NETHERTON'S SYNDROME

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A 6-year-old Libyan boy had diffuse erythema at birth, and later developed pruritic, maculo-papular, papular, circinate, double-edged, scaly lesions, suggestive of ichthyosis linearis circumflexa (ILC). His scalp hair were brittle and sparse with partial patchy alopecia, showing changes of trichorrhexis invaginata, these associations being characteristic of Netherton's syndrome. The boy had slighly stunted growth, a feature which has not been recorded in previously reported cases.

Key words : Netherton's syndrome, Ichthyosis linearis circumflexa (ILC), Trichorrhexis invaginata.

Netherton's syndrome is characterized by ichthyosis linearis circumflexa (ILC) or non-bullous ichthyosis associated with abnormality of hair growth, mainly showing trichorrhexis invaginata. Sometimes atopic manifestations, aminoaciduria and mental retardation may also be present. The inheritence pattern is autosomal recessive.[1] Netherton[2] described his first case in 1958, with congenital non-bullous ichthyosis, associated with sparse, brittle hair showing nodose swellings (bamboo hair). Further cases were reported by Marshall and Brede in 1961, and by Wilkinson et al in 1964, who described the changes of hair as trichorrhexis invaginata.[3] In 1969, Altman and Stroud[4] described specific skin changes of ILC with trichorrhexis invaginata. A 6-year-old Libyan boy with this condition was seen by us recently.

Case Report

A 6-year-old Libyan boy had generalized erythema and scaly lesions on the hands and feet since birth, with episodes of regression and exacerbation. His hair were normal at birth but at the age of two years, these started becoming brittle and he developed irregular patches of partial alopecia on the scalp. He also had moderate to intense itching. There were no seasonal variations. One elder brother (18 years age) also had the similar condition. The parents were normal but related (first paternal cousins). At the time of admission, the erythema was diffuse, mainly on the face, trunk, thighs, hands and feet, along with maculo-papular, papular, circinate, double edged, scaly lesions on the trunk, thighs, hands and feet. Mild to moderate lichenification was also noticed on both antecubital and popliteal fossae. Scalp hair were short and brittle with partial patchy alopecia. There were similar changes in the eyebrows and eyelashes. The patient's growth was subnormal according to his age, but there was no mental deficiency. Teeth and nails were normal.

Complete blood picture, erythrocyte sedimentation rate, urine, stools, liver and renal function tests, serum proteins, blood sugar, urinary aminoacids, and x-ray chest did not reveal any abnormality.

Biopsy from the margin of a lesion showed hyperkeratosis, parakeratosis, irregular acanthosis and spongiosis at some places. Upper dermis showed perivascular infiltration of lymphocytes.

Microscopic examination of the scalp hair revealed changes of trichorrhexis invaginata.

The patient showed moderate improvement in the skin lesions after alternate use of local emollient and hydrocortisone cream with oral antihistaminic for two weeks. Later the patient was lost to follow-up.

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Comments

Ichthyosis linearis circumflexa (ILC) first described by Comel[5] in 1949, is characterized by erythematous migratory plaques with double edged scales, distributed mainly on the trunk and extremities. The lesions start at birth or shortly thereafter, with universal erythema, which lessens within a few years and develops into the typical lesions. Sometimes it is associated with pruritus.[1,2,6,7] A case of ILC was reported from Benghazi (Libya) by Kanwar et al[7] in 1987 with similar cutaneous changes and pruritus but without any abnormality of the hair. By 1969, five of the 14 reported cases of ILC had hair abnormality.[4] Dimitrova and Georgiewa[8] described one case with pili torti out of his 5 cases, Stankler and Cochrane[9] reported two sisters with trichorrhexis invaginata, and Schnyder and Wiegand[10] described trichorrhexis invaginata in two cases. By 1976, sixty cases of ILC were reported,[6] and those with trichorrhexis invaginata have been described as Netherton's syndrome.

Mental retardation has been reported in five cases of Netherton's syndrome.[1] We could not detect any mental retardation in our patient. His growth was slightly stunted, a feature not seen in previously reported cases.

In our patient the family history was suggestive of autosomal recessive inheritance. Similar mode of inheritance was found in most of the reported cases.[1]

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