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ICHTHYOSIS HYSTRIX– A CASE REPORT

Dermatology

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ABSTRACT

Ichthyosis hystrix, a rare autosomal dominant disorder of keratinization, usually presents with verrucous scaly lesions predominantly over the extensor aspects of the body. We report a solitary case of ichthyosis hystrix- Curth Macklin type with no family history and lesions involving the entire body with no specific extensor predilection.

KEYWORDS

Ichthyosis Hystrix, Systematized Epidermal Nevus, Rare

Introduction:

Ichthyosis hystrix otherwise known as systematized epidermal nevus[1] when generalized, is an uncommon disorder of abnormal epidermal differentiation[2] which has an autosomal dominant or sporadic inheritance[3]. The Curth-Macklin type is usually associated with diffuse palmoplantar keratoderma, but in our patient, there was widespread involvement, sparing the palms and soles.

Case report:

A 35 year old male patient presented to our skin out-patient department with complaints of dark, raised lesions all over the body for the past 7 years. Patient was born of a non-consanguinous marriage via normal vaginal delivery. There was no history of a similar condition in the family. He developed a small papular eruption over right gluteal region at 2 months of age. The lesions progressively enlarged to involve the entire body and has become significantly worse over the past 7 years. He gives a positive history of itching. The patient also experienced 9-10 episodes of seizures in the past. There was no history of any erythema or blister formation prior to the onset of these lesions.

On examination, bilateral, symmetrical, pigmented , verrucous, adherent, scaly plaques were noticed all over the body, equally over the flexor and extensor aspects, including the scalp. The lesions were not distributed in a blaschkoid pattern. The palms, soles and genitalia were spared. There was residual pigmentation over the forehead. The lesions on the lower limbs were extremely verrucous in appearance. Ophthalmological, neurological, ENT and skeletal abnormalities were ruled out.

Biopsy from the lesion showed hyperkeratosis, acanthosis and papillomatosis.

Based on the clinical and pathological findings, we arrived to a diagnosis of Ichthyosis hystrix.

Discussion:

Ichthyosis hystrix is an infrequently occurring genodermatosis with the defect in the gene encoding keratin 1[4]. Hystrix is derived from the Greek word which means "porcupine-like"[2], referring to the classical presentation of hyperkeratotic, verrucous, spine like lesions, chiefly involving the extensor aspects of the extremities but can also involve the entire body and given the term, systematized epidermal nevus. The lesions are usually distributed along the Blaschko's lines[5,6]. These plaques have a tendency to coalesce, imparting a cobble-stone appearance to the lesions.

Five types are included under ichthyosis hystrix : Curth-Macklin, Lambert, Rheydt, Brocq and Bäfverstedt[7].

Brocq type is associated with erythroderma and blistering prior to the onset of the scaly lesions[7]

Rheydt type is associated with deafness[7].

Bäfverstedt type is associated with diffuse involvement of the face with follicular hyperkeratosis[7].

In Lambert type, there is sparing of the face, genitalia, palms and soles. Patients are normal at birth and begin to develop lesions after 7 weeks of age[7].

The Curth-Macklin type has diffuse lesions all over the body involving the palms and soles[7].

Epidermal nevus syndrome is the association of systematized epidermal nevus with neurological, cardiovascular, ocular and urogenital abnormalities in addition to cutaneous manifestations[8].

Histopathology usually shows the characteristic features of hyperkeratosis, acanthosis and papillomatosis without the presence of perinuclear vacuolization and binucleate keratinocytes which differentiates this condition from epidermolytic hyperkeratosis(EHK)[2].

Another differential in addition to EHK is bullous congenital ichthyosiform erythroderma. Both these conditions can be ruled out by the presence/history of blisters and pathological examination.

Treatment is symptomatic and for cosmetic purposes. Topical keratolytics and emollients can be applied to bring down the scaling. Systemic retinoids have been found to be of some benefit[9].

CONCLUSION:

This case has been reported not only because of its rarity, but also because of its atypical presentation, wherein the lesions appeared diffusely without a blaschkoid distribution, spared the palms, soles and genitalia and was associated with a history of recurrent seizures emulating an epidermal nevus syndrome.

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CONFLICT OF INTEREST: The authors declare that they have no conflict of interest.

Legends to figures:

Figure 1: Bilateral symmetrical pigmented, verrucous plaques over the trunk and upper limbs

Figure 2: Extremely verrucous lesions over bilateral lower limbs

Figure 3: Histopathological picture under low power showing hyperkeratosis, acanthosis and papillomatosis.

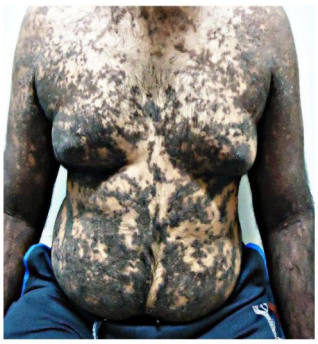


FIGURE 2

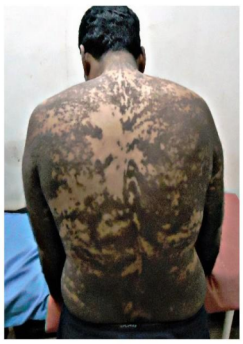
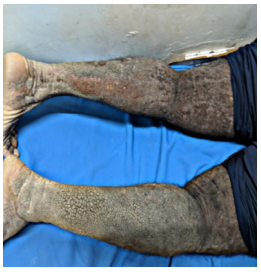
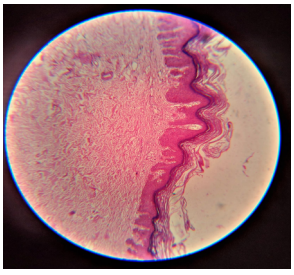


FIGURE 3





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