VASCULAR DISEASE, VASCULITIS

FEBRILE ULCERONECROTIC MUCHA-HABERMANN DISEASE - A CASE REPORT AND REVIEW OF THE LITERATURE

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Background: A 47-year-old male patient who is characterized by the sudden onset of ulceronecrotic skin lesions associated with high fever and systemic symptoms such as intermittent high temperature, hepatic dysfunction, anemia and hypoalbuminemia and alimentary tract hemorrhage.

Observation: This patient was diagnosed as severe febrile ulceronecrotic Mucha-Habermann disease(FUMHD). He was treated with topic antiseptics and oil quality protective agents after surgical debridement twice a week, as well as immunosuppressive therapy using high-dose methylprednisolone (1 mg/kg/day) and supportive treatments of plasma infusion, whole blood transfusion and albumin transfusion. These therapies as well as intensive supportive care proved to be effective in abating fever and inducing a dramatic improvement of ulceration and in arresting the appearance of new lesions. However, methotrexate (10 mg/wk) successfully cleared his systemic symptoms. The FUMHD is a rare and potentially lethal type of pityriasis lichenoides et varioliformis acuta (PLEVA, about 50 cases described to date), characterized by the sudden onset of ulceronecrotic skin lesions associated with high fever and systemic symptoms such as intermittent high temperature, hepatic dysfunction, anemia and hypoalbuminemia. Skin biopsy showed a combination of the classic features of PLEVA and an allergic vasculitis. Patients with FUMHD should be treated with combined therapy of both glucocorticoids and methotrexate. This patient highlights the exceptional response of FUMHD to methotrexate. Surgical debridement and intensive supportive care are recommended. And to those who couldn't afford expensive treatment with intravenous immunoglobulin, plasma infusion and whole blood transfusion and albumin transfusion form alternatives.

Key message: Febrile Ulceronecrotic Mucha-Habermann Disease, Pityriasis Lichenoides et Varioliformis Acuta