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Pili gemini – a common phenomenon of uncommon presentation and familial background

Broshtilova V, M. Gantcheva***

** Department of Dermatology and Venereology, Faculty of Medicine, Sofia Medical University*
*** Institute of Experimental Morphology, Pathology and Anthropology with Museum, Bulgarian Academy of Science*

Abstract: A 28-year-old man with diffuse alopecia and hair loss is presented. Confronting examination of the other male family members defined the diagnosis of familial pili gemini. Thus, a peculiar genetic item to a dermatological condition that appears with a rather unusual presentation is described. Further observations and genetic studies are needed to define the scientific significance of this phenomenon.

Key words: pili gemini, familial background

Introduction

The first description of multiple hairs is given by Flemming in 1883 [4]. The phenomenon was seen on the beard and for a long time attributed only to this location. Hair shafts dysplasia is admitted to be a consequence of splits of the follicular germen [8]. Currently, confusion on the clinical picture and pathogenesis of the various forms of hair shafts anomalies exist [1].

Herein, a familial case of different-sized hairs with single cuticles, growing from the same follicular matrix and emerging through a single pilary canal is discussed.

Case report

A 28-year-old male sought treatment of diffuse alopecia and hair loss, accentuated in the last few months. He had an elder brother with the same symptoms. Their father had hair loss and baldness since the age of 40. They all complained of permanent dandruff

and greasy hair. On physical examination the hairs in the frontal region of the scalp looked three or four times thicker than average. A magnifying glass revealed closely bundled hairs present in one follicle. They grew as a whole, surrounded by a common sheath. On dermoscopy two hair shafts came out of a single follicular opening (Fig. 1). Confronting visits of the other family members proved the same clinical picture. Histology taken by the scalp of our patient revealed two hair shafts converged towards the base, without fusion, arising from subdivided bulb surrounded with a common sheath (Fig. 2). Transverse sections showed two hairs surrounded by own cuticles, enclosed in the outer root sheath. The diagnosis of presumably autosomal dominant familial pili gemini of the scalp was suggested. Recommendations on appropriate daily care were provided.

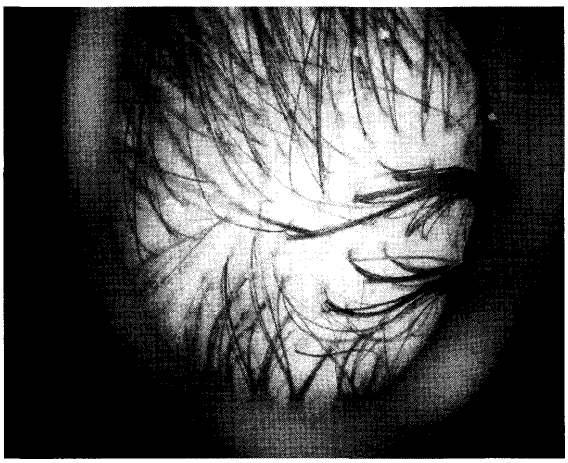

Fig. 1. Dermoscopy findings of two hair shafts coming out from a common follicular opening

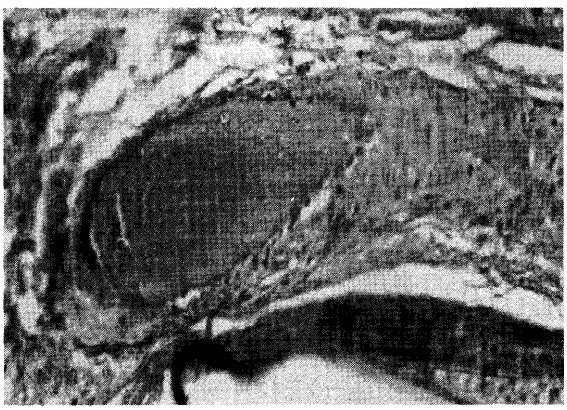

Fig. 2. A subdivided hair bulb with two hair shafts converged towards the base, without fusion, surrounded with a common sheath (HE, × 400)

Discussion

The hair has the most elaborately balanced structure with even minor functional deviations forming various aberrations [10]. The basic rules of hair morphogenesis are often break and on gross inspection of the skin incomplete hairs are present in one follicle or a bundle of hairs protrude from the follicular mouth [9]. Multiplicity of hairs may result either from remnants of old hairs in the follicle as an exaggeration of the normal cyclic activity of the papilla (trichostasis spinolosa) [12] or of splitting of the papilla matrix during the anagen of the hair to form compound follicle[7].

Double-tipped kinetic papilla produces two different-sized hair shafts with separate cuticles emerging through a single pilary canal [8]. This condition is known as pili gemini. The phylogenetic arrangement of hairs in groups of three or five gives the very common phenomenon of compound hairs – several hairs contained in the same follicle[2]. Two main mechanisms have been incriminated to cause the condition. According to Koelliker [5] accessory follicles grow down from the neck of the central one, while Rabl [11] favored the view of secondary merging of the superficial parts of independent follicles. This second opinion is widely recognized as the main reason for appearing of tufted hair folliculitis, a condition that is probably a consequence of staphylococcal scalp infection[3]. The most important differentiation of pili gemini and compound hairs is the level of hair follicle units merging [1]. The superficial merging comes out with tufted hair folliculitis, while matrix papilla tip merge forms two separate hair shafts with own cuticles, defining the pili gemini phenomenon.

Contra version exists as for the epidemiology and pathogenesis of hair shaft anomalies. Pili gemini can be observed both in hair dysplastic conditions such as cleidocranial dysostosis [7] and trichorinophalangeal syndrome, and in normal hair. According to Pincus, who first described the condition in 1951[9], the occurrence of the anomaly is really uncommon. The author pointed out that minor aberrations which furnish the basis for actual subdivision of the hair shaft usually affect the beard region and most often split the tip of the papilla. Thus, a complete subdivision of the hair shaft can result only if the split extends down to the neck of the papilla, at the site of inner rooth sheath formation. Therefore, the labiality of the hair matrix seems to play the most important role in the pathogenesis of multiple hairs. If the papilla tip split maintains during the whole anagen phase, hair shafts remain separate and form pili Gemini [13]. When the same papilla changes its shape repeatedly, it can produce hair shafts with bifurcations at irregular intervals defining the pili bifurcati. There is then no absolute distinction between the completely divided multiple hairs and the partly merged composite hairs [3]. Therefore, pili bifurcati can be considered a special case of pili gemini.

We presented a family with pili gemini that affect the scalp region. The location is not often described, which can be due to either low incidence or overlooking of the phenomenon. The signs of seborrheic dermatitis are probably secondary to male-pattern hair loss, seen in our patients. Of great interest is the familial background, which seems to be of autosomal dominant trait. To date, there were no other observations and genetic investigations in this area.

This anecdotal case report is presented to revive further knowledge of a well-known hair shaft anomaly presented at an unusual genetic background and atypical localization. Thus, we dare add a peculiar genetic item to a dermatological condition that appears with a rather unusual presentation. Further observations and genetic studies are needed to define the scientific significance of the phenomenon described.

References

1. Camacho, F., R. Happle, A. Tosti, D. Whiting. The different faces of pilli bifurcate. – Eur. J. Dermatol., 2000, 10: 337-340.
2. Cambiaghi, S., M. Barbareschi, G. Cambiaghi et al. Scanning electron microscopy in the diagnosis of pili multigemini. – Acta Derm. Venereol., 1995, 75: 170-171.
3. Elston, D. Tufted folliculitis. – J. Cutan Pathol., 2011, 38: 595-596.
4. Flemming, W. Ein Drillingshaar mit gemeinsamer innerer Wurzelscheide. Monatshefte prakt. – Dermatol., 1883, 2: 163-167.
5. Koelliker, A. Handbuch der Gewebelehre des Menschen. 6th Ed. Leipzig, Wilhelm Engelmann 1889, 1: 221-245.
6. Lee, J., Y. Kim, H. Kang. The nevoid pili multigemini over the back. – Eur. J. Dermatol., 2005, 15: 99-101.
7. Mehregan, A. H., W. S. Thompson. Pili multigemini. Report of a case in association with cleidocranial dysostosis. – Br. J. Dermatol., 1979, 100: 315-322.
8. Pinkus, F. The story of a hair root. – J. Invest. Dermatol., 1947, 9: 91–93.
9. Pincus, H. Multiple hairs (Flemming-Giovannini). Report of two cases of pilli multigemini and discussion of some other anomalities of the pilary complex. – J. Invest. Dermatol., 1951, 17: 291-301.
10. Price, V. H. Structural anomalies of the hair shaft. Pili multigemini. – In: Orfanos C. E., Happle R., eds. Hair and hair disease. Heiderlberg: Springer-Verlag, 1990: 414-415.
11. Rabl, H. Histologie der normalen Haut, in Handbuch der Hautkrankheiten, ed. Franz Mracek. Wien, Alfred Hoelder, 1902, 1: 50.
12. Strobos, M. A., M. F. Jonkman. Trichostasis spinulosa: itchy follicular papules in young adults. – Int. J. Dermatol., 2002, 41: 643-646.
13. Whiting, D. A. Hair shaft defects. Pili multigemini and pili bifurcati. – In: Olsen E. A., ed. Disorders of hair growth. Diagnosis and treatment. New York: McGraw-Hill, 1994: 118.