

# Keratitis–ichthyosis–deafness (KID) syndrome in an adult in sub-Saharan Africa

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## ABSTRACT

KID syndrome is a congenital disorder combining keratitis, ichthyosis, and deafness. Herein, we report a case diagnosed in adulthood. A 29-year-old male patient with deafness and progressive loss of visual acuity in early childhood was consulted because of generalized scaly lesions that worsened over time. A skin examination revealed large blackish ichthyosiform scaly patches on the limbs, marked follicular keratosis on the trunk, palmoplantar keratoderma, small papules, sometimes hyperpigmented, on the mediofacial area, and significant xerosis. Dental anomalies such as malocclusion and several decayed teeth were present. The combination of ichthyosis lesions, deafness, and visual abnormalities led to the diagnosis of KID syndrome. Emollients and keratolytics were applied with a slight improvement in the skin lesions. The uniqueness of this observation was the late diagnosis of KID syndrome. Ichthyosis associated with neurosensory deficits should evoke KID syndrome despite the difficulty of molecular diagnosis in developing countries.

**Key words:** Keratitis–ichthyosis–deafness syndrome, Adult, Case report, Sub-Saharan Africa

## INTRODUCTION

Keratitis–ichthyosis–deafness (KID) syndrome is a rare genetic disorder combining keratitis, ichthyosis, and deafness [1]. Originally described by Burns in 1915, the acronym *KID* was proposed by Skinner in 1981, and its etiology was discovered by Van Geel et al. and Richard et al. in 2002 [2–5]. In addition to the characteristic clinical triad, dental abnormalities and susceptibility to infection are often reported. Approximately one hundred cases have been reported, including four cases in sub-Saharan Africa (Cameroon, Togo) [6–9]. Herein, we report a case diagnosed in adulthood.

## CASE REPORT

A 29-year-old male living in Abidjan was consulted because of generalized scaly lesions present since childhood. The onset of the symptoms was at the age of

one year. The non-pruritic scaly lesions were located on the dorsal aspect of the hands and feet with extension and thickening, associated with significant skin xerosis. At two years of age, the cutaneous symptoms were associated with severe deafness and mutism (with otolaryngologic follow-up). The patient also reported progressive loss of visual acuity and corneal opacity at four years of age. An ophthalmic examination revealed cataract and vascular keratitis. His medical history was unremarkable. The patient had no skin lesions at birth, no other chronic diseases, and no parental consanguinity. A physical examination revealed generalized scaly lesions on a sometimes-erythematous background. They were pityriasiform on the trunk and scalp, ichthyosiform, thick, hyperpigmented, and scaly on the upper and lower extremities (Figs. 1a – 1d). An examination of the neck revealed follicular keratoses (Fig. 2a) and palmoplantar keratoderma (Fig. 2b). The hair and nails were normal in appearance.

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**Figure 1:** Generalized scaly lesions: a) ichthyosiform scales on the left upper extremity; b) ichthyosiform scales of right upper extremity; c) ichthyosiform scales of the thighs (posterior surfaces); d) ichthyosiform squamous epithelium separating into flaps.



**Figure 2:** (a) Follicular keratosis of the neck. (b) Plantar keratoderma. (c) Vascular keratitis. (d) Multiple calculus, caries, malocclusion, tooth 41 in lingual position.

An examination of other systems, especially the ophthalmologic and stomatologic systems, revealed vascular keratitis (Fig. 2c) and multiple calculus deposits, caries and malocclusion of the lingual teeth (Fig. 2d). The diagnostic hypotheses were lamellar ichthyosis, progressive symmetric erythrokeratoderma and KID (keratitis–ichthyosis–deafness) syndrome. The association of ichthyosis lesions, deafness, and visual and dental abnormalities led to the diagnosis of KID syndrome. Dermatologic treatment was symptomatic with the use of emollients and keratolytics with a slight improvement in the skin lesions. ENT and ophthalmologic follow-up was continued. The patient was lost to follow-up after two months.

## DISCUSSION

KID syndrome is a rare ectodermal dysplasia characterized by autosomal dominant mutations of the GJB2 gene encoding connexin 26 [10]. Herein,

we report a case of KID syndrome associated with severe deafness, mutism, severe ichthyosis with plantar keratoderma, ocular involvement (cataract and vascular keratitis), and oral anomalies (calculus, caries, and malocclusion) in an adult. The majority of cases have been reported in children. Wonkam et al. (Cameroon) reported two cases in children aged two years, Barruet et al. (Togo) reported one case in a sixteen-year-old, and Kombaté et al. (Togo) reported one case in a nine-year-old [7-9]. To the best of our knowledge, this description of KID syndrome is the first of its kind in Côte d'Ivoire. The late diagnosis in our case could be explained by the lack of knowledge of this pathology by the general practitioners and the sociocultural prejudices of the patients. The low socioeconomic level of the patients would lead them to prefer the care of traditional therapists, which would favor delaying the diagnosis of the disease. We were unable to perform genetic analysis on the index case and his parents due to our limited technical resources. This analysis would have provided a diagnostic argument of certainty and confirmed the mode of transmission. Indeed, some authors have shown that KID syndrome is generally associated with sporadic inheritance, yet there are familial forms with autosomal dominant and recessive transmission [7,11-13,14]. In their work in Togo, Kombaté et al. suggested that KID syndrome described in a child from a consanguineous marriage was likely to be sporadic [9]. In our case, there was no concept of consanguinity. Genetic analysis of the parents in the Kombaté study and in our case would, therefore, have been useful. The hair and nails may be affected, as reported by Cammarata-Scalisi et al. and Caceres-Rios et al., in the form of progressive scarring alopecia and onychodystrophy [10,15]. In our case, the hair and nails appeared normal. Raghavon et al. (India) reported a case of a four-year-old with alopecia of the scalp, eyelashes, and eyebrows [16]. Kombaté

et al. (Togo) also reported diffuse scalp alopecia [9]. Dental anomalies, although not well described, may also be found in patients, as in our case. Caceres-Rios et al. found 25% dental abnormalities in their study in Mexico [15]. Multiple caries and pyogenic granulomas, right oral mucosal ulceration with missing teeth, and dental dysplasia were described in studies by Korolenkova et al. (Russia), Homeida et al. (U.S.), and Wonkam et al. (Cameroon), respectively [7,17,18]. KID syndrome is associated with an increased risk of chronic and opportunistic skin infections (bacterial or fungal) and, in severe cases, sepsis [13,19,20]. Benign and malignant skin tumors, such as trichilemmoma and mucosal squamous cell carcinoma, may develop in at least 12% of patients, as may squamous cell carcinoma from dermal or lingual lesions [11-13,20]. Treatment of skin lesions remains symptomatic. Treatment with retinoids may be attempted to improve the skin barrier, as well as the use of keratolytic agents and antifungal and antibacterial agents if there is a risk of infection [11,13]. In our case, the use of emollients and keratolytics allowed the skin lesions to begin to improve.

## CONCLUSION

KID syndrome remains a rare entity. We have reported the first case of KID syndrome in Ivory Coast. It is necessary to go beyond ichthyosis. Auditory neurosensory deficits and ocular involvement should be considered KID syndrome in our working context. A multidisciplinary collaboration would allow to enrich the semiology of the associated disorders. The difficulties in conducting genetic and molecular analyses in developing countries such as ours are an obstacle to confirming the autosomal transmission of this syndrome.

## Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

The authors certify that they have obtained all appropriate patient consent forms, in which the patients gave their consent for images and other clinical information to be included in the journal. The patients understand that their names and initials will not be published and due effort will be made to conceal their identity, but that anonymity cannot be guaranteed.

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