Case Report

Keratosis follicularis spinulosa decalvans

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Abstract A case of keratosis follicularis spinulosa decalvans in young Kashmiri male is presented. This rare disorder with genetic and clinical heterogeneity, inherited in an X-linked recessive pattern is secondary to mutation in SAT1 gene located at Xp22.1. Although multigenerational families with male to male transmission suggestive of autosomal dominant inheritance have been reported, we present a case with no family history.

Key words
Keratosis follicularis spinulosa decalvans, young male.

Introduction

Keratosis follicularis spinulosa decalvans (KFSD) is a rare disorder of keratinization characterized by spiny follicular hyperkeratosis resulting in scarring alopecia, with ocular involvement.[1-7] It was first reported by Lameris[2] and described by Siemens.[3] The disease may be sporadic or familial and caucasions are more commonly affected.[5] We here describe this rare disorder in a young male patient.

Case Report

A 27-year-old man presented with the complaints of follicular skin eruptions since three years of age. The lesions started initially over the trunk and gradually extended over the face, neck, scalp and limbs (**Figure 1**). The lesions on the scalp and face turned pustular which healed with scarring alopecia. By the age of 12 years, the patient had lost most of the scalp hair. Patient also reported redness and watering of both eyes with photophobia since early childhood. The patient was born after a full-term normal delivery. There was no history of similar lesions in any of the patient's sibs.

Examination revealed cicatricial alopecia over the scalp and face with small, discrete, spiny papules with follicular plugging present on a normal looking skin over the trunk and limbs. The nails were normal while slight hyperkeratosis was seen over the palms and soles. Redness of the conjunctiva was seen in both eyes.

Routine hematological and other laboratory studies were normal. Histopathological examination of skin biopsies obtained from the trunk showed prominent hyperkeratosis and follicular plugging with mild inflammatory infiltrate in the dermis. Based on these clinical and histological findings the patient was diagnosed as a case of KFSD and started on low dose oral isotretinoin. Significant improvement was noted after three months.

Discussion

Keratosis follicularis spinulosa decalvans (KFSD) is a rare disorder of keratinization

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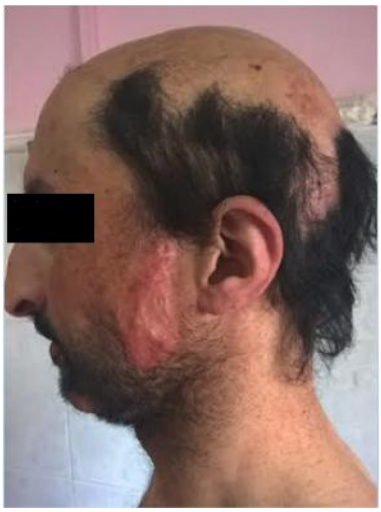

Figure 1 Scarring alopecia on scalp and left cheek.

characterized by involvement of skin and eyes. First reported by Lameris and described by Siemens, the disorder may occur in both sporadic or familial forms with apparent sparing of females.[1-7] The diagnosis of KFSD in our patient was made on the basis of characteristic cutaneous, ocular and histological features. Other rare abnormalities that have been reported in association with KFSD include deafness, physical and mental retardation, hypoplastic nails, failure to thrive and recurrent infections.[8,9] None of these features were seen in our patient.

The treatment of KFSD is frustrating, although several treatment modalities have been reported to be beneficial. These include topical corticosteroids, intralesional corticosteroids, oral retinoids (etretinate or isotretinoin), tetracycline and dapsone. Hair transplantation is an option in appropriate candidates with burnt out scalp disease.[1,5,10,11] In our patient significant improvement was seen with oral isotretinoin (0.25 mg/kg per day for 3 months) therapy and he is planned for hair transplantation for the management of scalp alopecia.

Dermatoscopy showed decreased hair density with loss of follicular openings, hyperkeratotic perifollicular white scales, perifollicular erythema and occasionally perifollicular pustules.

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