Ind J Dermatol Venereol Leprol 1995; 61 : 312-3

KERATITIS, ICHTHYOSIS AND DEAFNESS (KID) SYNDROME

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Keratitis, ichthyosis, deafness (KID) syndrome is a genetically determined disorder. The present case is having marked photophobia, bilateral corneal ulceration with vascularisation, neurosensory deafness and skin changes.

Key Words : Keratitis, Ichthyosis, Deafness

Introduction

Keratitis, ichthyosis and deafness (KID) syndrome is characterised by keratitis with corneal neovascularisation, hyperkeratosis and neurosensory hearing defect.[1] Burns recognised this entity in 1915 and described it as generalised congenital keratoderma with keratitis and deafness.[2] Skinner et al in 1981 proposed the name Keratitis, Ichthyosis, Deafness (KID) syndrome to this entity.[1] Since then about 40 cases had been described in literature.[3]

Case Report

A nine-year-old daughter of non-consanguineous parents was brought with photophobia and redness of eyes and peculiar skin appearance. The child was born at term after uneventful antenatal period by normal labour. There was no history of any drug intake or infection during the pregnancy. At about 3 months of age, the child developed generalised erythema and peculiar appearance of skin. From the age of seven occasional redness of the eyes and photophobia were noticed. The performance at school was below average.

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Physical examination showed dry hyperkeratotic and prickly skin. Perioral skin was thick and wrinkled. The skin over both elbows and knees was rugose. Chronic cheilitis and perleche were present. The dorsum of the hands and wrists showed an appearance similar to that of acanthosis nigricans. The palms and soles were hyperkeratotic. The patient had alopecia, sparse eyebrows and eyelashes. The teeth were normal. Her nails were thickened and white. There was moderate hypohidrosis and intolerance to heat.

The girl showed marked photophobia, bilateral corneal ulceration with vascularisation along the periphery. The visual acuity was reduced markedly. Slit lamp examination showed corneal epithelial and sub-epithelial opacity. Hearing was impaired and audiogram revealed neurosensory deafness. The systemic examination, complete haemogram, urinalysis and skeletal survey were all within normal limits.

Discussion

The association of cutaneous hyperkeratosis with neurosensory hearing defect and keratitis constitutes the KID syndrome.[4] The skin is usually hyperkeratotic and non-scaly.[5] Histopathologically there is saw-toothed epidermal morphology with basket weave hyperorthokeratosis and follicular plugs.[4]

This appearance is markedly different from the usual histopathology of ichthyosis and hence many authors believe that the term ichthyosis is not suitable for these hyperkeratotic lesions. In our patient also there is non-scaly hyperkeratosis with follicular keratosis and perioral wrinkling.

The clue to the diagnosis of KID syndrome is the dual sensory impairment.[5] Loss of visual acuity develops in the first year of life, resulting from corneal vascularisation and opacification of corneal stroma,[5] the cause of which is unknown. Neurosensory hearing defect also starts from early life. In our patient, even though there are marked visual and hearing disturbances, we are not sure about the time of their onset.

The syndrome is genetically determined and transmitted by an autosomal gene.[3] The scarcity of familial cases of the syndrome could be explained by a low rate of procreation in affected patients.

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