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Keratosis Follicularis Spinulosa Decalvans in a Female Patient- A Case Report

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Abstract
Keratosis follicularis spinulosa decalvans is X-linked genodermatosis with occasional autosomal dominanant inheritance. It is characterized by keratotic papules and cicatricial alopecia mainly affecting the males with females being carriers with milder symptoms. Other features of the syndrome include widespread keratosis pilaris, progressive cicatricial alopecia of the scalp, eyebrows and eyelashes, photophobia, blepharitis, conjunctivitis and corneal dystrophy. Here we present a rare case of KFSD in a female.

Keywords: *keratosis follicularis spinulosa decalvans, follicular keratotic papules, scarring alopecia.*

Introduction

Keratosis follicularis spinulosa decalvans is a rare genodermatosis chiefly characterized by widespread keratosis pilaris, progressive cicatricial alopecia of the scalp, eyebrows and eyelashes and a preponderance in males. Photophobia, blepharitis, conjunctivitis and corneal dystrophy are characteristic ancillary findings. It is most often inherited as an X-linked trait mainly affecting the males with females being carriers having milder symptoms[1,2]. We present a rare case of keratosis follicularis spinulosa decalvans in a female.

Case Report

A 21-year-old girl born from non- consanguineous marriage presented with the complaints of itchy, skin colored papules over the body with loss of hair since childhood. The patient was normal at birth. At the age of six months, she started developing pinhead sized skin colored raised lesions over the eyebrows followed by loss of eyebrow hair and eyelashes. This was followed by appearance of similar lesions over the scalp followed by loss of hair in patches. After 3-4 months, similar itchy lesions started developing over the rest of the body in the cephalocaudal manner. There were no eye

complaints. The patient never developed axillary hair. Pubic hair developed at the age of 13 years. The nails, mucosae, palms and soles were unaffected. On examination, there was generalized involvement of the body with relative sparing of the central face, flexures, hands and feet in the form of hyperpigmented to skin colored discrete follicular papules with small, dirty white loosely adherent scaling and punctate atrophy at places. Over the scalp, there were similar lesions with patches of scarring at places. Ocular examination was normal. Histopathological examination revealed hyperkeratosis and spongiosis with exocytosis of several lymphocytes and a conical parakeratotic scale over the spongiosis. With the above findings, a diagnosis of keratosis follicularis spinulosa decalvans was made. The patient was started oral isotetinoin, topical salicylic acid-steroid combination, moisturizers and anti-histaminics.

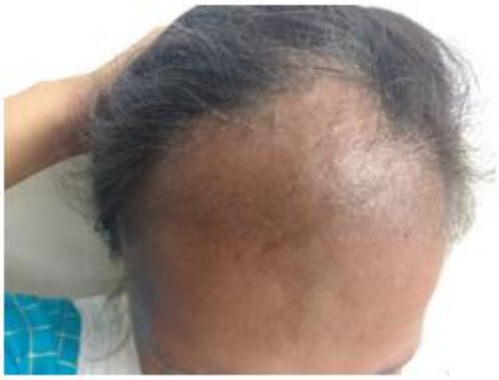
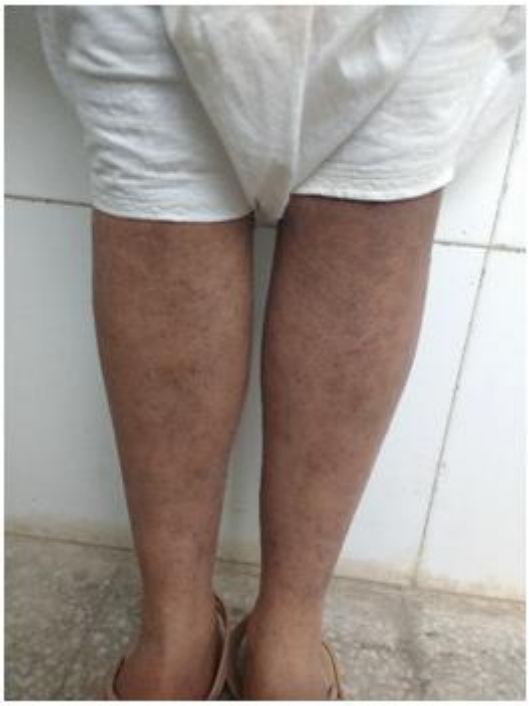


Figure 1 & 2 showing scarring alopecia of the scalp with keratotic follicular papules and absent axillary hair



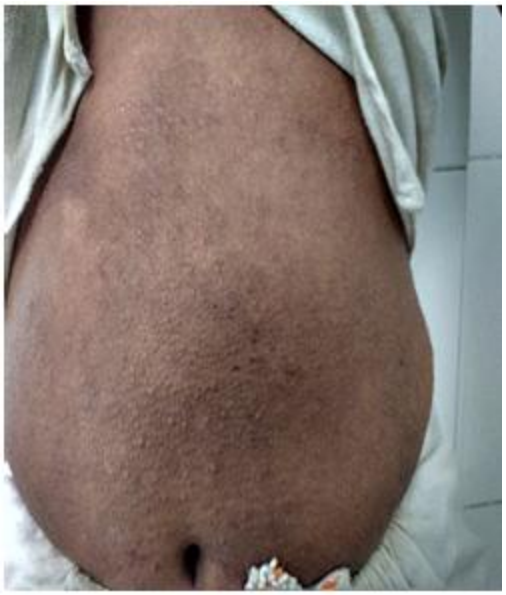
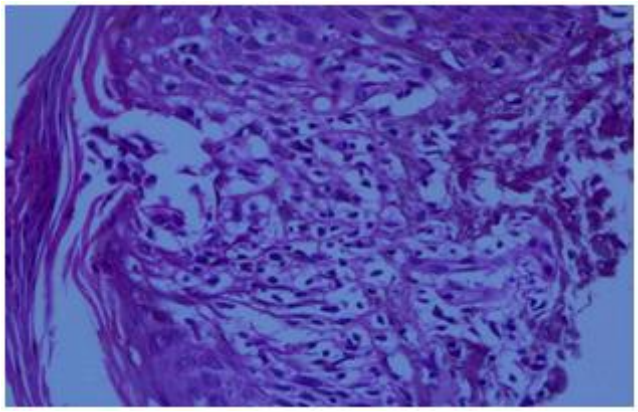


Figure 3 & 4 showing scaly keratotic papules over the trunk and legs.



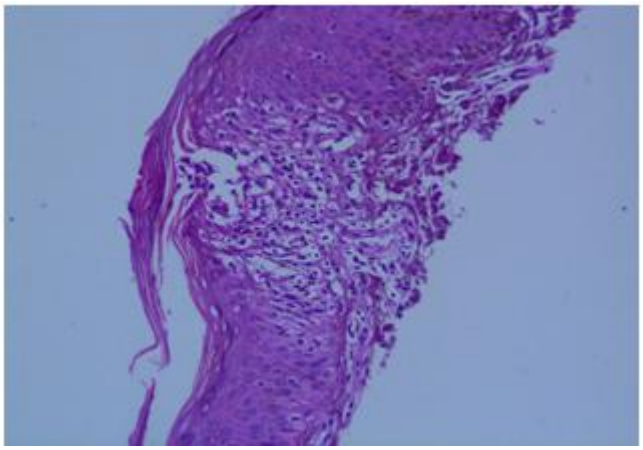


Figure 5 & 6 showing hyperkeratosis and lymphocytic infiltration in the dermis on histopathology.

Discussion

Keratosis Follicularis Spinulosa Decalvans (KFSD) was first described by Macleods. Siemens first used this mainly affects males with females being carriers having milder symptoms. Autosomal dominant inheritance has been suggested in familial cases. The process of lyonization (non-random X chromosome inactivation) may explain expression of KFSD in females[2,3]. The candidate gene suggested is the membrane-bound transcription factor protease site 2 (MBTPS2) which is required for cleavage of sterol regulatory element-binding proteins (SREBPs). Altered SREBP cleavage impairs cholesterol and lipid homeostasis in the skin causing defective epidermal differentiation[4].

KFSD comes under the umbrella term Keratosis pilaris atrophicans (KPA) which includes three rare and distinct clinical entities representing the scarring types of keratosis pilaris. These include Keratosis pilaris atrophicans facei (KPAF), Atrophoderma Vermiculatum (AV) and Keratosis Follicularis Spinulosa Decalvans (KFSD)[5]. KFSD startsin the early years of life starting on the face and progressing towards the trunk and limbs.It begins with numerous horny follicular plugs and milia first on the nose and cheeks and later on the eyebrows, scalp, neck and body. Erythema is variable and often very faint. Scarring alopecia of the scalp, eyebrows and eye lashes becomes apparent in childhood and progresses until puberty. Facial lanugo hair is absent. There is keratosis pilaris of the body that resembles that of non-atrophic type but many follicles appear empty. Axillary and pubic hair thinning is frequently observed. Patches of eczema, particularly on the scalp may be seen. After puberty, the disease becomes static. Photophobia is a regular feature caused by sub epithelial opacities in the bowman's membrane and is found predominantly in children. Visual prognosis is good[6].Other associated features include palmoplantar keratoderma with predilection to calcaneal region, high periungual cuticles, atopy, deafness, mental retardation, acne keloidalis nuchae and tufted hair folliculitis, aminoaciduria and woolly hair[3,4].

The differential diagnosis includes Ichthyosis follicularis alopecia photophobia (IFAP) syndrome, lichen planopilaris and lichen spinulosus. IFAP syndrome is characterized by non-scarring alopecia in contrast to KFSD[1,2]. Furthermore, ocular features like photophobia and corneal dystrophy are essential for the diagnosis of IFAP[3]. Graham Little Piccardi Lasseur Syndrome, a variant of lichen planopilaris presents with scarring alopecia of the scalp, non-scarring alopecia of axillae and pubic region and keratotic follicular spinous papules over a body. The distinction from KFSD is made mainly on histopathology which reveals changes suggestive of lichen planus including vacuolar degeneration and interface dermatitis[3]. Lichen spinulosus has a predilection for acral areas unlike keratosis pilaris, which is frequently limited to the upper aspects of the arms and legs. Furthermore, the horny spine characteristic of lichen spinulosus can be removed, leaving behind a tiny funnel-like orifice in the papule, whereas an entire individual lesion can be removed with the plug in keratosis pilaris[2].

So far no effective therapy is known to work for KFSD. Frequent application of topical keratolytics and emollients improves skin texture. Antibiotics such as tetracyclines, sulfonamides (dapsone), macrolides, penicillins and rifampin have been used at therapeutic doses and found to be ineffective. Topical and intralesional corticosteroids have been tried and cause only transient improvement. Oral acrtretin, etretinate and isotretinoin have also been

used but with variable results[5]. Divya et al[8] have reported excellent improvement with oral isotretinoin in a female patient of KFSD. It is likely that retinoids, which are useful in disorders of keratinization, act by down regulating the process of follicular hyperkeratosis and inflammation. Laser assisted hair removal with the long-pulse non-Q-switched ruby laser has been found to be useful in progressive or recalcitrant KFSD[5].

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