

Pityriasis Lichenoides Chronica (PLC) Turning into Febrile Ulceronecrotic Mucha-Habermann Disease: A Case Report.

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Abstract: Pityriasis Lichenoides Chronica (PLC), Pityriasis Lichenoides Et Varioliformis Acuta (PLEVA), Febrile Ulceronecrotic Mucha-Habermann Disease (FUMHD) all represent clinicopathological spectrum of same disease entity. FUMHD is a rare febrile variant of PLEVA characterised by necrotic cutaneous ulcerations associated with high fever and systemic manifestations. Hereby we report a 11 year old male, who initially presented with lesions of PLC which healed with oral erythromycin for 8 weeks but in 3 months he again presented with generalised ulceronecrotic lesions on skin, continuous high grade fever and abdominal pain suggestive of Mucha-Habermann Syndrome. He responded only to systemic corticosteroids. Disease being a clinical spectrum, it is important to follow up the patients presenting with PLC and having a watchful suspicion as it can turn into potentially fatal MHD.

Keywords: Pityriasis Lichenoides Chronica, Pityriasis Lichenoides Et Varioliformis Acuta, Mucha-Habermann Syndrome, T-cell monoclonality.

I. Introduction

Mucha-Habermann disease (MHD) was described by Degos et al. in 1966¹. It is considered a severe variant of pityriasis lichenoides et varioliformis acuta (PLEVA) characterised by purpuric, papulonodular, polymorphic, ulceronecrotic and crusted lesions on skin and mucous membranes accompanied by high fever and other systemic manifestations like pneumonitis, abdominal pain, malabsorption, liver, CNS involvement and rheumatological manifestations². Pityriasis Lichenoides Chronica (PLC) is a milder chronic form of disease characterised by brownish-red scaly papules and plaques without ulceration and crusting. There are no systemic symptoms. However patients may exhibit a mixture of acute and chronic lesions sequentially or concurrently³. The etiology of this disease is unknown. Infectious agents like Toxoplasma gondii, Epstein-Barr virus, Cytomegalovirus (CMV), Parvovirus B19 have been implicated. Dominant T-cell clonality has been demonstrated⁴ and some consider it as clonal cytolytic memory T-cell proliferative response to foreign antigen. We report a case of a 11 year old male who presented with PLC and responded to oral erythromycin and topical potent steroids. But he again presented with features of MHD in 3 months finally responding to systemic steroids.

II. Case Report

A 11 year old male presented to us with 2 year history of itchy brownish red flat papules and plaques over extremities, trunk and few on face. His general condition was good without any systemic symptoms. Auspitz sign was negative but there were few feathery scales over the lesions. There were dystrophic changes in his finger nails. (fig.1). Histopathological examination of the lesion showed superficial perivascular lymphocytic infiltrate with extravasation of RBCs in papillary dermis and focal vacuolar change. Epidermis was infiltrated by lymphocytes and RBCs in foci. (fig.2). A diagnosis of PLC was made and he was given oral Erythromycin 500mg in divided doses for 4 weeks and topical Mometasone with Salicylic acid ointment. The lesions improved rather slowly in 3 weeks with post inflammatory hyperpigmentation (PIH). We discharged him with emollients. After about 2 months the same patient came with severe high grade fever and extensive necrotic papules and plaques with crusts all over the body which he claimed to appear in 3-4 days. He looked toxic and drowsy. (Fig. 4). CSF analysis showed no abnormalities. Complete blood count showed moderate lymphocytosis. His liver enzymes were elevated (SGOT 239 IU/L) with marginal elevation of serum urea (75 mg/dl) and creatinine (1.9 mg/dl). He was put on IV fluids, i.v Ceftriaxone and i.m. paracetamol for 5 days. But his condition deteriorated for 3 days. A provisional diagnosis of FUMHD was made and i.v Dexamethasone 8 mg/day was administered for 3 days followed by oral prednisolone 20 mg/day which was tapered by 5mg every 10 days. He improved dramatically within few days and lesions healed convincingly. We report this case as it was challenging and peculiarly converted from previous PLC to FUMHD.



Figure 1 brownish papules and plaques with dystrophic nails

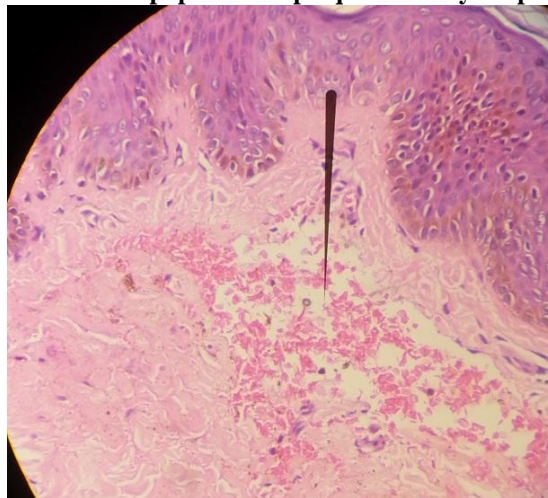


Figure 2 H&E staining 40X showing extravasatedRBCs in papillary dermis

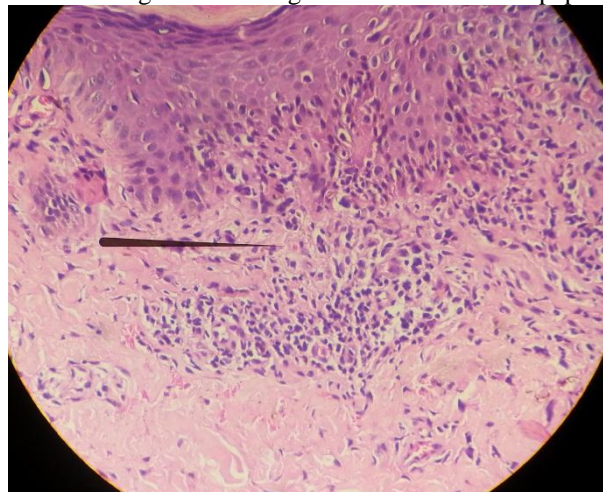


Figure 3H&E stain 40X showing focal interface vacuolar change and lymphocytic infiltration and extravasation in epidermis



Figure 4 the boy with toxic look with crusted plaques around lips, eyes and over chest and forearms



Figure 5,6 papules and plaques with central necrosis and crusting

III. Discussion

Mucha and Habermann described in 1919 and 1925, respectively a severe form of pityriasislichenoides characterised by sudden onset of cutaneous eruption with necrosis, crusting and severe constitutional symptoms. In 1966, Degos et al. described MHD with similar findings. Its etiology remains unknown and believed to be caused by infectious agents like EBV, Parvovirus B19, staphylococcus, Streptococcus and pseudomonas⁵. It is common in children and adolescents.

The cutaneous manifestations are widespread polymorphic, ulceronecrotic, crusted papules and plaques frequently secondarily infected and tend to resolve with hypochromic scar. The oral, genital, conjunctival mucosae as in this patient may be involved⁶. There may be liver, gastrointestinal disturbances similar to our patient. It can also lead to CNS manifestations, lymphadenopathy, cardiomyopathy, DIC, Pneumonitis⁷.

The diagnosis is based on presence of high fever, typical cutaneous lesions and biopsy showing changes of PLEVA with leucocytic extravasation in epidermis. There may be elevated ESR, CRP, and hypergammaglobulinemia⁷.

The prognosis is worst in adults but good in children⁶ as in our patient. There is no definitive treatment modality⁶. Most are treated with multiple therapeutic options like systemic glucocorticoids as in our case, antibiotics, acyclovir, methotrexate, IVIG, cyclosporine^{7,8}. Most recent studies have reported success with methotrexate with methyl prednisolone pulse therapy^{7,8}. Anti TNF- α agents can be tried as these patients have high TNF- α titers⁷. Our case who initially diagnosed as PLC later developed high fever, disseminated ulceronecrotic cutaneous lesions, typical histopathological findings pointed us to the diagnosis of MHD.

IV. Conclusion:

PLC, PLEVA and MHD represent a spectrum of diseases and can present in continuum. So any patient with PLC should be regularly and suspiciously monitored for PLEVA and MHD and managed accordingly.

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