

A Case of Keratosis Follicularis Spinulosa Decalvans

BİR KERATOZİS FOLİKULARİS SPİNULOZA DEKALVANS OLGUSU

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Summary

Keratosis follicularis spinulosa decalvans (KFSD) is characterized by follicular hyperkeratosis with inflammation followed by atrophy and refers to extensive keratosis pilaris associated with scarring alopecia of the scalp. Some cases show an associated corneal opacity and photophobia. Because it is seen rarely, we report a case of 17-year-old boy with follicular keratotic papules of the face, scalp, trunk and limbs, and cicatricial alopecia in the scalp. Physical examinations showed mild mental motor retardation and central corneal stromal opacity in the eyes. Based on the clinical findings and histopathologic examination, the patient was diagnosed as KFSD.

Key Words: Keratosis follicularis spinulosa decalvans,
Keratosis pilaris atrophicans,
Mental retardation, Corneal opacity

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Özet

Keratozis folikularis spinuloza dekalvans (KFSD) foliküler hiperkeratoz ve inflamasyonu takiben atrofi ile karakterize olup, yaygın keratozis pilaris ve baş saçlı derisinin skatrisiyel alopesi ile kendini gösterir. Bazı olgularda korneal opasite ve fotofobi de birlikte bulunabilir. Nadir görülmesi nedeniyle yüz, baş saçlı derisi, gövde ve ekstremitelerinde foliküler keratotik papüller ve baş saçlı derisinde skatrisiyel alopesisi olan 17 yaşında bir erkek olgu sunuyoruz. Fizik muayenede orta derecede mental motor retardasyon ve gözlerde santral stromal korneal opasite saptandı. Klinik bulgular ve histopatolojik inceleme sonuçlarına dayanılarak olguya KFSD tanısı kondu.

Anahtar Kelimeler: Keratozis folikularis spinuloza dekalvans,
Keratozis pilaris atrofikans,
Mental retardasyon, Korneal opasite

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Keratosis pilaris atrophicans (KPA) defines a group of cutaneous disorders characterized by follicular hyperkeratosis and scarring (1). KPA encompasses all 3 disorders: keratosis pilaris atrophicans faciei (KPAF), atrophoderma vermiculatum and keratosis follicularis spinulosa decalvans (KFSD) (2,3). KFSD is characterized by follicular hyperkeratosis with inflammation followed by atrophy

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and refers to extensive keratosis pilaris associated with a scarring alopecia of the scalp (2). Here we report a case of KFSD because it is seen rarely.

Case

A 17-year-old boy was referred for evaluation of keratotic papules in the skin and scarring alopecia of his scalp. Onset of the skin disease was in early childhood but scalp involvement had occurred in the teen years. Over the years, he has had recurrent flares of pustular folliculitis in the scalp. On the last occasion, Staphylococcus aureus was isolated and the pustules resolved with a course of systemic antibiotic therapy. Family history disclosed follicular keratotic papules in his sister on the upper and lower extremities. The findings of dermatologic examination were remarkable for fol-

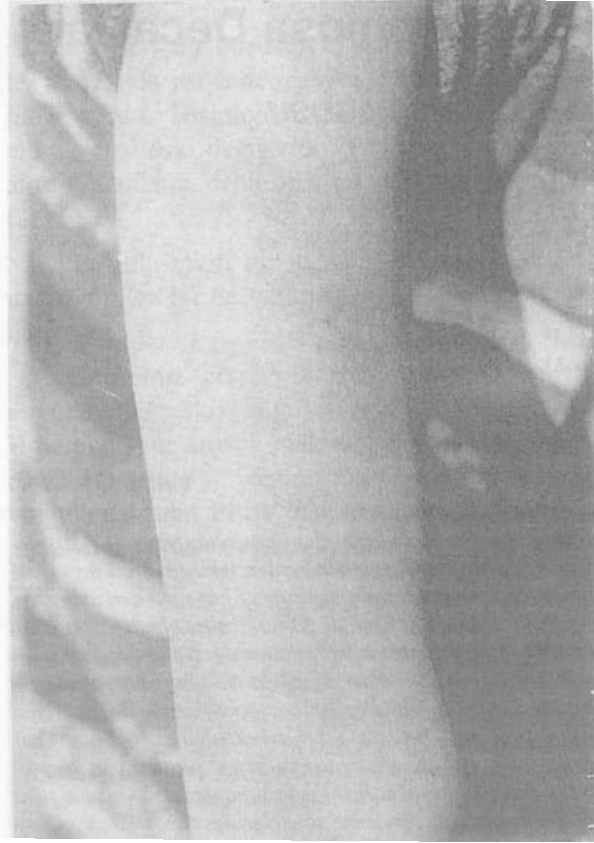


Figure 1. Follicular keratotic papules of the upper extremity.



Figure 2. Follicular keratotic papules and cicatricial alopecia of the scalp.

licular keratotic papules of the face, scalp, trunk and limbs (Figure 1), and cicatricial alopecia in the scalp (Figure 2). Physical examinations showed

mild mental motor retardation and central corneal stromal opacity in the eyes. Routine biochemical and hematological examinations were normal. The biopsy specimen of the trunk showed diffuse sclerosis of dermis (Figure 3) and of the scalp showed destruction and atrophy of hair follicles (Figure 4). The patient was diagnosed as KFSD and oral acitretin (0,5mg/kg) was given.

Discussion

KFSD, also called keratosis follicularis decalvans, was described in 1905 for the first time by Lameris in the Netherlands (4). Many cases occur sporadically; but severe manifestations are found in males, thus supporting an X-linked inheritance pattern (2). Symptoms of KFSD were not seen in male and female members of our case's family however there were keratotic papules in his sister. Symptoms are not present at birth but may develop in early childhood. The male patients generally improve

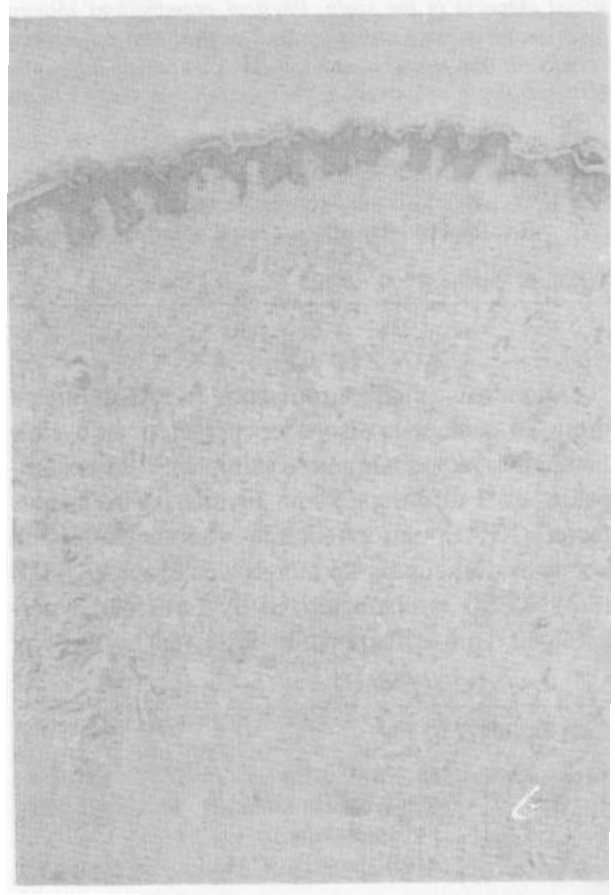


Figure 3. Atrophy and loss of the hair follicles and dermal fibrosis (H.E X 40).

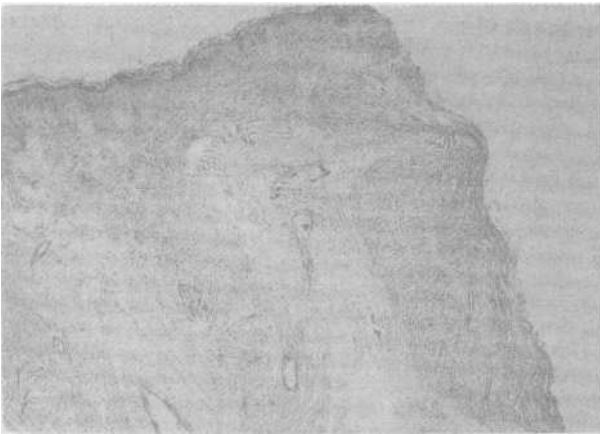


Figure 4. Hair follicle damage, perifollicular fibrosis and multinucleated foreign body giant cells were seen (H.E X 40).

spontaneously at puberty. The end stage of follicular papules in KFSD is characterized by atrophy and associated with loss of hair, especially from the scalp, eyebrows and eyelashes (3). Cicatricial alopecia of the scalp and eyebrows starting during childhood or up to early adolescence is the hallmark of the disorder (2). Some authors indicated clinical and genetic heterogeneity in KFSD. One of the major problem is a variant characterized by persisting pustular elements, especially on the scalp and instead of improvement, this variant exacerbates in puberty. Oranje et al (4) suggested that a better name for this form would be folliculitis spinulosa decalvans (FSD). In our case, pustular eruptions were first seen in puberty, therefore he could also be accepted as FSD. The cause of the recurrent folliculitis is still uncertain. *Staphylococcus aureus* may be isolated from the pustules, as in our case. An abnormal host response to an infection of the follicle has been postulated as an etiologic factor (2).

Many patients show marked but not pathognomonic photophobia and corneal dystrophy associated with the condition (3-5). Ophthalmologic examination showed central corneal stromal opacity in our case. The process is also associated with hyperkeratosis of the calcaneal regions of the soles and high eponychium of the fingernails (3). There was no hyperkeratosis of the soles and high eponychium of the finger nails in our patient.

Although physical examination showed mild mental motor retardation in our patient, to the best of our knowledge is not mental retardation associated with KFSD. On the other hand association of KPAF and different ectodermal defects, multiple congenital anomalies, atrophy and mental retardation were reported (6). We think that there is much confusion on KPA entities, especially because different overlapping syndromes have been described.

No effective therapy is currently available for this inflammatory hyperkeratotic follicular syndrome. Systemic antibiotic therapy will often prevent further extension of the disease, but only for as long as it is administered (2). Response to therapy including keratolytics and corticosteroids is limited (1). Isotretinoin and tretinate are moderately effective (3). Isotretinoin was used in four patients for 4 months at 1 mg/kg. Three of the patients had no to slight improvement, while one flared. Tretinoin cream 0,1% enhanced the redness when used alone but well tolerated when used in combination with a topical corticosteroid (1). Dermabrasion and laser resurfacing techniques may potentially be helpful. Atrophy in the final stages of the disease cannot be treated by anything other than grafting (3). After one month of acitretin therapy (0,5mg/kg), our patient gave up the retinoid therapy because of the desquamation of the palms and soles. There was no improvement in KFSD lesions during the therapy period.

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