Pseudomonilethrix type II and pili bifurcati

Editor

We present the case of a 2-year-old boy who was brought to our unit for evaluation of his hair problem. The hair, especially at the occipital and temporal areas of the head, was dry, thin and very fragile, in the context of a generalized hypotrichosis (fig. 1), the length of the hairs was not more than 1.5 cm. The physical and psychological development of the boy was within normal limits. Both parents had normal hair and the personal and family history did not have associated diseases. We could not appreciate keratosis pilaris at the scalp, nape of the neck, or extensor surfaces of the upper arm and thigh. Laboratory test including blood count test, general biochemistry, urinary test, immunoglobulins, thyroid profile and autoantibodies showed no abnormalities. Microscopic examination of hair showed that the hair shaft has spindle-shaped swellings by constricted internodes of normal appearance. In our case pili bifurcati was associated, instead of pili torti (figs 2 and 3). Pseudomonilethrix type II was diagnosed. We started treatment with minoxidil 2% 1 mL/day, and advised the parents to avoid any trauma of the hair with great improvement in 3 months.

Pseudomonilethrix is a rare dysplastic disorder of the hair shaft that is inherited as an autosomal dominant
trait with high penetrance but variable expressivity. It was first described by Bentley-Phillips and Bayles in 1973 and clinically affects the whole scalp as a generalized hypotrichosis or just the occipital area. Pseudomonilethrix usually starts in the first months of life, although, in some cases, it does not become apparent until childhood. No follicular papules can be seen on physical examination.2

Pseudomonilethrix has been classified in three different types: I, familiar pseudomonilethrix of Bentley-Phillips (autosomal dominant inheritance); II, acquired pseudomonilethrix in dysplastic disorders with hair fragility (inheritance profile depending on the dysplastic disorder); and III, iatrogenic pseudomonilethrix.3

Diagnosis can be easily made by microscopic examination of the hair shaft. Elliptical nodes form along the hair shaft at variable intervals and separated by narrower internodes. Differential diagnosis with monilethrix is obligated. Monilethrix is an autosomal dominant disorder characterized by a beaded appearance of the hair due to periodic thinning of the shaft.4 The phenotype results in hair fragility and patchy dystrophic alopecia. Keratosis pilaris is almost invariably associated with monilethrix unlike pseudomonilethrix. Concomitant ectodermal defects have been reported in the literature as dental abnormalities, juvenile cataracts, and some neurologic defects that appear as a cognitive deficiency or epilepsy. Pili torti or partial forms of trichorrhexis nodosa have been associated with the second type of this hair disorder. There are no previous reports of pili bifurcati and pseudomonilethrix in the literature, as the clinical case that we present. The avoidance of trauma is the most effective method of managing this anomaly.5

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References

Adult-onset Langerhans cell histiocytosis confined to the skin

Editor

Langerhans cell histiocytosis (LCH) is a clonal, pleomorphic disease of unknown aetiology, with the accumulation of local or disseminated atypical histiocytic cells staining positively for S-100 and CD-1a, and causing damage in the bones, lungs, mucocutaneous structures and endocrine organs.1 The condition is generally diagnosed in infancy and childhood, but onset in adulthood can occasionally occur. The eruption is usually diffuse or manifests as part of a multisystemic disease, and up to 25–50% of patients with LCH will present initially with a cutaneous rash.1,5 Cutaneous manifestations are heterogeneous and similar both in children and adults.1,2 LCH confined to the skin is uncommon among reported cases of adults.2-8 We describe a rare case of adult-onset LCH presenting as cutaneous lesions located symmetrically on the scalp, groin and inframammarian areas, and review the literature for previous adult cases of isolated cutaneous LCH.

A 38-year-old woman presented to our dermatology department with a-two-year duration of red, painful skin ulcerations in the inframammary and inguinal areas located bilaterally, and severe crusting and scaling of the scalp. Skin examination showed diffuse yellowish crusting with erosions involving the scalp and extending beyond the hairline onto the forehead (fig. 1) and symmetrically distributed sharp-edged, ulcerated plaques with a small

References

Fig. 1 Erythematous, erosive, ulcerated plaques on right inframammarian fold.