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KERATOSIS FOLLICULARIS SPINULOSA DECALVANS ASSOCIATED WITH ACNE KELOIDALIS NUCHAE

Ilteris Oguz Topal[1], Ilknur Mansuroglu Sahin[2], Betül Berberoglu[3], Mehmet Ozer[2]

[1]*Department of Dermatology, Okmeydani Research and Training Hospital, Istanbul, Turkey*
[2]*Department of Pathology, Okmeydani Research and Training Hospital, Istanbul, Turkey*
[3]*Department of Pathology, Batman State Hospital, Istanbul, Turkey*

Corresponding author: Dr Ilteris Oguz Topal *drilteris@yahoo.com*

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Abstract
Keratosis follicularis spinulosa decalvans (KFSD) is a keratinization disorder characterized by diffuse follicular hyperkeratosis, progressive cicatricial alopecia, corneal dystrophy, and photophobia. Acne keloidalis nuchae (AKN) is a syndrome of chronic folliculitis that manifests as follicular-based pustules and papules on the occipital region of the scalp, which may eventually lead to cicatricial alopecia.
Various diseases such as cutis laksa, deafness, aminoaciduria, mental retardation, and atopy have been reported to be associated with KFSD, but AKN is a rare cutaneous manifestation. Herein, we report the case of a patient with KFSD associated with AKN. He was presented to our clinic with follicular-based pustules and papules that had been progressively advancing for five years that were now manifesting as cicatricial alopecia.

Key words: Keratosis follicularis; cicatricial alopecia; acne keloidalis nuchae

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Introduction

In 1905, Lamaris first described keratosis follicularis spinulosa decalvans (KFSD) as ichthyosis follicularis [1]. Later, Siemens provided more detailed phenotypic characteristics of the disease and first used the term KFSD in 1926 [2]. This condition is characterized by diffuse keratosis pilaris, cicatricial alopecia, and photophobia and is associated with cutis laksa, deafness, aminoaciduria, mental retardation, and atopy [3]. However, only very rarely has acne keloidalis nuchae (AKN) been associated with KFSD in the literature. Herein, we report a case of KFSD with acne keloidalis lesions.

Case Report

A 22-year-old male patient was admitted to our outpatient clinic because of hair loss that had begun five years earlier. He had consulted with several doctors and had used various antibiotics for what was considered to be acne vulgaris. The patient, who had also been treated with oral doxycycline for a period of time, said that he had received some benefits from this treatment but that the hair loss was nevertheless gradually increasing.

A dermatological examination revealed a cicatricial plaque with a diameter of 7x8 cm, follicular tufting, especially on the periphery of the plaque, erythematous papules and pustules, and partial follicular crusts on the vertex of the scalp (Fig. 1A). There was also a wide range of keloidal hard papules and pustules measuring a few millimeters in diameter on the patient's neck (Fig. 1B). In addition, persistent erythema was detected on the cheeks, and millimetric follicular papules were present on his face, including the eyebrows (Fig. 2A). The thinning of his lateral eyebrows was also remarkable. Furthermore, diffuse millimetric follicular papules, suggestive of keratosis pilaris, were present on the trunk and extremities (Fig. 2B). However, the patient's oral mucosa and nails were normal. In the biopsy specimen taken from the pustular lesion on the vertex region, the epidermis was normal, but prominent lymphocytic inflammation was present which was destroying the follicle epithelium. In addition, neutrophils and plasma cells were seen in the dermis (Fig. 3A, B).
The biopsy taken from the neck showed epidermal acanthosis and perivascular mixed- type infiltrates along with neutrophils and fibrosis in the dermis (Fig. 4).

The stains were negative for bacteria and fungi, but the clinical and histopathological features of the nuchal lesions were consistent with AKN.

Moreover, an ophthalmological examination showed mild blepharitis, and a topical antibiotic was prescribed for treatment. We also established that there were no similar clinical features in the family of the patient, and the findings of a psychiatric evaluation were normal. The patient said that the lesions, which were consistent with keratosis pilaris, had been present since his childhood. Taking into account all of the clinical and laboratory results, we then diagnosed the patient with sporadic KFSD and initiated isotretinoin treatment.

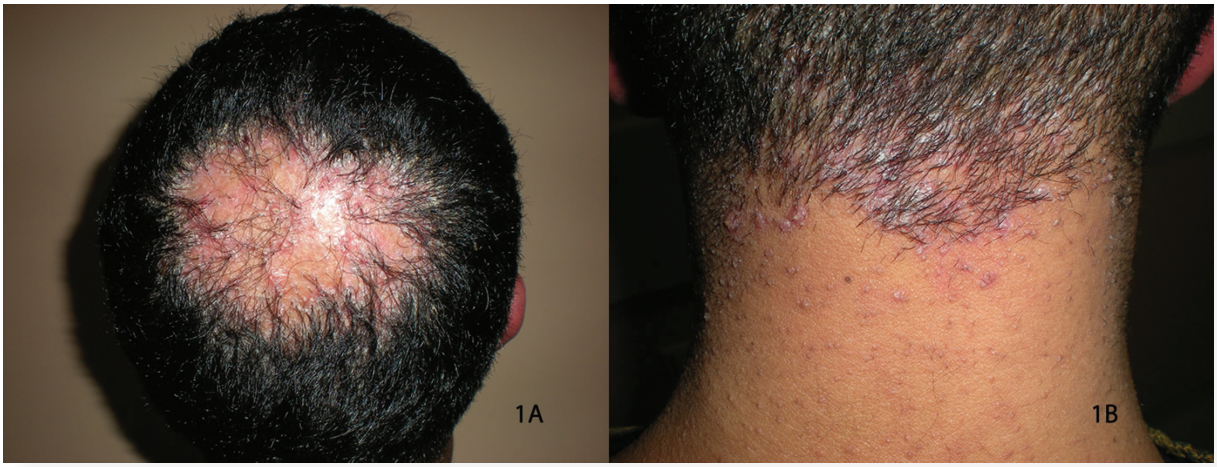

Figure 1A. Vertex of the scalp with scarring alopecia and tufted folliculitis. **B.** Acne keloidalis affecting the nuchal region.

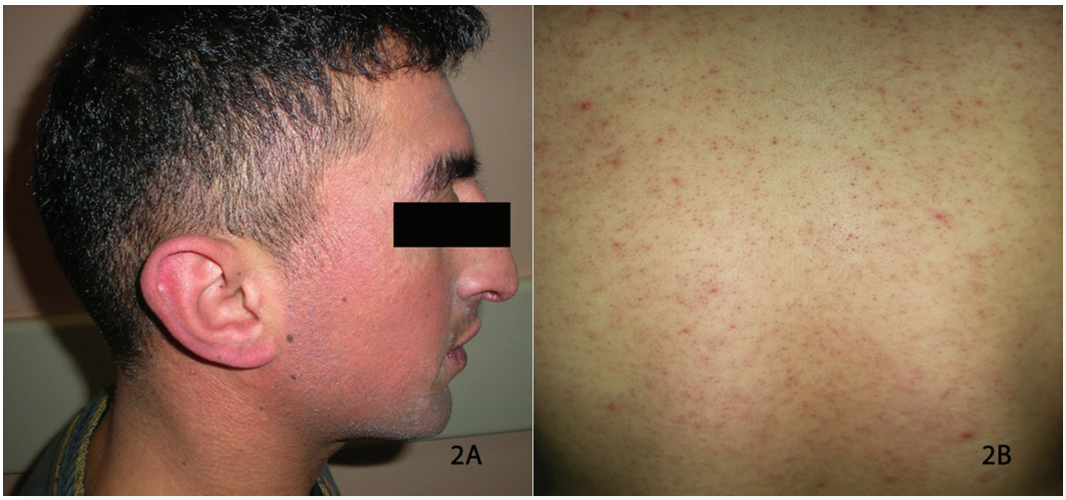

Figure 2A. Millimetric follicular papules affecting the lateral eyebrows and face. **B.** Keratosis pilaris on the trunk.

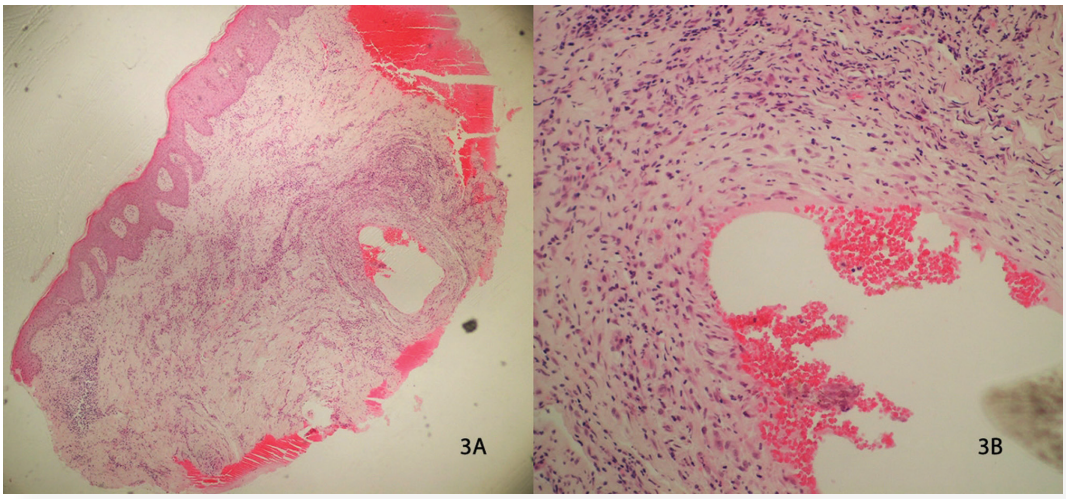

Figure 3A and B. Scalp histology showing the prominent, inflammatory perifollicular lymphocytic infiltrate in the dermis (H&Ex40 and H&Ex200).

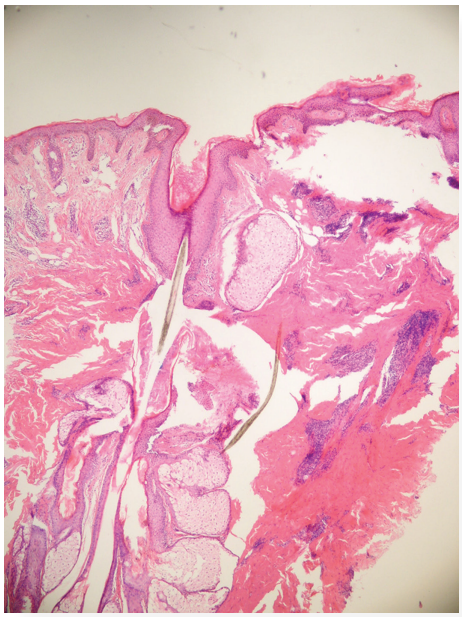

Figure 4. Histopathology of a nuchal papule showing the epidermal acanthosis and the perivascular mixed-type infiltrate along with the neutrophils and fibrosis in the dermis (H&E x40).

Discussion

Keratosis follicularis spinulosa decalvans is considered to be a keratinization disorder, and follicular hyperkeratosis, variable degrees of inflammation, and atrophic scars are characteristic features of this skin condition [4,5]. Follicular hyperkeratosis first appears on the cheeks and face in childhood, and keratosis pilaris lesions occur due to excessive accumulation of keratin in the hair follicles. In addition, progressive cicatricial alopecia may occur on the scalp, eyebrows, and eyelashes [6].

The mechanism of alopecia is not known. At first, it was suggested that the potential mechanism could be keratinization and follicular destruction associated with inflammation in the hair follicles that was caused by hyperkeratosis and hypergranulosis in the infundibulum and isthmus [6,7]. Follicular destruction is caused by hyperkeratosis as well as abnormal cytokine formation stemming from inflammatory reactions. Ophthalmologic findings of the disease include photophobia and corneal dystrophy. The photophobia is thought to be caused by conjunctivitis and hyperkeratotic lesions on the eyelids that occur because of corneal trauma [8]; however, this symptom usually regresses after puberty [7]. However, not all of the published cases include reports of photophobia in connection with alopecia. Palmoplantar hyperkeratosis, clinodactyly, and arachnodactyly should also be considered when examining the hands and feet in KFSD cases. In addition, nail examinations have revealed subungual hyperkeratosis, onycholysis, and cuticular hypertrophy, and gingival hypertrophy and dental agenesis may be seen in the oral mucosa [2,7,9,10]. Other clinical associations may include deafness, wooly hair, aminoaciduria, cutis hyperelastica, and mental retardation [8,9,11].

Acne keloidalis nuchae is a chronic disorder associated with folliculitis that is often seen in the neck area of young adult males. Grouped papules and pustules occur in the first stages of the disease, and in the later stages, keloidal papules, nuchal hypertrophic scars, chronic abscesses, and hair loss can be seen [12,13].

The exact cause of AKN is not yet known, but frequently suggested etiological possibilities are short haircuts along the posterior hairline and penetration of the cut hair into the skin, as in patients with pseudofolliculitis. In addition, constant irritation by shirt collars, a chronic low-grade bacterial infection, seborrhea, and increased testosterone levels are other possible causes [14,15].

In histopathological examinations of KFSD lesions, superficial intra- and perifollicular fluid along with neutrophil infiltration are present during the acute phase. In advanced stages, lymphocytic infiltration is accompanied by follicular destruction as well as concentric perifollicular fibrosis. Some factors should be considered when trying to distinguish between KFSD and AKN lesions histopathologically. For example, KFSD is classified under the lymphocytic group of primary cicatricial alopecias by the North American Hair Research Society, whereas AKN is classified under the group of alopecias in which mixed infiltration takes place. Furthermore, the presence of especially advanced-stage follicular plugs that can cause perforations in the follicular infundibulum, the formation of an abscess in the follicular environment, and hypertrophic scars in the dermis are all important indications of AKN [16].

It is extremely rare to find KFSD associated with AKN, and to date, only two cases have been reported in the literature. One of these cases, reported by Goh et al.,[5] had suffered from cicatricial alopecia and nuchal papular eruptions for ten years. Additionally, acute suppurative folliculitis was identified via a biopsy of the patient's scalp and neck. As in our case, cicatricial alopecia and concurrent nuchal follicular papules had been present for five years. Furthermore, the thinning of the lateral eyebrows in their patient was also accompained by, diffuse follicular papules on the face and body. Other cases pointed to acne keloidalis as well as tufted folliculitis [17]. In our case, follicular tufting was also found in the areas where cicatricial alopecia had developed, and this type of tufting can occur as the result of diseases associated with superficial purulent inflammation that cause cicatricial alopecia of the scalp. We believe this is what took place in our patient since we found fibrous scars caused by cicatricial alopecia and inflammation of the hair follicles.

In conclusion, our case featured a unique association between AKN and KFSD. Lesions related to AKN are an immune response to the presence of a stimulating factor, and we believe that KFSD was the stimulating factor in our patient. In other words, the KFSD increased the predisposition of the AKN. However, it is not known how AKN lesions develop nor is it clear how KFSD caused cicatricial alopecia of the scalp. Therefore, further investigations are needed to explore these topics.

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