Case report of mucha-habermann disease

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ABSTRACT

Pityriasis lichenoides et varioliformis acuta (PLEVA), also known as a Mucha-Habermann disease, is an uncommon, idiopathic, and acquired dermatosis. The disease is characterized by erythematous, scaly, papules, and polymorphic lesions which often progresses to hemorrhagic necrosis and heals with varioliform scarring. A febrile ulceronecrotic variant of PLEVA, also termed pityriasis lichenoides (PL) with ulceronecrosis and hyperthermia (PLUH) or febrile ulceronecrotic Mucha-Habermann disease (FUMHD), is a severe variant of PLEVA. The disease is characterized by the acute onset of large, more destructive, coalescent papules, leading to ulceronecrotic skin lesions associated with high fever and other systemic symptoms. In spite of the presence of multiple treatment modalities with variable success rate, the disease has poor prognosis. Here, we report the case of a 17-year-old male patient who presented with typical features of FUMHD and responded well to systemic administration of corticosteroid therapy.

Key words: *Pityriasis lichenoides, Systemic corticosteroid, Ulceronecrosis*

Pityriasis lichenoides (PL) is an uncommon, acquired spectrum of cutaneous dermatoses that pose various challenges to patients as well as treating clinicians. It is a difficult disorder often poses a challenge to diagnose, categorize, and treat. In spite of these inherent obstacles, PL merits awareness because it has been considered as a variant of parapsoriasis and also possesses potential to progress to cutaneous lymphoma [1] or an ulceronecrotic presentation, both of which are associated with a significant risk of mortality. The spectrum of PL presentations is delineated along a continuity of multiple variants [2], which includes pityriasis lichenoides chronica (PLC), pityriasis lichenoides et varioliformis acute (PLEVA), and febrile ulceronecrotic Mucha-Habermann disease (FUMHD), also known as pityriasis lichenoides with ulceronecrosis and hyperthermia (PLUH). The disease is very rare, worldwide, its incidence and prevalence are not defined. As per the literature, around 70 cases of FUMHD were reported (till 2016) and few anecdotal case reports are also present from India [3]. We report a case of 17-year-old male patient presenting with this rare disease with ulceronecrosis and responded beautifully to a course of oral steroid over a period of 6 weeks.

CASE REPORT

A 17-year-old male student presented to our department with chief complains of fever and multiple painful ulcerative lesions over the body. The illness started as high-grade fever without chills, malaise, and body ache for 5–7 days and development of skin lesions as reddish raised papules on the 3rd day of fever. The lesions first started on trunk, then gradually involved limbs and head and neck area. Over a period of next 2 days, these lesions increased in number and size, with central necrosis in few of them which further progressed to form ulcer and covered with thick adherent crust.

There was no history of intake of any drug, vaccination, upper respiratory tract infection, systemic disease before the onset of illness, or any sexual contact. There were no associated symptoms of any other system involvement such as joint pain, dysuria, respiratory, abdominal, or neuropsychiatric problems, and there was no history of similar illness in the past inpatient or any other family member.

On general physical examination, the patient was average built and well nourished. Fever was (101°F) with tachycardia (pulse 90 beats/min) at the time of examination. Systemic examination of other systems and rest of the vitals were within normal limits. The peripheral nerves were palpable and not enlarged. On detailed cutaneous examination, around 40–50% of body surface area was involved with almost bilateral and symmetrical distribution of polymorphic generalized cutaneous lesions. The lesions were seen as discrete to coalescent, erythematous, edematous papules, plaques, and punched out ulceration with adherent hemorrhagic crusting (Figs. 1 and 2). These ulcers were multiple in number, round to polygonal in shapes, with size varying from 1 to 10 cm. They had well-defined punched out margins, with necrotic tissue crusting on the floor, and base was free from the underlying tissue. The lesions were mild-to-moderate tender and indurated. The scalp, hair, and nail were normal.

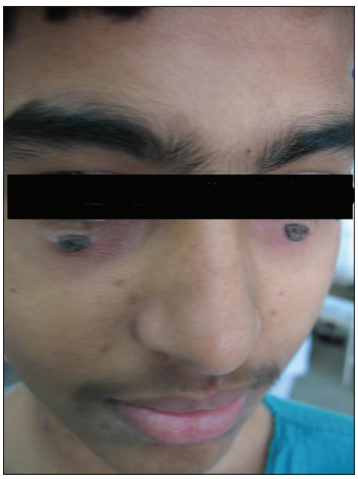

Figure 1: Nodulo crusted lesions over face

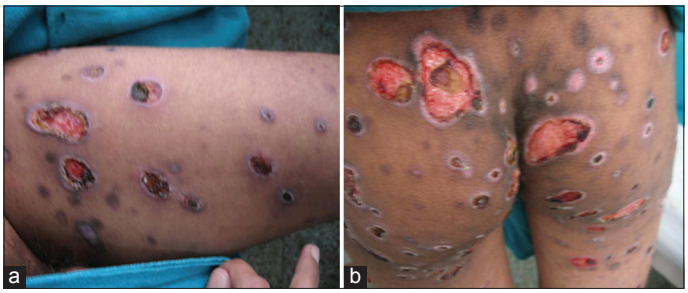

Figure 2: Multiple large ulcerative lesions with crust and post-inflammatory changes over thighs (a) and buttocks (b)

Based on the clinical presentation of the patient, the differential diagnosis of necrotizing skin infections - erythema nodosum leprosum necroticans, malignant syphilis, and ecthyma gangrenosum and cutaneous vasculitis - Wegener's granulomatosis, pyoderma gangrenosum, and ulceronecrotic variety of PLEVA were kept.

On laboratory investigation, the complete blood count showed leukocytosis (total leukocyte count of 14,050 cells/L) and erythrocyte sedimentation rate of 32 mm. Other parameters such as C-reactive protein, blood sugar, urine examination (routine and microscopic), liver and kidney function tests, electrocardiogram, X-ray chest, and ultrasonography of abdomen were found to be normal. Venereal disease research laboratory test, Treponema pallidum hemagglutination test, viral markers (hepatitis B and C) human immunodeficiency virus test, antinuclear antibody test, and antinuclear cytoplasmic antibody test (ANCA-against both proteinase 3 and myeloperoxidase) were non-reactive/negative.

Gram stain from the skin lesions showed predominate neutrophils, but culture from ulcer base, blood, urine, and throat swab failed to grow any organism. The slit smear examination was negative for acid-fast bacilli. Skin biopsy from the ulcer showed mild edema and spongiosis of the epidermis (Fig. 3a and b). Dermal changes included the presence of moderate edema, moderately dense inflammatory infiltrate predominantly composed of lymphocytes, few eosinophils, and extravasated red blood cells. Direct immunofluorescence study was not performed because of unavailability.

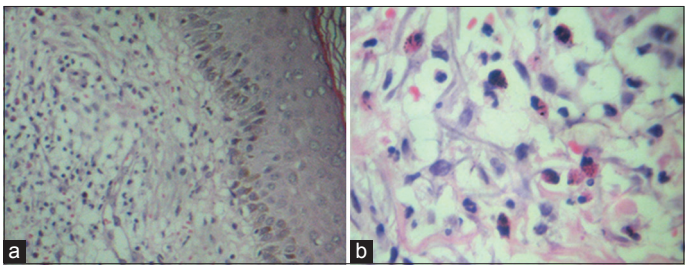

Figure 3: Histopathology (a and b) showing epidermis with mild edema, spongiosis. Dermal edema with dense inflammatory infiltrate predominantly composed of lymphocytes, few eosinophils, and extravasated red blood cells

The final diagnosis of an ulceronerotic variant of PLEVA was made after correlating the clinicopathological scenario. The patient was treated promptly with oral antibiotics, systemic corticosteroids 1.5 mg/kg body weight, and supportive treatment. The patient responded well to oral corticosteroids over a period of 2 weeks, which allowed us further to taper the dose of steroid and eventual withdrawal of the same in 6 weeks duration. The papules and plaques healed with post-inflammatory hyperpigmentation, whereas ulceronecrotic lesions healed with varioliform scarring. The patient did not suffer from any recurrent episodes of similar illness in 1-year follow-up period.

DISCUSSION

PLUH represents a fulminant and potentially lethal variant of PLEVA. In 1916, Mucha, and in 1925, Habermann [4] reported an acute form of PL characterized by the abrupt onset of papulovesicular eruptions and described it as PLEVA or Mucha-Habermann disease. In 1966, Degos [5] first described a rare variant of PLEVA, also known as FUMHD.

PLEVA and PLUH occur most commonly in children, adolescents, and young adults. Majority cases of FUMHD occur in the second or third decade of life with a male predominance [6]. The cutaneous lesions of PLEVA usually develop in crops, may be asymptomatic or mild pruritic in nature. They are polymorphic and may heal with varioliform scarring. These patients can also have accompanying constitutionals symptoms without major systemic signs. FUMHD, on the other hand, often starts as classic PLEVA but goes on to develop widespread ulceronecrotic lesions which may develop in new crops over many months, it is associated with a fulminating course and high fatality rate, particularly in adults and immunocompromised patients. Although the exact etiopathogenesis for PLUH is unknown, several reports have suggested the possibility of hypersensitivity to an infectious agent [7]. The elevation of microbe-specific antibody titers, deposition of immune complexes in dermal vessels, familial outbreaks [8], and associated constitutional symptoms have been offered as potential evidence for infectious etiology of the disease.

Another postulated mechanism for PLEVAs pathogenesis is lymphocytic proliferation. Lopez-Estebaranz *et al.* [9], however, implied that FUMHD represents an inflammatory disorder rather than a T-cell lymphoproliferative process as they did not detect abnormalities in T-cell receptor gene analysis of DNA

from skin lesion and peripheral blood [9]. Immune complex-mediated vasculitis [4] has also been implicated as one of the etiopathogenesis mechanisms.

The management of PLUH is controversial and a considerable number of therapies have been tried with varied success rate. These include systemic corticosteroids [10], antibiotics, acyclovir, dapsone, methotrexate [10,11], cyclosporine [12], psoralen plus ultraviolet A, and tumor necrosis factor-alpha inhibitors (such as infliximab and etanercept) [13]. A combination of these agents has been tried in some patients. Prognosis of the condition is variable at times fatal, which is more common in adults as compared to the children [3].

In our patient, systemic corticosteroids in dosages >1 mg/kg per day, along with oral antibiotics, were used. The patient responded to this very well without any and on immunosuppressive or biologics.

CONCLUSION

PLUH or FUMHD is a severe disease, being a rare presentation of PLC, associated with a significant risk of mortality. Timely diagnosis, prompt management, and appropriate treatment according to an individual patient are important. In this patient, systemic corticosteroids in dosages >1 mg/kg per day, along with oral antibiotics, were used and proved lifesaving.

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