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Keratosis Follicularis Spinulosa Decalvans: Case Report

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Abstract **Case Report**

Follicular spinulosic decalvans keratosis is a rare X-linked disease affecting both the skin and eyes. The aim of this report is to describe this pathology which manifests itself as progressive scarring alopecia of the scalp, with keratosis pilaris.

Keywords: Follicular spinulosic keratosis, scarring alopecia, keratosis pilaris.

INTRODUCTION

Keratosis follicular spinulosa decalvans (KFSD) is a rare hereditary, X-linked keratinization disorder characterized by diffuse follicular hyperkeratosis, progressive scarring alopecia of the scalp, eyebrows and eyelashes. Very few cases of KFSD have been reported in the Moroccan literature; here we present a case of severe form of KFSD, a rare cause of scarring alopecia in a Moroccan child.

CASE REPORT

8 years old child, with no history, who had diffuse hair loss since the age of 3 months with hair loss of eyelashes and eyebrows, and photophobia. Dermatological examination notes the presence of alopecic patches of irregular contours confluent in places at the frontal level. occipital temporo-parietal with presence of a tuft of hair at the vertex, complete depilation of eyelashes and eyebrows, on dermoscopy: multiple black dots, leukotrichic hairs, split hairs, sleeve appearance, follicular papules at the forehead with erythema on the cheeks, the presence of beauty lines at the nails, with palmoplantar keratoderma, a biopsy of the scalp showed follicular ostias dilated by keratotic plugs sheltering or not hair shafts, the residual hair follicles are often dilated by keratotic plugs with atrophic or altered appearance, their epithelial sheaths are often atrophic, the dermis also harbors some light inflammatory infiltrates, mainly lymphohistiocytic, responsible for superficial capillary vessels and sometimes discreetly responsible for a few follicles without lichenoid reaction, concluding in follicular keratosis siemens decalcifying spinolosis. The patient benefited from a biological assessment including a blood count and liver function returning to normal then put on oral retinoids but the patient does not have the means.

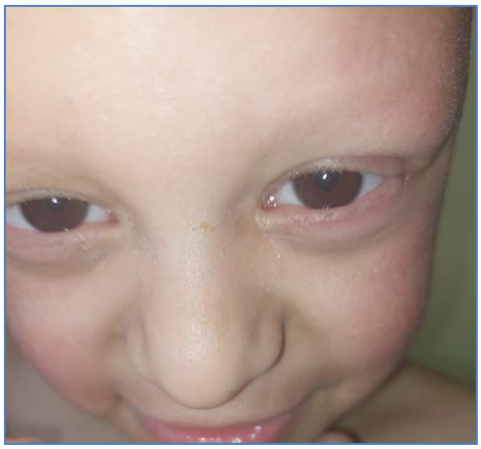

Figure 1: Clinical photo showing depilation of eyelashes and eyebrows

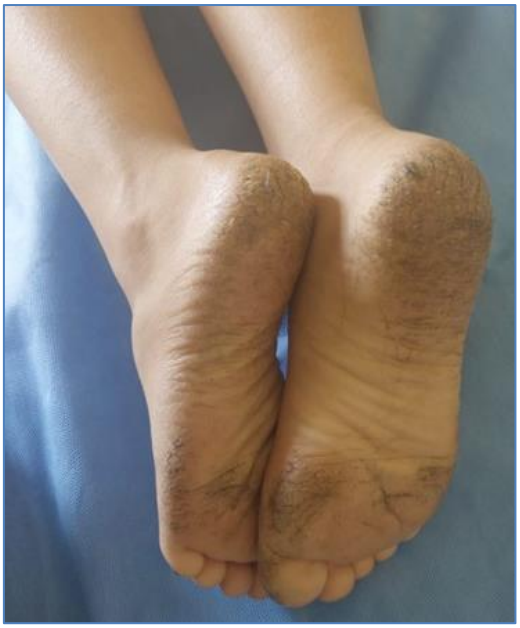

Figure 2: Clinical photo of plantar keratoderma

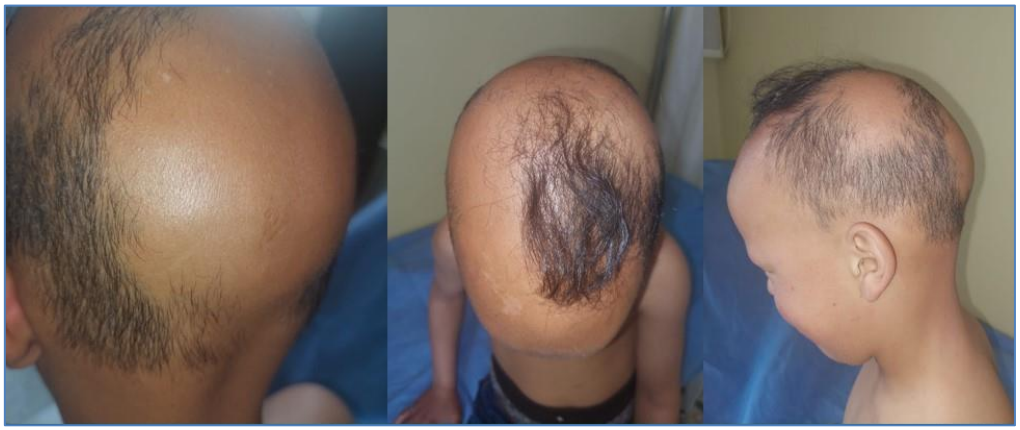

Figure 3: Alopecic patches with irregular contours confluent in places at the frontal occipital temporoparietal level with the presence of a tuft of hair at the vertex

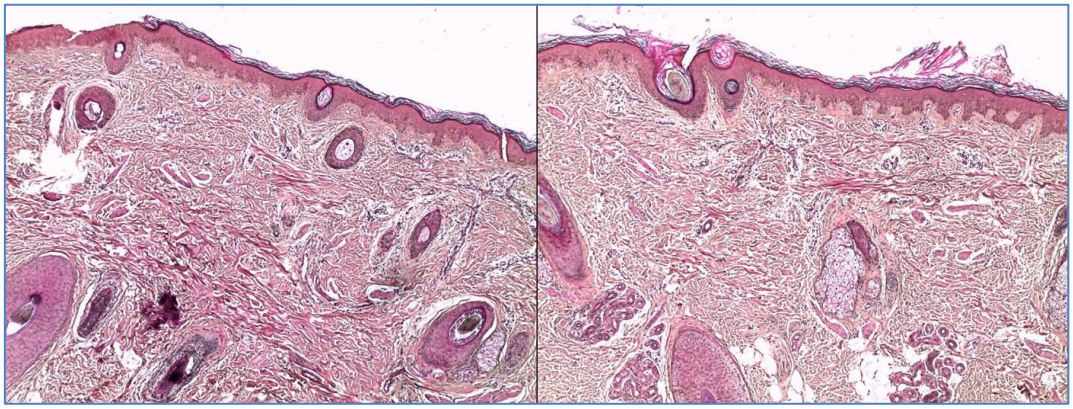

Figure 4: HES G x 50: Follicular ostia dilated by a keratotic plug with or without a hair shaft, reduction of pilosebaceous follicles to dermal level (non-fibrous), atrophic hair sheaths, hair shafts sometimes absent or dystrophic, slight non-lichenoid mononucleated inflammatory infiltrates

DISCUSSION

Keratosis follicularis spinulosa decalvans (KFSD) is a type of scarring alopecia that is grouped under a broader term keratosis pilaris atrophicans (KPA). This condition, along with keratosis atrophicans faciei and atrophoderma vermiculatum, represent closely related disorders [2], first described by Macleod [6], but

the term KFSD was used by Siemens in 1926 The gene has been mapped to Xp21.2-p22 The suggested candidate gene is the gene for membrane-bound transcription factor protease site 2, required for cleavage of sterol regulatory element-binding proteins (SREBPs). An alteration of the cleavage of SREBP impairs cholesterol and lipid homeostasis in the skin, causing defective epidermal differentiation; sporadic cases have also been described [3, 4]. This disease begins in the first years of life, with follicular hyperkeratosis first on the face, then progresses to the trunk and limbs and can spread to other parts of the body. Palmar-plantar keratoderma, photophobia, corneal abnormalities and atopy may be associated [5]. The clinical picture is clearer in boys than in girls. biopsy is generally unnecessary. Recently trichoscopic signs have been reported consisting of diffuse perifollicular hyperkeratosis, with absence of follicular ostia. tufted folliculitis with visualization of elongated vessels. Trichoscopy of the eyebrows and eyelashes presented yellow dots and dystrophic hairs [6]. Histological examination would show a voluminous horny plug dilating and obstructing the pilosebaceous ostium, a peripheral inflammatory reaction with fibrosis or granulomatous infiltrate [7]. The most important differential diagnoses of follicular spinulosa decalvans keratosis are: KID syndrome (keratosis, ichthyosis, deafness), atrichia with papular lesions and hereditary mucoepithelial dystrophy [5], Graham Little Piccardi Lasseur syndrome (GPLLS) [4].

KFSD has been described in association with various conditions, including cutis laxa, "big pinnae", clinodactyly, arachnodactyly, Noonan syndrome, deafness, aminoaciduria, mental retardation, Down syndrome, congenital glaucoma, lenticular cataract, hepatomegaly and bilateral inguinal hernia, a greater propensity for recurrent systemic bacterial infections has also been reported [8]. Treatment of KFSD is generally unsatisfactory and difficult and, until now, no effective therapy is known. Etretinate and isotretinoin have been reported, but should be used early in the disease, dapsone has been tried with good results. Topical treatment mainly includes keratolytics and emollients [9]. Laser-assisted hair removal with the non-Q-switched long-pulse ruby laser has been shown to be useful in progressive or recalcitrant KFSD [10].

CONCLUSION

Spinal decalving keratosis is an unusual form of keratosis pilaris, which must be considered in the face of any keratosis pilaris associated with scarring alopecia in children.

Conflict of Interest: The authors declare that they have no conflicts of interest.

Source of Support: Nil

Conflict of Interest: The authors declare that they have no conflicts of interest.

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