ICHTHYOSIS LINEARIS CIRCUMFLEXA

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A case of ichthyosis linearis circumflexa presented with multiple serpiginous polycyclic lesions and characteristic double edged scales. The patient had unusual features like pruritus and thickening of the palms and soles. One younger sib of the patient appeared to have lamellar ichthyosis. The response to therapy was not satisfactory.

Key words : Ichthyosis linearis circumflexa.

The four major types of ichthyoses are ichthyosis vulgaris, X-linked ichthyosis, lamellar ichthyosis and epidermolytic hyperkeratosis. Some other types of ichthyoses are also of clinical interest as these may be associated with other systemic manifestations.[1] Ichthyosis linearis circumflexa (ILC) is an extremely rare type of ichthyosis.[1] Since the initial description of this disorder by Comel in 1949,[2] less than 70 cases have been reported.[3] In the present communication, a case of ILC, the first from Arab world is reported.

Case Report

A 6-year-old male child had severely itchy erythemato-squamous lesions on the trunk and extremities since birth. These lesions were recurrent, varied in location and were still evolving and frequently got secondarily infected. No seasonal variation was observed. The child's mother had a full-term pregnancy and uncomplicated delivery. There was no family history of consanguinity. The lesions were multiple, serpiginous, polycyclic with double-edged scales and pustules, present on the trunk and proximal extremities. The face, scalp and eyebrows showed scaling in a sebaceous pattern. There was thickening of the palms and soles and mild hyperkeratosis in the flexures. During stay in the hospital, the lesions were observed to extend peripherally quite rapidly. The child's general growth and health as well as his teeth, hair, eyes and skeletal system were unaffected. The nails were normal.

Complete blood cell count, erythrocyte sedimentation rate, liver and renal function tests, urinalysis, serum protein electrophoresis, serum vitamin A levels, blood sugar, chest X-ray and serum immunoglobulins were normal. Urinary aminoacid levels were also normal. Pus culture revealed profuse growth of *Staphylococcus aureus*. Histopathology showed hyperkeratosis with parakeratosis, acanthosis and papillomatosis. Granular cell and basal cell layers were normal. Dermis was oedematous with a moderate perivascular round cell infiltrate.

One of the younger sibs of the patient, aged 6 months also had generalised erythema with large scales all over including the cubital and popliteal fossae, intertriginous regions and the palms and soles. There was matting of hair on the scalp with secondary bacterial infection.

Comments

Ichthyosis linearis circumflexa (ILC) is a rare autosomal recessive disorder.[1] Generalised erythema and scaling are seen at birth or shortly thereafter and then fade. Subsequently, polymorphic and serpiginous erythematous lesions bordered by a double-edged scale appear on the trunk and proximal extremities. There is lichenification of the popliteal and cubital fossae and the dorsum of the wrist. The face and the eyelids are diffusely red and scaly and the pattern of the lesions slowly changes.[1] Our patient

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had all the typical features characteristic of ILC. Apart from these, there were some other uncommon features such as severe pruritus. Pruritus has been referred to only three times in the literature in patients of ILC.[4] The thickening of palms and soles and frequent cutaneous infections however, have not been described earlier.

Some reported cases of ILC have shown abnormalities of hair, trichorrhexis invaginata and signs of atopy. This collection of symptoms is referred to as Netherton's syndrome.[4,5] In our patient, no hair abnormalities were observed and there was no atopic diathesis.

Another interesting feature was involvement of the younger sib of the patient who had clinical features of lamellar ichthyosis. Though in the initial stages, it may be difficult to exclude it,[6] the changing circinate lesions are highly characteristic of ILC. However, a 6-month follow up has not shown any such lesions, though the erythema and scaling have become less.

The course of ILC is characterized by partial remissions.[1] Topical corticosteroids and systemic antihistaminics have been temporarily effective. Photochemotherapy[3] and heavy doses of vitamin A[7] which have been reported to be beneficial in patients of ILC were of no effect.

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