CASE REPORTS

EPIDERMOLYTIC HYPERKERATOSIS WITH CLUB FOOT DEFORMITY

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A case of epidermolytic hyperkeratosis is reported. The patient had extensive verrucous scaling resembling porcupine man. There was associated bilateral foot deformity in the form of congenital club foot.

Key words : Epidermolytic hyperkeratosis, Club foot.

Epidermolytic hyperkeratosis is the least common of the four major types of ichthyosis.[1] The incidence of this rare disorder has not been reliably ascertained.[2] In the present communication a case of epidermolytic hyperkeratosis associated with bilateral congenital club foot is reported. Review of literature did not reveal any such association.

Case Report

A 14-year-old male student was admitted with verrucous scaling over the entire cutaneous surface almost since birth, along with deformities of the feet. The child was the product of a full term uncomplicated delivery. History of consanguinity in the parents was present, the maternal and paternal grandfathers of the patient being brothers. The child was excessively red and scaly at birth, and had repeated blisters especially on the legs which frequently got secondarily infected. The bony deformity became more prominent when the child started standing and walking. Milestones of physical and mental development were normal.

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The thick, dark, verrucous scales were present on most of the cutaneous surface. These were more pronounced on the knees and elbows. At places the scaling was almost warty and was present in a linear manner. There was thickening of the skin in the flexures and hyperkeratosis presented a furrowed appearance. The scaling was also quite pronounced on the neck. Palms and soles were normal and did not show any keratoderma. No bullae were observed. Face was not affected significantly, it had only dry and pityriasiform scaling. Scalp hair were normal. Nails showed prominent longitudinal ridging. Mucous membranes and eyes were normal.

The foot deformity was clinically and radiologically confirmed to be talipes equinocavovarus (club foot).

Routine laboratory investigations were normal. Histopathological examination of the skin from the back revealed variable degree of hyperkeratosis, papillomatosis and acanthosis. The cells of granular layer and prickle layer showed vacuolization. The keratohyalin granules in the granular cell layer were increased. The dermis showed a mild chronic inflammatory infiltrate.

The patient was treated with topical keratolytics with which there was some improvement.

However, on stopping the local therapy the scaling reappeared.

Comments

When the patient was admitted in the ward he had massive verrucous scaling on some areas of the body resembling almost the so called "porcupine man". According to Pinkus and Mehregan[3] severe cases of epidermolytic type of ichthyosiform erythroderma may show patches and streaks of heavy verrucous hyperkeratosis of the porcupine man type, as in our patient. Similar morphological changes occurring in a segmental pattern are termed as ichthyosis hystrix.[3] Families have been described in which both the diffuse and the segmental forms occurred. However, examination of the parents and sibs of the patient did not reveal any ichthyosiform changes.

The other interesting feature in our patient was the presence of bilateral club foot since birth. Though there are some rare syndromes in which ichthyosis may be associated with skeletal deformities,[1] club foot in association with epidermolytic hyperkeratosis has so far not been reported. In most cases of club foot, the aetiology is unknown.[4] Genetic factors are not operative.

References

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