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Case report-Olgu sunumu

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Monilethrix

Moniletriks

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Abstract

Moniletriks is hair shaft defect seen as mono symptomatic or with some ectodermal anomalies. Patients are usually normal at birth, symptoms emerge after a few months. There is not an effective treatment. Here a five-year-old moniletriks case is reported, having hair, eyebrows and eyelashes growing defect.

Keywords: Monilethrix, hair shaft, hair disease

Özet

Moniletriks monosemptomatik olarak ya da bazı ektodermal anomalilerle birlikte görülen kıl şaftı defektidir. Hastalar çoğu zaman doğumda normaldir ve doğumdan bir kaç ay sonra belirtiler ortaya çıkar. Etkili bir tedavisi bulunmamaktadır. Burada saçları, kaşları, kirpikleri büyümeyen ve kollarında keratozis pilarisi olan beş yaşındaki moniletriks olgusu sunulmuştur

Anahtar sözcükler: Moniletriks, kıl şaftı, saç hastalığı

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Introduction

Monilethrix is a rare autosomal dominant dysplastic disease of the hair shaft and was described in 1879 for the first time it usually occurs in the first months of life, but sometimes can be seen in childhood or in even later ages. Hair is of normal appearance at birth but this hair is lost. After 2-3 months dry, brittle, short, no more than 2-3 centimeters long hair takes place. Hair is fragile and characterized by follicular papule [1-3]. Here, we present a case of monilethrix with review of the literature.

Case report

A five-year-old female patient who applied to our outpatient clinic with complaints of non-elongation in hair, eyelashes and eyebrows. Investigation of clinical background revealed that complaints had begun approximately at when she was 4 months old. In dermatological examination, thinning, shortening and drying of hair shaft with a diffuse thinning of the scalp was seen. Family history of the patient was unremarkable. In addition, 0.3 to 0.5 mm in diameter skin-colored papules with follicular localization on the scalp, eyebrows and eyelashes regions and pilar keratosis on arms was seen in dermatological examination. The patient's nails, oral mucosa, and eye examination revealed no pathological findings (Figures 1 and 2). In the laboratory investigation, complete blood count, biochemistry analysis, thyroid function tests and urinalysis were normal. Microscopic examination of hair showed symmetrical and identical in size

elliptical nodes in the hairs haft that were separated by narrow internodes. Based on these findings, the patient was diagnosed as moniletriks.

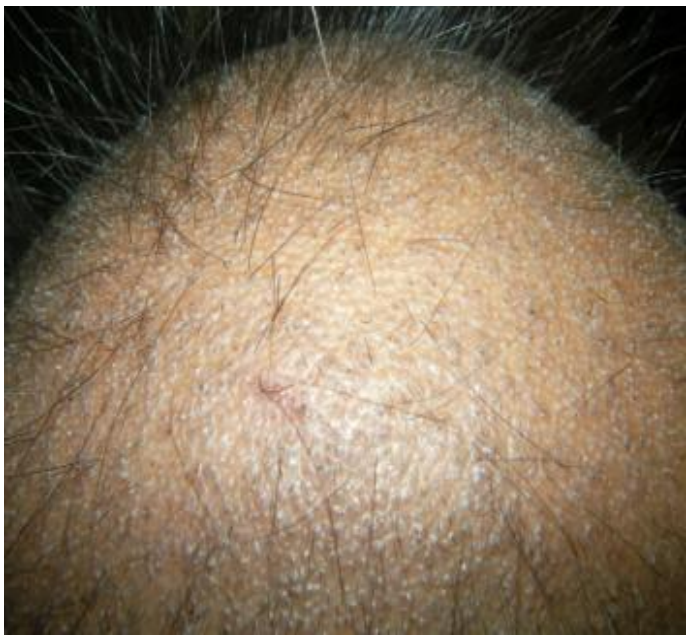

Figure 1. Papules with follicular localization on the scalp.

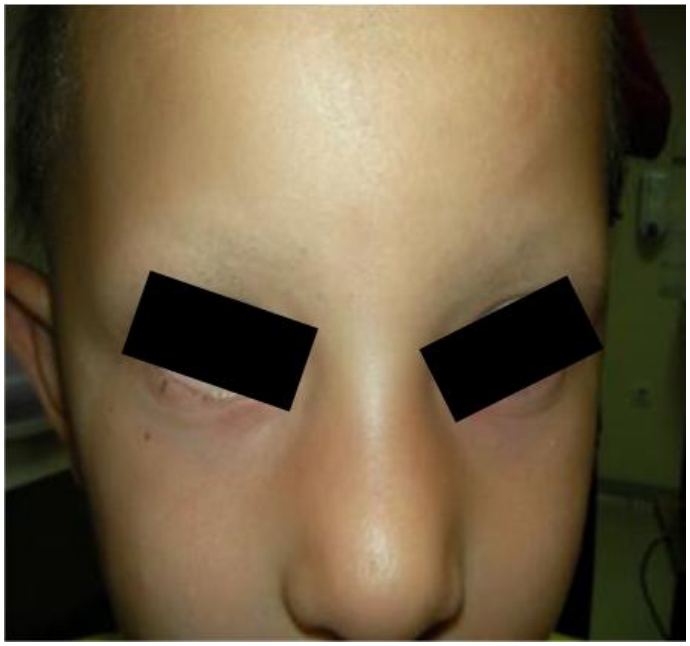

Figure 2. Sparse eyebrows and eyelashes, papules with follicular localization.

Discussion

Monilethrix is often autosomal dominant inherited but autosomal recessive inheritance of this disease is also reported and is characterized by the hair shaft disorder. Sporadic cases due to de novo mutations can be seen without a family history [1, 4]. In our patient, there was no family history. Mutations in the human hair keratin genes (hHb1 and hHb6) may be responsible for the pathogenesis of monilethrix. As a result of these mutations, hair breakage at different degrees, hair loss and follicular hyperkeratosis can be seen. It is

estimated that mutations mainly occurs in germinative cells of hair cortex, this defect causes impaired cycle and deterioration of diurnal rhythm which cause shrinking and expanding sections of the hair shaft. Growing, structure and number of cortical cells in large segments of the hair is normal. Thinner segmented portion of the hair is more transparent, contoured and has a smaller number of cortical cells. There is no medulla in histological examination of this section [1, 4-7]. In monilethrix, there are lanuga hairs in the neonatal period which has normal appearance. Asymptomatic changing of the hair after the birth starts within 6 weeks-1 year [2, 3]. In our patient, the complaints began at the four months after the birth. Often occipital region, nape or even the entire hair is affected in monilethrix. Eyebrows, eyelashes, axillary and pubic hair of patients may be affected. Follicular papules on neck and nape can be seen. Hair is fragile and rarely longer than 1-2 cm. Keratosis pilaris and koilonychia, rarely mental retardation, growth retardation, syndactyly, cataracts, tooth, nail anomalies may accompany the disease [8, 9]. In our patient, all the hair, eyebrows and eyelashes were involved and had keratosis pilaris on the arms. Trichogram and light microscop plays an important role in the diagnosis of monilethrix. Significantly increased anagen hair is seen in tricology examination. Nodal structures is in the form of beads and intermodal structures that appear lighter and 0.7 to 1 mm in thickness between the nodal structures were seen in examination of the hair shaft [10, 11]. The prognosis of monilethrix is variable. Although in many cases disease is persistent, in some cases it may regress in adolescence. In some cases reduction in disease during the summer months, pregnancy with age has been reported. In addition, reduction in disease in a case after the first menstrual cycles and after iron treatment in case with iron deficiency anemia has been reported in the literature [7, 12, 13]. There is no effective treatment for monilethrix. There are cases who were treated with systemic corticosteroids, oral retinoids, vitamins, peeling, and topical agents such as minoxidil in the literature. In addition, avoiding from hardly crawling and brushing of hair is very important for the protection of existing hair [12-17].

In light of this literature it is understood that monilethrix is disease with childhood-onset and genetic and hormonal factors are accused in the etiology, yet there is no an effective treatment protocol and prognosis may differ between patients. Here we present a rare case who has monilethrix with only cutaneous involvement to contribute to the literature.

References

1. Bentley-Philips B. Monilethrix and pseudo-monilethrix. In: Orfanos CE, Happle R, eds. Hair and hair diseases. Berlin: Springer-Verlag 1990; 423-41.
2. Sotiriou E, Sotiriadis D. Monilethrix. Case report. Eur J PediatDermatol 2002: 12; 101-4.
3. Singh G, Siddalingappa K, Nischal KC, Naik LC, Lokanatha K. Monilethrix in three generations. Indian J Dermatol 2008; 53: 99-100.
4. Horev L, Djabali K, Green J, Sinclair R, Martinez-Mir A, Ingber A, Christiano AM, Zlotogorski A. De novo mutations in monilethrix. Exp Dermatol 2003; 12: 882-5.
5. Baker H. An investigation of monilethrix. Br J Dermatol 1962; 74: 24-30.
6. Dawber R, Neste DV. In: Hair and Scalp Disorders. Martin Dunitz Ltd, Gillingham, UK, 1995; 54-60.
7. Gebhardt M, Fischer T, Claussen U, Wollina U, Elsner P. Monilethrix--improvement by hormonal influences? Pediatric Dermatology 1999; 16: 297-300.
8. Koçak M, Karakayalı G, Pençe B, Kundakçı N. Moniletriks (Case Report). T Klin Dermatoloji 1993; 3: 15-7.
9. Berker DAR, Messenger AG, Sinclair RD. Disorders of Hair. Rook's Textbook of Dermatology. Burns T. Eighth Edition. Blackwell Science 2010; 66.1-66.100.
10. Zlotogorski A, Christiano AM. Mapping of monilethrix and papularatrichia [letter]. Dermatology 1995; 190: 89-92.
11. Erbağcı Z, Erbağcı I, Erbağcı H, Erkılıç S, Tuncel AA. Severe Monilethrix

Associated with Intractable Scalp Pruritus, Posterior Subcapsular Cataract, Brachiocephaly, and Distinct Facial Features: A New Variant of Monilethrix Syndrome? Pediatric Dermatology 2004; 21: 486-90.
12. Zalka AD, Goldsmith LA, Scott GA, Tunnessen WW Jr. Picture of the month. Monilethrix. Arch PediatrAdolesc Med 1995; 149: 549-50.
13. de Berker D, Dawber RP. Monilethrix treated with oral retinoids. Clin Exp Dermatol 1991; 16: 226-8.
14. Harper J, Orange A, Prose N. Hair disorders. In: Olsen EA, editor. Text book of pediatric dermatology. Vol 2. London: Blackwell Science 2002; 1463-90.
15. Thai KE, Sinclair RD. Keratosis pilaris and hereditary koilonychia without monilethrix. J Am AcadDermatol 2001; 45: 627-9.
16. Wolff H. Diseases of Hair. Braun-Falco's Dermatology Braun-Falco O, Plewing G, Wolff HH, Landthaler M, editors. Third edition. Springer MedizinVerlag Heidelberg, Italy 2000; 1129-40.
17. Tamayo L. Monilethrix treated with the oral retinoid Ro 10-9359 (Tigason). Clin Exp Dermatol 1983; 8: 393-6.