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Keratitis - Ichthyosis - Deafness Syndrome (KID): A Case at the Bamako Dermatology Hospital

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Abstract **Case Report**

Summary: The syndrome of keratitis - ichthyosis - deafness (KID) is a rare genetic disorder associating: keratitis, ichthyosis and deafness. We report a case at the dermatology hospital in Bamako (Mali). A 6-month-old boy consulted at the Bamako Dermatology Hospital for a generalized rash evolving since birth associated with psychomotor retardation. Dermatological examination showed warty-looking hyperkeratotic lesions on the trunk, keratoderma palmo -plantar of velvety aspect. Nails were thickened with brown color and onychodysplasia. The axillary folds were the seat of a cupboard covered with whitish plaster; the same lesions were found on the scalp. The ophthalmological examination noted corneal conjunctival xerosis and keratitis. The otorhinolaryngological examination revealed sensorineural deafness. Diagnosis of keratitis - ichthyosis - deafness syndrome (KID) was selected based on the clinical criteria (keratitis, ichthyosis and sensorineural hearing loss).

Keywords: Syndrome, keratitis, ichthyosis, deafness, Bamako, Mali.

INTRODUCTION

The syndrome of keratitis - ichthyosis - deafness (KID) is a rare genetic disorder associating: keratitis, ichthyosis and deafness. This syndrome has been known for a long time since it was first described in 1915 by Burns [1].

The mode of genetic transmission of this condition remained uncertain for a long time. The reported cases were most often sporadic, but the possibility of an autosomal dominant transmission was suspected in view of the observation of a few family cases [2].

This syndrome is a severe condition for several reasons. Cutaneous involvement is generally very unsightly and is frequently complicated by superinfection phenomena. The impact on social life is generally major [3].

We report a case at the dermatology hospital in Bamako.

OBSERVATION

A 6-month-old boy consulted at the Bamako dermatology hospital for a generalized eruption evolving since birth associated with psychomotor retardation. There was no notion of consanguinity, nor any family history of a similar condition. On examination, the child presented with delayed psychomotor development, photophobia and moderate acute malnutrition. Dermatological examination showed warty-looking hyperkeratotic lesions on the trunk (figure 1), keratoderma palmo -plantar with a velvety appearance (figures 2). Nails were thickened with brown color and onychodysplasia (figure 3). The axillary folds were the seat of a plaque covered with a whitish coating (figure 4); the same lesions were found on the scalp.

Figure 1

Figure 2

Figure 3

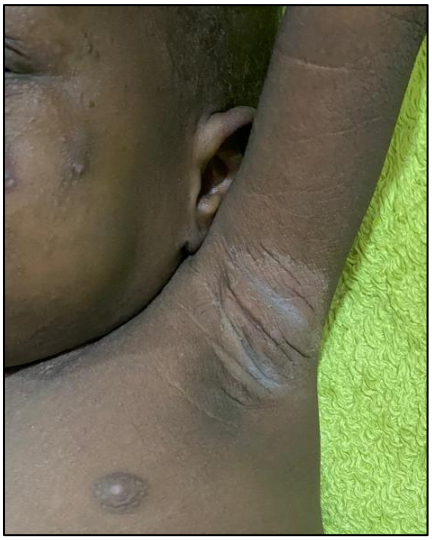

Figure 4

The tongue was red and the labial commissures were the seat of angular cheilitis. The ophthalmological examination noted corneal conjunctival xerosis and keratitis. The ENT examination found sensorineural deafness. The diagnosis of KID syndrome was made based on the clinical criteria (keratitis, ichthyosis and sensorineural hearing loss). The standard assessment was normal; the assessments (molecular analysis of the GJB2 gene, histopathology and immunohistochemistry) were not carried out. The evolution was marked by the occurrence of multiple abscesses and chronic infections that led to the abandonment of the marital home by the father, following social prejudice.

DISCUSSION

To our knowledge, this is the first case of the syndrome of keratitis - ichthyosis - deafness (KID) described in Mali. It is a severe condition and it for several reasons. Cutaneous involvement is generally very unsightly and is frequently complicated by superinfection phenomena. Keratitis can lead to blindness and deafness is usually severe. Finally, patients are at high risk of developing sometimes fatal squamous cell carcinomas. These most often occur between the ages of 30 and 40, on the lower limbs and buttocks. More exceptionally, squamous cell carcinoma of the tongue may occur [3-5].

Diagnosis is essentially clinical and confirmation is provided by molecular examination, which we do not have.

In terms of heredity, familial cases are more common than initially described. However, they remain a minority compared to sporadic cases, which could be explained by the fact that patients find it difficult to start a family because of their very unsightly appearance [3].

Our case is from a non-consanguineous marriage and without a family history of keratitis - ichthyosis - deafness (KID). This is probably a sporadic case. The management of this syndrome in low-income countries such as Mali represents a crucial challenge for the practitioner and an economic challenge for the family, hence the need to equip our various departments with genetic and immunohistochemical examinations.

CONCLUSION

The syndrome of keratitis - ichthyosis - deafness (KID) is a genodermatosis which sometimes poses a diagnostic problem in our structures given its rarity. The prognosis is grim and marked by recurrent infections.

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