Ind J Dermatol Venereol Leprol 1996; 62 : 379-80

ICHTHYOSIS LINEARIS CIRCUMFLEXA

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A case of ichthyosis linearis circumflexa in a 9-year-old boy is reported here for its rarity

Key Words : Ichthyosis linearis circumflexa, Comel - Netherton's syndrome

Introduction

This rare and distinctive entity was recognised by clinicians for many years but confusion existed due to a lack of proper descriptive terminology. It was Comel in 1949, who first coined the term ichthyosis linearis circumflexa giving it a clearcuit individuality.[1] Netherton in 1958 reported an association of bamboo hair (trichorrhexis invaginata) and ichthyosiform erythroderma.[2] Atopic diathesis was also reported in some patients. Since the initial description of this disorder by Comel in 1949, less than 70 cases were reported by 1987.[3]

Case Report

A 9-year-old boy, the second issue of a non-consanguineous couple reported to us with generalised scaly lesions since his infancy. On examination the child was found to be of average built with wide spread erythematous, serpiginous, annular or polycyclic, scaly eruptions with double edged scales at the periphery of the lesions (Fig.1). The lesions are continuously changing their patterns and shape with a bizarre migratory nature, involuting in a week or two after reaching their maximum size leaving no scarring, atrophy or pigmentary changes. The lesions are distributed mainly over the trunk (Fig. 1) with hyperkeratotic flexures in the extremities. Hair, nail, genitalia and mucous membrane was not involved. The child was not atopic. Routine physical examination, X-ray chest, Mantoux test, complete haematological check up did not reveal any abnormality. Histopathological examination from the periphery of the lesion showed hyperkeratosis, papillomatosis with upward projection into the epidermis and suprapapillary thinning (Fig.2). Collection of homogenous material in the horny layer was also present (Fig.3). These features coroborated well with our clinical diagnosis.[4]

Fig. 1. Typical lesions of ILC distributed mainly over the trunk.

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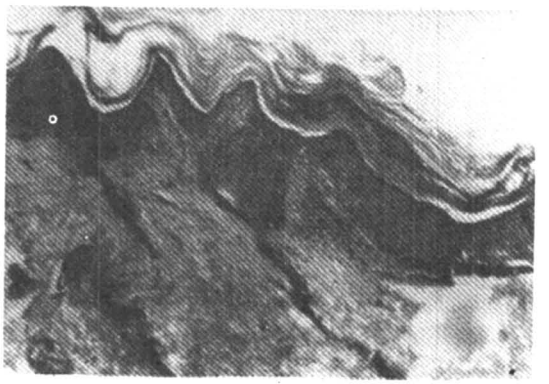

Fig. 2. Hyperkeratosis, papillomatosis and suprapapillary thinnig (H&Ex65).

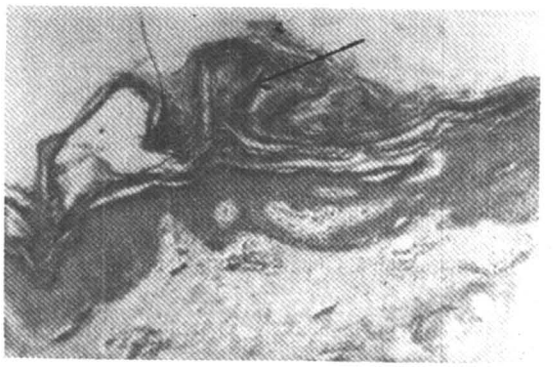

Fig. 3. Collection of homogenous material in the horny layer (marked here with an arrow) (H&E x 65).

Discussion

ILC is a very rare autosomal dominant disorder. Generalised erythema and scaling are seen at birth or shortly thereafter.Subsequently polymorphic, serpiginous, erythematous, slowly changing lesions with double-edged scaly border appear on the trunk and proximal extremities. Lichenification of the popliteal and cubital fossae and red, scaly face and eyelids are also seen. Till date only 4 cases are reported in Indian journals. In 1985 Chandwala et al reported a brother and sister from Pune, both in their teens, with this rare disease.[5] The other two were from Libya. Kanwar et al in 1987 reported a 6-year-old boy from Benghazi and in 1989 Khatri et al reported another male patient of same age from Tripoli.[6,7]

Clinically and histologically ours was a classical case of ILC but associations like hair abnormalities, atopy, palmoplantar hyperkeratosis and hyperhydrosis, pruritus, stunted growth and mental retardation where absent in our case.

To the best of our knowledge, except for the cases reported by Chandwala, ours is the only other Indian case of ILC and the very first one reported from eastern India.

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