Pityriasis Lichenoides et Varioliformis Acuta (PLEVA; Mucha-Habermann disease): A Case Report

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History

G.C. an 8 year old boy from Senglea, presented in July 1983 with crops of erythematous papules rapidly evolving into vesicles and ulcers with haemorrhagic crusts. Lesions healed spontaneously with post-inflammatory hyperpigmentation, but heavily infected ones left scars. The rash started on the face but soon became generalized.

Examination

The child looked remarkably well in spite of the rash and a temperature of 100°F. There were several cutaneous lesions at various stages of evolution (Fig. 1). The general examination was otherwise negative.

Investigations

A freshly blistered lesion was sampled for routine histology. This showed a mononuclear infiltrate surrounding blood vessels and invading a degenerating epidermis (Slide No.: 3546/83).

The E.S.R. was 35 mm in the 1st hour (Wintrobe method). The Hb, WBC count and differential, blood film, urinalysis and agglutinin titration were normal. A throat swab yielded no pathogens and his ASOT was

less than 200 IU/ml.

Treatment

He was treated initially with systemic antibiotics and topical astringents. Subsequently he received oral prednisolone for a period of six months (2.5 mg in the morning on alternate days) and topical adrenocorticosteroids for the more troublesome lesions. The rash subsided and did not relapse on withdrawal of the steroids.

Comment

This child had Pityriasis Lichenoides. In the milder form of this disease, called Pityriasis Lichenoides chronica, individual lesions evolve slowly and remain papulosquamous, inviting clinical confusion with psoriasis or lichen planus. In the more severe form, called Pityriasis Lichenoides et Varioliformis Acuta, the lesions evolve much more rapidly and epidermal involvement produces papulonecrotic lesions which may be misdiagnosed initially as chickenpox.

Histologically, the disease is a lymphoid vasculitis. This may be the result of a hypersensitivity reaction to an infective organism but so far all attempts to isolate a culprit have failed.

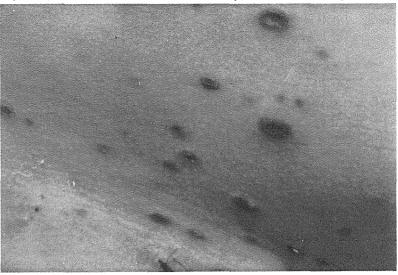


Fig. 1. Papulonecrotic lesions in the right iliac fossa

d.a.