Clinical History

The patient was a 2-year-old girl with no past history of interest. She was referred to our outpatient clinic because her parents had reported sparse, very slow-growing hair since birth; her hair had never grown long enough to reach below the neck. The girl's mother had had hair of similar characteristics in childhood, and its appearance had improved with age.

Physical Examination

The hair was blonde, thin, short, and sparse, with a diffuse loss of hair density predominantly in the parietal regions (Figure 1). The hair was not fragile and there were no scalp abnormalities. Pulling on a lock of 10 hairs was completely painless. There were no abnormalities of the nails, eyebrows, eyelashes, eyes, or teeth.

Complementary Tests

Trichography was performed and demonstrated that all the hairs removed lacked epithelial sheaths and presented dystrophic bulbs in anagen phase, with a ruffled cuticle on the part of the hair shaft closest to the bulb (Figures 2 and 3), and no shaft abnormalities.

What Was the Diagnosis?

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Diagnosis

Loose anagen hair syndrome.

Clinical Course and Treatment

The parents were informed that the disorder would improve spontaneously with age and that care should be taken to avoid damaging the hair. Adjuvant treatment was started with a hydroalcoholic solution of minoxidil, 2%, 1 mL per day, and oral supplements of L-cystine, 500 mg per day, and biotin, 5 mg per day.

Discussion

The loose anagen hair syndrome is a type of hair dysplasia first described by Zaun\textsuperscript{1} in 1984. It is due to a lack of adhesion of the hair to the hair follicle, leading to a painless loss of hair on gentle traction,\textsuperscript{2,3} but there is no increase in hair fragility.

The loose anagen hair syndrome has only been reported in white individuals. It occurs mainly in 3-to-6-year-old girls with fine, blonde hair, which is often sparse, though coverage is sufficient to avoid the appearance of conspicuous areas of alopecia.\textsuperscript{4} As hair growth is very slow and stops on reaching a certain length, the girls do not need to have their hair cut, as it usually never grows below the level of the shoulders. The areas of the scalp most severely affected are the vertex and the occipital area.

The loose anagen hair syndrome usually occurs sporadically, although familial cases have been reported.\textsuperscript{5} This syndrome has not be consistently associated with any other disorder, although there are case reports in patients who are also suffering other diseases, such as Noonan syndrome, nail–patella syndrome, trichorhinophalangeal syndromes, and other ectodermal dysplasias.\textsuperscript{3,4}

With regard to the pathogenesis, it has been suggested that alterations exist in the follicular adhesion molecules, such as desmosomal desmoglein or E-cadherin,\textsuperscript{1} leading to weak anchoring and slow growth of the hair.

The diagnosis is fundamentally clinical and is based on the finding of easily and painlessly plucked hair in a patient with a compatible clinical history. However, it should be remembered that this sign varies over time, and that there are periods during which the hair-pull test is negative; in these cases, if the clinical findings are suggestive, the hair-pull test should be repeated at regular intervals or a trichogram performed on hair pulled out by force. The trichogram will provide diagnostic confirmation: it shows most of the hair to be in anagen, absent epithelial sheaths, deformed bulbs that are angled with respect to the hair shaft, and a cuticle that is rolled up on itself or ruffled at the level of the shaft closest to the bulb.\textsuperscript{5,6}

The principal differential diagnoses are diffuse alopecia areata, trichotillomania, hereditary hypotrichosis simplex, and female-pattern androgenetic alopecia.\textsuperscript{3,4}

There is no effective treatment for loose anagen hair syndrome, although the thickness, length, and pigmentation of the hair improve spontaneously with age. All trauma to the hair, such as pulling when combing or vigorous brushing, should be avoided and it would also seem reasonable to add oral supplements of biotin, 5 to 10 mg per day, and L-cystine, 500 mg per day.\textsuperscript{2,6}

Conflicts of Interest

The authors declare no conflicts of interest.

REFERENCES