Pediatric Dermatology Vol. 23 No. 3 255–258, 2006

Folliculitis Spinulosa Decalvans: An Uncommon Entity within the Keratosis Pilaris Atrophicans Spectrum

Vito Di Lernia, M.D. and Cinzia Ricci, M.D.

Operative Unit of Dermatology, 1st Medical Department, Arcispedale Santa Maria Nuova, Reggio Emilia, Italy

Abstract: Folliculitis spinulosa decalvans is an uncommon condition characterized by follicular hyperkeratosis, followed by scarring alopecia. We report a 12-year-old boy affected by keratotic papules of the scalp and keratosis pilaris of the limbs who developed erythema, pustules, and scale crusts on the scalp associated with scarring alopecia. Histologic examination showed follicular and interfollicular hyperkeratosis, follicular plugging, mild inflammation, and focal scarring. A transient remission of the inflammatory changes on the scalp was obtained after treatment with isotretinoin. The follicular spinulous hyperkeratosis persisted. A severe relapse of the scalp inflammation was observed during a 2-year follow-up.

Folliculitis spinulosa decalvans (FSD) is a term proposed in 1994 by Oranje et al (1) to identify a distinct clinical entity in the group of diseases with keratosis pilaris atrophicans (KPA), characterized by persisting pustular elements associated to typical keratotic papules of the scalp and scarring alopecia, which exacerbates at puberty when inflammation and persisting pustular elements occur on the scalp. Although its nosologic position must be confirmed, it can be considered to belong to the group of the skin diseases characterized by keratosis pilaris and atrophic scarring, together with keratosis follicularis spinulosa decalvans (KFSD), keratosis pilaris atrophicans faciei, and atrophoderma vermiculatum.

This condition has previously been reported as persisting inflammatory variant of KFSD.

We report a new sporadic occurrence of this rare condition.

CASE REPORT

A 12-year-old boy was referred to us for evaluation of a progressive loss of scalp hair. He was the product of a full-term, uncomplicated pregnancy and delivery. His parents were not consanguineous and no history of affected family members was obtained. For 4 years before being seen in our clinic, the patient had been noted to have dry, scaly scalp. Prior treatments included systemic and topical antibiotics and vitamins. When he was 1 year old, he was diagnosed as having atopic dermatitis, which regressed at age 4 years.

Physical examination showed a healthy-appearing young boy. The scalp examination disclosed erythema, scaling, follicular hyperkeratosis, pustules, and crusts (Fig. 1). A patchy scarring alopecia involving the parietal regions of the scalp was also present. He also had extensive redness of the interfollicular skin of the face and horny keratotic papules on the extensor surfaces

Address correspondence to Vito Di Lernia, M.D., Unità Operativa di Dermatologia, Arcispedale Santa Maria Nuova, via Risorgimento 80, 42100 Reggio Emilia, Italy, or e-mail: vito.dilernia@asmn.re.it.

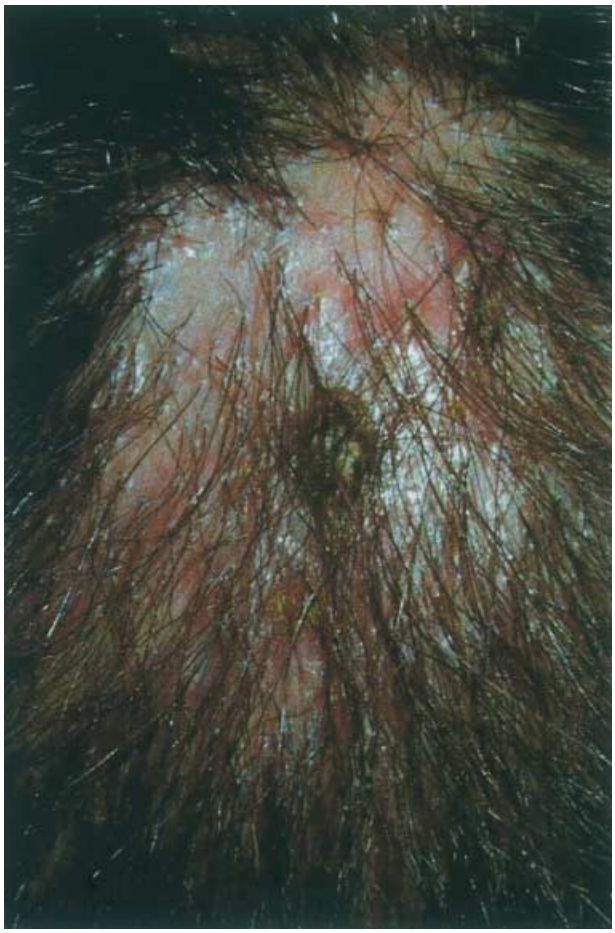

Figure 1. Redness, pustules, crusts, and scarring alopecia on the scalp. Keratotic follicular papules are present at the periphery of the atrophic patch.

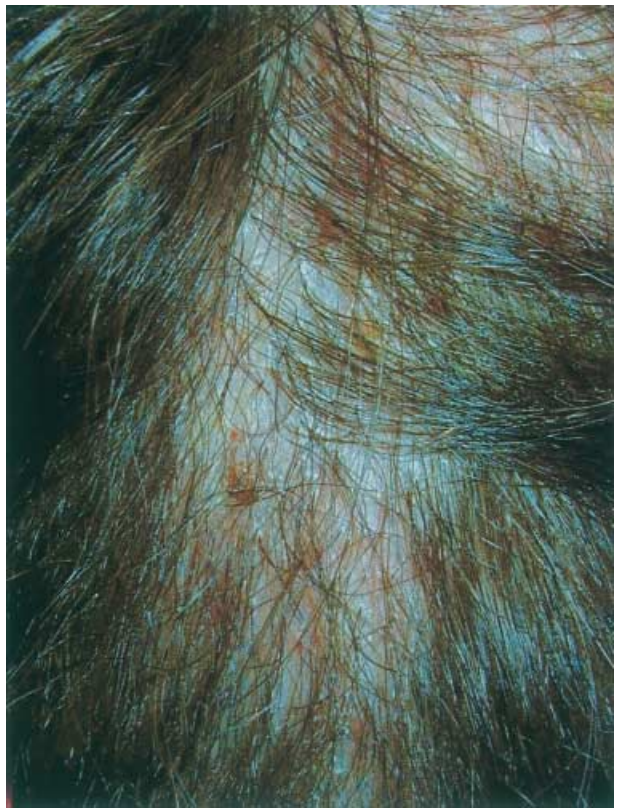

Figure 2. Regression of the inflammation and pustules with residual scale crusts and persistent keratotic papules after treatment with oral isotretinoin.

of the arms and thighs. No loss of eyebrows or eyelashes was noted. The nails, teeth, and mucous membranes were normal. The results of the remainder of the physical examination were normal.

Laboratory studies, including a complete blood count, serum chemistries, and serum amino acid profiles were unremarkable. Although the patient denied ocular symptoms, ophthalmologic consultation found squamous blepharitis and mild inflammation of the conjunctivae. Cultures grew normal skin flora. A biopsy specimen was obtained from the scalp, in particular from the inflamed edge of a plaque and revealed hyperkeratosis and hypergranulosis of the upper follicle with follicular plugging. In the papillary and reticular dermis, there was an inflammatory infiltrate of polymorphonuclear cells, plasma cells, and lymphocytes. A decreased number of terminal follicles was also observed.

Over the ensuing months, the patient experienced recurrent flares of his scalp lesions, as manifested by inflammation, pustules, and discomfort. Keratolytics and topical steroids did not prove useful, nor did a 2-week course of clarithromycin (250 mg two times a day). Isotretinoin, 0.6 mg/kg body weight/day, was then used for 20 weeks. This treatment gave good control of the inflammation and the pustular eruption; the progressive scarring alopecia appeared adequately managed as well, whereas the hyperkeratotic follicular papules on the scalp persisted (Fig. 2). Flares of inflammation and pustular folliculitis continued to occur in the following 2 years and were treated with topical steroids; the application of keratolytics such as salicylic acid and urea resulted in a smoother skin surface, without clearing of the papular eruption, which is still present. After a 2-year follow-up a new, severe flare of inflammation occurred. A cycle of dapsone 50 mg/day did not prove useful and the patient is again being treated with isotretinoin 0.5 mg/kg body weight/day with initially better results.

DISCUSSION

Keratosis follicularis spinulosa decalvans was described in 1905 by Lameris (2) in the Netherlands under the

TABLE 1. *Classification of Keratosis Pilaris Atrophicans, from Oranje et al (1), Modified*

Type/inheritance*	Clinical characteristics	Association/remarks
Keratosis pilaris atophicans faciei/ AD?	Keratosis pilaris atropicans in the eyebrows and keratosis pilaris at other sites	Noonan syndrome, woolly hair
Atrophoderma vermiculatum/AR?	Follicular papules and reticulated atrophy (honeycomb)	Lichen spinulosus, Down syndrome
Keratosis follicularis spinulosa decalvans/x-linked	Follicular papules, absent eyebrows and lashes, scarring alopecia, remission at puberty	Photophobia, corneal dystrophy, nail abnormalities
Folliculitis spinulosa decalvans/ AD?AR?	Follicular papules, persisting inflammation with pustular elements on the scalp, and scarring alopecia exacerbating at puberty	Blepharitis and conjunctivitis (?)

*AD, autosomal dominant; AR, autosomal recessive; ?, uncertain.

name of ichthyosis follicularis. The term KFSD was first used in 1926 by Siemens (3), who described some Dutch patients and others of the original Bavarian family. The skin manifestations usually appear during the first weeks or months of life and are characterized by follicular hyperkeratosis of the skin, especially in the region of the face, associated with scarring alopecia and absence of the follicules of the hair, eyelashes, and eyebrows. Spontaneous improvement at puberty is commonly observed. Eye symptoms can be associated, the most typical of which are photophobia and corneal distrophy. An X-linked inheritance has been confirmed. The clinical picture is more distinct in boys than in girls, who frequently have milder forms. Expression of KFSD in girls can be explained by nonrandom X-inactivation (Lyon hypothesis) (1,4).

Some of the sporadic instances of KFSD reported in the literature do not conform to the classical type (5–7). In 1987 Guillet raised the question of different clinical variants in the spectrum of KFSD, suggesting a subset represented by a sporadic type, clinically severe and characterized by follicular atrophy (8). In 1994 Oranje (1) suggested the name of FSD to define a clinical entity characterized by keratosis pilaris atrophicans, but differing from KFSD by more pronounced erythema and inflammation, occurrence or exacerbation at puberty with eruption of pustules, crusting and scaling on the scalp. The cause of the recurrent folliculitis is still uncertain. *Staphylococcus aureus* had sometimes been isolated from the pustules (6,9); however, in our patient repeated cultures proved negative. Systemic antibiotic therapy has appeared useful episodically (9) to prevent further spreading of the disease, but was ineffective in our patient. However, the true existence of FSD as a distinct disease is still questionable. The gene of KFSD has been located to Xp21.2-p22.2 (4); multipoint analysis placed the gene defect between *DXS16* and *DXS269* (10). Genetic investigations will confirm whether FSD can be added to the group of clinical entities sharing KPA (KFSD, keratosis pilaris atrophicans faciei, and atrophoderma vermiculatum) (Table 1) or represents merely a developmental stage or a distinctive clinical pattern of KFSD.

Our patient had a nonfamilial, papular, hyperkeratotic disorder of scalp hair associated with scarring alopecia occurring in prepubertal age and exacerbating at puberty. Severe inflammation and pustular formation on the scalp were associated with keratotic papules. The clinical and histologic findings of this patient are consistent with those of FSD, the clinical entity identified by Oranje (1). Oral retinoids appeared at first, helpful, with a transient remission of the inflammation and cessation of the spreading scarring alopecia, even though no improvement in the follicular hyperkeratosis was noted and a severe relapse occurred 2 years after the end of treatment. The efficacy of retinoids appears to be controversial in the instances of FSD and KFSD previously reported in the literature (6,7,11–13). This may be in part resulting from the confusion of the two conditions. Resolution of inflammation and pustules has been reported after therapy with dapsone in a 27-year-old man with FSD. This patient had previously been treated with isotretinoin without effect (13). Of interest, blepharitis and conjunctivitis were observed in this man and in our patient.

More than a disease entity, keratosis pilaris (KP) and KPA represent skin symptoms associated with different diseases, mostly of ectodermal origin (1). Thus, in the differential diagnosis it is necessary to consider skin conditions that may have alopecia and follicular keratotic papules, such as Down syndrome, Noonan syndrome, cardiofaciocutaneous syndrome, and monilethrix. A condition closely resembling KFSD is IFAP syndrome, an X-linked disease that shares nonscarring alopecia, follicular hyperkeratosis, and photophobia.

Our unusual patient highlights the difficulties in trying to classify on clinical grounds all patients sharing KPA; in particular, the autonomy of a clinical entity such as FSD with different onset and severity with respect to KFSD remains an unresolved and controversial issue.

REFERENCES

1. Oranje AP, Van Osch LDM, Oosterwijk JC. Keratosis pilaris atrophicans. Arch Dermatol 1994;130:500–502.
2. Lameris HJ. Ichthyosis follicularis. Ned Tijdschr Geneeskd 1905;41:1524.
3. Siemens HW. Keratosis follicularis spinulosa decalvans. Arch Dermatol Syphilol 1926;151:384–387.
4. Van Osch LD, Oranje AP, Keukens FM et al. Keratosis follicularis spinulosa decalvans. J Med Genet 1992;29:36–40.
5. Rand R, Baden HP. Keratosis follicularis spinulosa decalvans. Arch Dermatol 1983;119:22–26.
6. Richard G, Harth W. Keratosis follicularis spinulosa decalvans. Therapy with isotretinoin and etretinate in the inflammatory stage. Hautarzt 1993;44:529–534.
7. Baden HP, Byers HR. Clinical findings, cutaneous pathology, and response to therapy in 21 patients with keratosis pilaris atrophicans. Arch Dermatol 1994;130:469–475.
8. Guillet G, Labouche F, Cambazard F et al. Keratoses folliculaires décalvantes: discussion nosologique de la maladie de Siemens. A propos de trois cas. Pediatrie 1987: 437–440.
9. Romine KA, Rothschild JG, Hansen RC. Cicatritial alopecia and keratosis pilaris. Arch Dermatol 1997;133:381–384.
10. Osterwijk JC, Nelen M, van Zandoort PM et al. Linkage analysis of keratosis follicularis spinulosa decalvans, and regional assignment to human chromosome Xp21.2-p22.2. Am J Hum Genet 1992;50:801–807.
11. Puppin D, Aractingi S, Dubertret L et al. Keratosis follicularis spinulosa decalvans: report of a case with ultrastructural study and unsuccessful trial of retinoids. Dermatology 1992;184:133–136.
12. Herd RM, Benton EC. Keratosis follicularis spinulosa decalvans: report of a new pedigree. Br J Dermatol 1996;134:138–142.
13. Kunte C, Loeser C, Wolff H. Folliculitis spinulosa decalvans: successful therapy with dapsone. J Am Acad Dermatol 1998;39:891–893.