for future cases and highlighting the necessity for personalized, multimodal care approaches. This case underscores the importance of comprehensive reporting in enhancing the collective knowledge and management strategies for this rare and complex condition.

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Footnote

Reporting Checklist: The authors have completed the CARE reporting checklist. Available at https://tp.amegroups.com/article/view/10.21037/tp-23-520/rc

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Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://tp.amegroups.com/article/view/10.21037/tp-23-520/coif). L.X. receives supports from the Shanghai Jiao Tong University School of Medicine: Nursing Development Program (grant number: XH3295) and 2023 Shanghai Jiao Tong University School of Medicine Nursing Research Top Project (grant number: Jyh2303). The other authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient's legal guardian for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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