GENETICS AND GENODERMATOSES

KERATOSIS FOLLICULARIS SPINULOSA DECALVANS: DIAGNOSIS AND THERAPEUTIC EVALUATION

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Background: Keratosis Follicularis Spinulosa Decalvans (KFSD) is an X-linked genodermatosis characterized by scarring alopecia and follicular hyperkeratosis.

Material and Methods: This paper reviews the different aspects of KFSD, including pathogenesis, clinical, histological, differential diagnosis and different therapeutic modalities and their impact on the prognosis of the disease.

Results: A nine year old male and his sister five years old who born from a first-degree consanguineous marriage visited our outpatient department with complaints of rough skin over the scalp and over the body since five years for the boy and two year for the girl in association with total loss of scalp and eyebrow hair in the boy and partial loss in the girl. At birth, their parents noted the absence of scalp and eyebrow hair, which gradually, over the next three to four years grew to some measure and eventually became scanty. Physical examination disclosed multiple follicular flesh-colored horny papules over the scalp, eyebrows, cheeks and both upper and lower limbs. A closer view of the scalp, cheek and eyebrow revealed fine scaling and areas of scarring alopecia, punctuate atrophy , hair loss of eyebrow in the boy and hair loss of the lateral half of eyebrows in the girl. The teeth, nails, palms and soles were found to be normal. The boy had history of photophobia but not in the girl

Conclusion: So far no effective therapy is known to work for KFSD. Frequent application of topical keratolytic agents and emollients improve 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 were tried but caused transient improvement. Etretinate and isotretinoin have also been used but with variable results.