FIBROSING ALOPECIA IN A PATTERN DISTRIBUTION IN TWO BROTHERS WITH PILI MULTIGEMINI

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Summary. Pili multigemini is a rare hair shaft defect. Simply it means the presence of several hairs in a single follicle; each hair has its own inner root sheath and the hairs are surrounded by a common outer root sheath. Fibrosing alopecia in a pattern distribution is a distinct type of cicatricial alopecia which is currently classified as a subtype of lichen planopilaris, characterized by inflammation and fibrosis with accelerated hair loss in the distribution of typical male or female pattern hair loss. We present 25- and 26-year-old brothers with increase hair loss of two years duration. The clinical and histological examination of the scalp area revealed the presence of pili multigemini hair follicles defect combined with fibrosing alopecia in a pattern distribution, involving the vertex area of the scalp in both patients. In our opinion the hair follicle defect pili multigemeni may have an impact over the pathogenesis of the fibrosing alopecia in a pattern distribution in our two cases.

Key words: *pili multigemini, cicatricial alopecia, androgenic alopecia, two brothers*

INTRODUCTION

Pili multigemini hair follicles, also called compound hairs, describe a condition where several separate hair shafts arising from multiple divided matrices and papillae emerging through a single pilosebaceous canal, each hair has its own inner root sheath and the hairs are surrounded by a common outer root sheath [5, 15]. The hair looks like a bunch of flower stalks in a vase. Pili multigemini is listed as a rare disease by the Office of Rare Diseases of the National Institutes of Health.

Fibrosing alopecia in a pattern distribution (FAPD) is a newly described variety of cicatricial alopecia characterized by inflammation and fibrosis with progressive hair loss in the distribution of typical male or female pattern hair loss. Clinically,

FAPD presents with perifollicular erythema, follicular hyperkeratosis and loss of follicular orifices in the midscalp area.

The presence of these both entities in the two brothers raises the question of a cause-and-effect relationship between the two diseases. Pili multigemini is a hair shaft defect not associated with increased hair falling or hair fragility, but it may play a role in the induction of an inflammatory reaction which can lead to the destruction of the abnormal hair follicle resulting in cicatricial alopecia

CASE REPORT

A 26-year-old young man and his 25-year-old brother were presented to our department with progressive hair loss of two years duration, mainly from the vertex area of the scalp, associated with moderate pruritus. Positive family history for alopecia androgenica was reported.

The two patients have been treated with hair growth stimulating products and minoxidil 5% for many months with no effect.

Physical examination revealed androgenetic alopecia type II according to Norwood/Hamilton classification system with increased telogen/anagen hair ratio. In the vertex scalp area there was a decrease in follicular density, perifollicular erythema and perifollicular hyperkeratosis were also observed (Fig. 1 A, B).

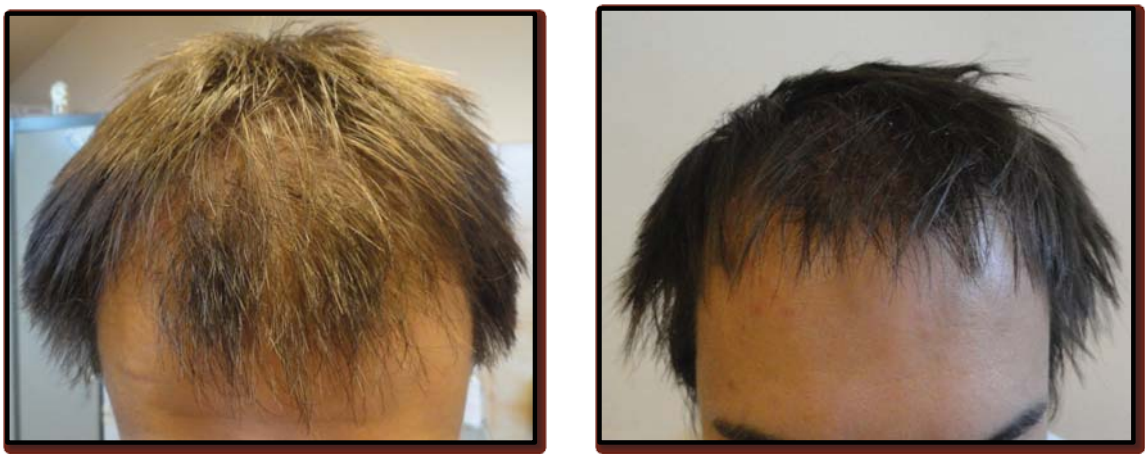

Fig.1. (A, B) Two brothers with progressive hair loss, mainly from the vertex area of the scalp

Dermoscopy of the scalp revealed different hair diameters, many velus hairs. In the vertex area of the scalp we observed follicular hyperkeratosis, dilated capillaries and telangiectasia, skin atrophy and loss of follicular osteum (Fig. 2 A, B).

Dermoscopy from different scalp area of both patients revealed a condition where several separate hair fibers bunch together and emerge from the skin through a single hair canal (Fig. 3).

Histopathological examination of the affected area showed a lymphocytic inflammation targeting the upper portion of the hair follicle (Fig. 4).

Histopathological examination of the pili multigemini bearing area of the scalp revealed several separate hair shafts, each hair had its own inner root sheath and was surrounded by a common outer root sheath (Fig. 5).

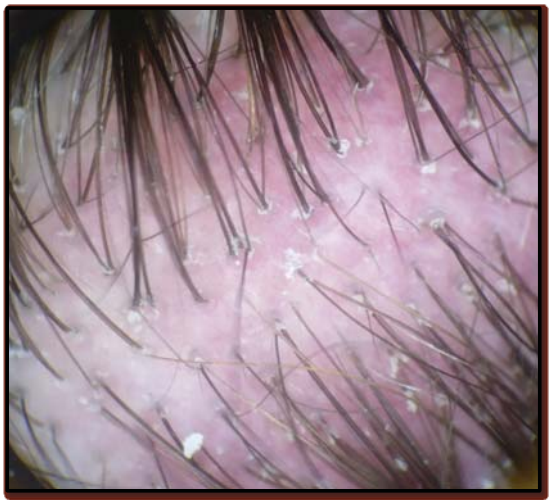

Fig. 2. A First patient's dermoscopy of the vertex area of the scalp revealed pili multigemini, perifollicular hyperkeratosis and erythema, skin atrophy and loss of follicular osteum, different hair diameters and many velus hairs. Dermoscopy x 10

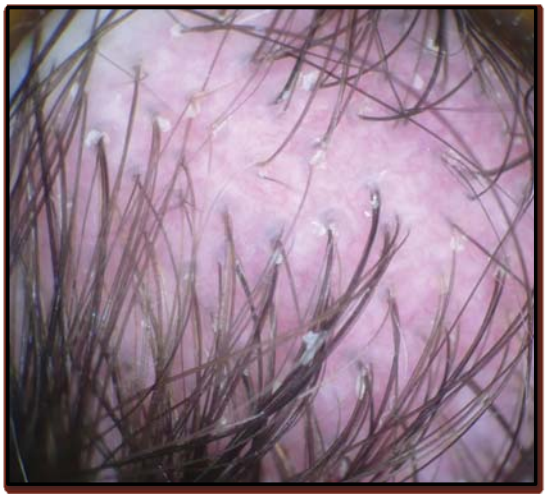

Fig. 2. B Second patient's dermoscopy of the vertex area of the scalp revealed pili multigemini, perifollicular hyperkeratosis and erythema, branching capillaries, velus hair, skin atrophy. Dermoscopy x 10

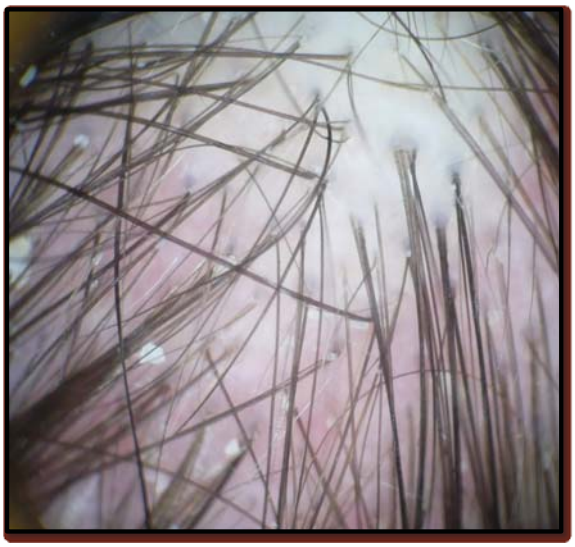

Fig. 3. Dermoscopy from the temporal area of the scalp: pili multigemini. Dermoscopy X 10

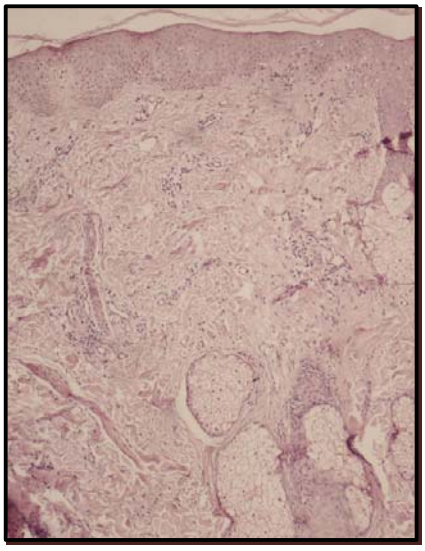

Fig. 4. Histopathological examination of fibrosing alopecia-hyperkeratosis, acanthosis, scattered lympho-histiocytic infiltration intermingled within coarse hyalinized collagen bundles. In the deep reticular dermis a fibrosing tract replacing the normal pilo-sebaceous unit is seen. H. E. stain x 100

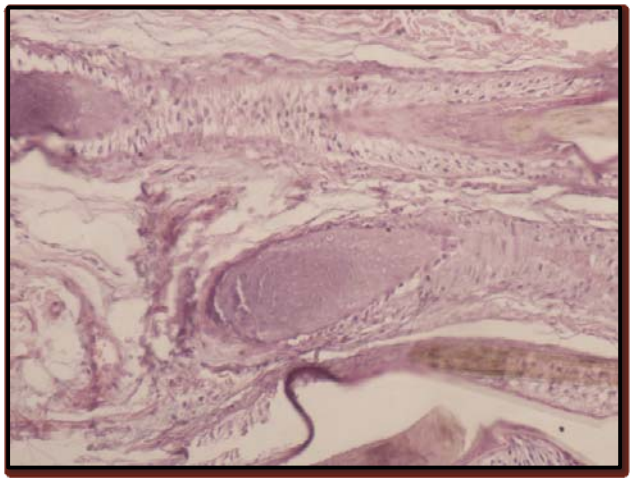

Fig. 5. Histopathological examination – Pili multigemini common bulb. H. E. stain x 400

Test for serum antinuclear antibody was negative.

Total vitamin D level was much lower than the normal limits in both patients – 28.44 and 42.35 nmol/L, respectively, for which supplementation with vitamin D was started.

According to the clinical and histological examination the diagnosis of pili multigemini in combination with fibrosing alopecia in a pattern distribution was performed.

DISCUSSION

Pili multigemini was first described by W. Flemming in 1883 [8], who found a triplet hair in the beard skin. The name was proposed by H. Pinkus in 1951 [13].

Pinkus indicated that two to eight shafts may grow from a single follicular matrix and papilla, the upper end of which is split into tips that correspond one-to-one with each hair shaft.

Multigeminate hairs have been usually found in the beard of an adult and the scalp of children [14]. Involvement of the entire back of healthy males was also reported [9].

The exact origin or mechanism of pili multigemini is still unknown. It was suggested that a subdivided papilla produce divided hairs, or multiple hairs may be due to the partial merging of several papillae, or that the reactivation of silent embryonic epithelial germs result in multigeminate hairs [5, 13].

The regrowth of multigeminate follicles after the forced extraction of the hair shafts from one side of the chin was observed. It was suggested that a fixed follicular malformation with the determining factor might reside within the area of the dermal papillae [11].

In the anagen phase, a kinetic dermal papilla changes its form from single-tipped to double-tipped and produced two hair shafts. The hair shafts emerge

separately through the same pilary canal - pili gemini. The process is very common, and may be observed both in dysplasias and in normal hair. It has been reported as occurring in cleidocranial dysostosis [11] and in the trichorinophalangeal syndrome [2].

Less frequently, the papilla splits into four or eight tips producing four to eight subpapillae which will produce 4 to 8 hair shafts that emerge separately from the same pilary canal - pili multigemini.

Currently, there is confusion between the terms “pili bifurcati” and “pili gemini”. When the one papilla changes into two and the hair shafts fuse again,this is called pili bifurcati [4, 18].

When the same papilla changes its shape repeatedly during the anagen phase, it can produce hair shafts with bifurcations at irregular intervals: pili multibifurcati. And, as it is also possible for one of the new papillae to split again in two, the hair shaft may be doubly bifurcated: pili bi-bifurcati [3, 7].

Another subject that should be differentiated from pili multigemini is the tufted hair folliculitis which is a characteristic localized scarring bacterial folliculitis of the scalp due to Staphylococcus aureus [1, 6]. Histopathological studies reveal perifollicular inflammation around the upper portions of the follicles sparing the hair root level [16]. Within areas of inflammation, several follicles converge toward a common follicular duct with a widely dilated opening. While the pathology of pili multigemini shows that deep in the skin several dermal papillas are closely situated with each producing a fiber, it should be noted that these separate hair follicle bulbs combine together into one hair canal towards the skin surface.

Fibrosing alopecia in a pattern distribution, initially described by M. S. Zinkernagel and R. M. Trueb in 2000, is a distinct type of cicatricial alopecia characterized by inflammation and fibrosis with accelerated hair loss in the distribution of typical male or female pattern hair loss [19].

Females are more commonly affected than males. All the effected men have had preceding male pattern baldness. The histological examination shows a lymphohistiocytic infiltrate around the isthmus and infundibular region of the hair follicle, concentric perifollicular lamellar fibrosis, sebaceous gland loss, and lymphocytic interface dermatitis with destruction of the basal keratinocytes [19]. These histological findings are identical to these seen in lichen planopilaris and, thus, FAPD is currently classified as a subtype of lichen planopilaris [10, 12, 17].

The end result is a reduction in the number of terminal hair follicles by either the underlying miniaturization process of pattern hair loss or by replacement of follicles by fibrous tracts.

Antiandrogen therapy with 1 mg daily of oral finasteride have been shown to calm down the progression of hair loss and decrease the scalp inflammation which

have led to the suggestion that androgens may play a role in the pathogensis of FAPD [19].

Regrettably, our two patients refused to be treated with finasterid because of the possible side effects.

CONCLUSION

In conclusion, we present two brothers with a rare genetic hair shaft abnormality – pili multigemini on the scalp, combined with fibrosing alopecia in a pattern distribution involving the vertex.

Their occurrence in two brothers with increased hair loss raised the question about the role of pili multigemini in the induction of inflammatory reaction against the abnormal hair follicle, producing the cicatricial pattern of alopecia in patients genetically predisposed to alopecia androgenetica.

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