ICHTHYOSIS LINEARIS CIRCUMFLEXA

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Two cases of ichthyosis linearis circumflexa occurring in the same family having classical migratory polycyclic skin lesions, and flexural hyperkeratosis since infancy but no hair abnormality or palmo-plantar hyperhidrosis are reported.

Key words : Ichthyosis linearis circumflexa.

Primary ichythyosiform dermatoses consist of a heterogenous group of hereditary disorders Some of these have associated systemic, especially neurological manifestations while most have only skin involvement.

Ichthyosis vulgaris and X-linked ichthyosis occur in about 1:300 persons and 1:6000 males respectively.[1] The incidence of lamellar ichthyosis and epidermolytic hyperkeratosis is 1:3,00,000.[1] Erythrokeratoderma variabilis and ichthyosis linearis circumflexa (ILC) are very rare. We are reporting two cases of ILC from the same family.

Case Reports

A 19-year-old girl and a 14-year-old boy, brother and sister, had pruritic skin lesions with history of secondary infection off and on, since the age of four weeks. Both of them were delivered normally at full term. There was no history of consanguinity. The lesions started over the face and the scalp, later affected the trunk and proximal parts of the extremities. The lesions were erythematous, serpiginous and polycyclic. Some had elevated borders and double-edged scales. Hyperkeratotic, brownish, lichenified lesions were present in both axillae and cubital fossae. Some lesions had pustules.

Only the brother had the scalp lesions. Hair, teeth mucous membranes, palms and soles were normal. The lesions changed their shape and sites by themselves. There were no seasonal variations. No one else in the family had similar illness. The sister had history of allergic bronchitis and solar sensitivity.

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General and systemic examination did not reveal any positive findings.

Histopathology revealed hyperkeratosis with parakeratosis, acanthosis and papillomatosis. Granular layer was normal. Basal cell layer was intact, but had reduced pigmentation. Upper dermis had a sparse mononuclear infiltrate.

Comments

Ichthyosis linearis circumflexa or dyskeratosis ichthyosis congenita migrans[2] is characterised by the triad of migratory, serpiginous and polycyclic lesions, hyperkeratosis of flexures and hyperhidrosis of the palms and soles.[2] It was first described in 1949 by Comel.[2] In 1961, Vineyard et al[2] reported the disease in a brother and a sister. Both sexes are equally affected. So far, all the reported cases have been caucasians.[2] The onset is at birth or during the first year. Inheritance is probably autosomal recessive. Our cases show all these features except hyperhidrosis of the palms and soles. Presence of hyperhidrosis of palms and soles is not seen in all patients.[2] Some reported cases have hair abnormalities similar to Netherton syndrome.[2] No hair changes were observed in our patients. Association of trichorrhexis nodosa with ILC suggests ILC may be a variant of lamellar ichthyosis.

References

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