A girl with loose anagen hair syndrome and concurrent uncombable hair syndrome

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Key words: loose anagen hair syndrome; uncombable hair syndrome.

INTRODUCTION

Loose anagen hair syndrome (LAHS) is characterized by easily extractable anagen hairs that lose the inner and outer root sheath during removal.[1] Patients with uncombable hair syndrome (UHS) have unruly, "spun-glass" hair that cannot be combed flat.[2] In 1996, Boyer et al[3] reported an instance of LAHS mimicking uncombable hair. In 2005, Lee et al[4] reported another instance of LAHS with concurrent features of uncombable hair syndrome. We report another patient with findings suggestive of both syndromes.

Abbreviations used:

LAHS: loose anagen hair syndrome
UHS: uncombable hair syndrome

CASE PRESENTATION

A 10-month-old Middle Eastern girl presented for evaluation of wispy, roughly textured scalp hair. The patient's hair appeared to be growing normally until this rough texture was noted at approximately 6 months of age. During this first visit at 10 months of age, areas of roughly textured scalp hair were noted without any appreciable hair loss.

The patient returned to the clinic at 23 months of age regarding patchy alopecia most evident on the left side of the scalp. At this time, the patient's scalp hair was noted to have the same wispy, straight, roughly textured hair as at the previous visit but with sparing of the central occipital scalp (Figs 1-3). Unlike the majority of her scalp hair, the central occipital scalp had curly, softer hair (Figs 2 and 3). During this second visit, it appeared as though the patchy alopecia was secondary to the patient's pulling out her hair.

Two weeks after the previous evaluation, the patient presented again due to increasing amounts of hair loss. Photos from her mother showed patchy hair loss on the scalp (Fig 4). The patient's mother also collected a bag of shed hairs for evaluation. No abnormalities of the eyelashes, eyebrows, fingernails, or toenails were noted.

Under dermatoscopic exam ination, the patient had normal-appearing scalp hair shafts without nodes or trichorrhexis. Her scalp skin appeared normal, without erythema or flaking. Microscopic examination of hairs showed many anagen hairs with the hair bulbs oriented at an acute angle to the hair shaft and a ruffled cuticle that resembled a crumpled sock (Fig 5). Microscopic examination also showed normal-appearing hair shafts (Figs 6 and 7). No abnormalities were seen pursuant to an examination with polarized filters (Fig 7). Electron microscopy was not available.

The patient was born at 40 weeks, and the mother had an uncomplicated pregnancy and vaginal birth. The child has been meeting regular pediatric milestones. The family history is significant for an aunt who has unruly hair. Her parents have no hair issues other than male pattern baldness in the father. The patient has 2 brothers without hair issues.

DISCUSSION

One of the first recorded descriptions of loose anagen hair syndrome was by Zaun in 1987.[5] Patients with LAHS often have thin hair, described as having a tacky or sticky feel, that can be easily extracted from the scalp with traction.[6] Girls between the ages of 2 and 9 years are most commonly diagnosed with the

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Funding sources: None.
Conflicts of interest: None disclosed.

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JAAD Case Reports 2020;6:92-5.
2352-5126

https://doi.org/10.1016/j.jdcr.2019.07.033

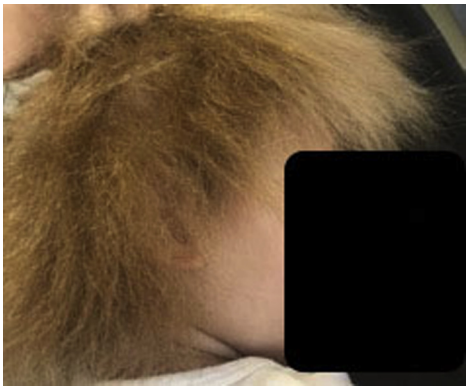

Fig 1. A picture taken in the office at 23 months of age showing unruly, kinked, light-colored hair.

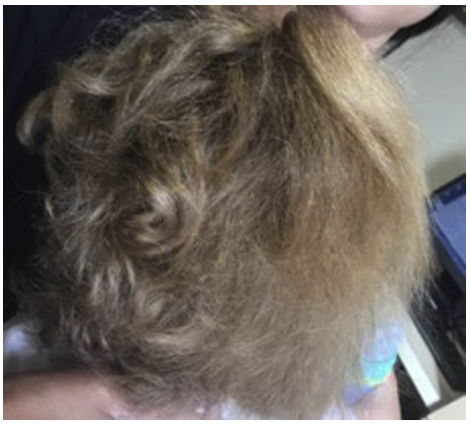

Fig 2. A picture taken in office at 23 months of age showing different hair texture in particular areas of the scalp.

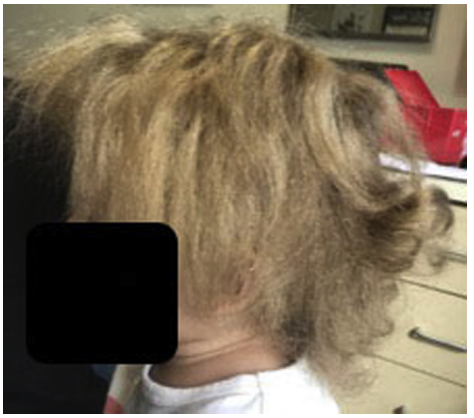

Fig 3. A picture taken in the office at 23 months of age showing a wool-spun texture over most of the scalp and a different, curly hair texture originating in the occipital scalp.

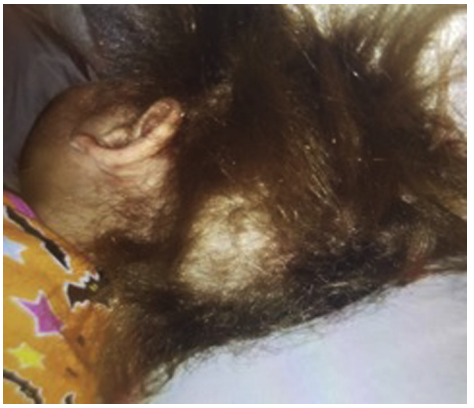

Fig 4. A picture taken by the patient's mother showing an area of hair loss over the occipital scalp. According to her mother, the patient had been pulling out her hair in this area.

condition, although it has been proposed that LAHS may be underreported in male patients who are more likely to cut their hair short.[7,8] Affected patients most commonly have light hair.[1] Both autosomal dominant familial and sporadic forms of syndrome inheritance have been described.[9] Its incidence has been estimated to be 2 to 2.25 occurrences per million per year.[6] Under light microscopy, hairs that are easily pulled from the scalp show ruffling of the hair cuticle, separation of the outer root sheath from the vitreous layer, and grooving of the shaft.[9,10] Scanning electron microscopy of hair specimens may show ridges that run longitudinally along the shaft, twisting of the hair along its longitudinal axis, triangular or other abnormally shaped cross sections, and a distorted bulb.[1,8,9,11] Recently, a genetic pathway related to loose anagen hair has been described for Noonan syndrome with loose anagen hair and another rasopathy involving the SHOC2 and PPP1CB proteins, respectively.[12] In the patient discussed in this case report, the many anagen hairs and clinically apparent hair loss found using light microscopy suggest LAHS. However, the unruly appearance of her hair and the extensive grooving are suggestive of UHS.

In 1973, Dupre et al[13] described *cheveux incoiffables,* meaning "unmanageable hair," and Stround and Mehregan[14] described a spun-glass hair syndrome. This later became known as *uncombable hair syndrome*. Patients have hair that sticks out from the scalp and cannot lie flat. They typically have a light hair color that kinks and twists. This abnormal hair texture is typically noticed before adolescence. This phenotype is often an isolated finding, but it can be found in association with other ectodermal dysplasias manifesting with abnormal teeth, eyes, nails, and sweat glands.[2,15] Another recent case report details a patient with UHS and congenital anonychia found to have mutations in both the RSPO4 and PADI3 proteins.[16] The differential

Fig 5. Easily plucked hair showing anagen bulb and rumpled sock appearance of the shaft.

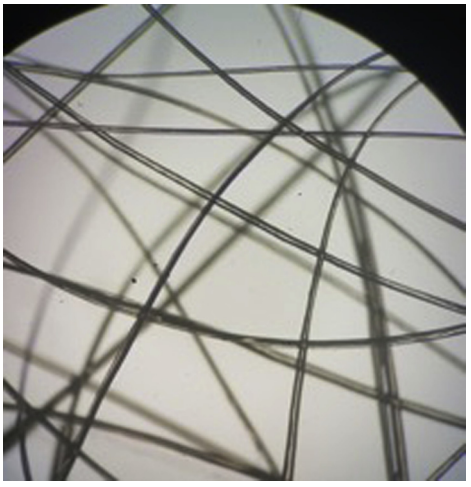

Fig 6. Hair sample under simple microscopic examination

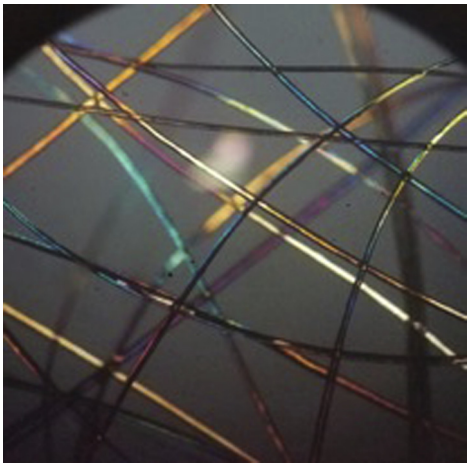

Fig 7. Hair sample under a polarized filter.

diagnosis includes pili torti, monilethrix, wooly hair nevus, and loose anagen hair syndrome. The diagnosis of UHS can be distinguished from these syndromes by clinical and microscopic examination.[2,17] Similar to LAHS, UHS has both inherited and sporadic forms described in the literature. However, unlike LAHS, scalp hair in UHS is not typically fragile, and the syndrome is not associated with hair loss.[2]

It is unusual for both of these syndromes to present in the same patient, although there are 2 other reports in the literature.[3,4] Both of the 2 previous cases and this case describe a young girl with clinical and microscopic features of LAHS and UHS simultaneously.

There are many similarities between this case and the 2 other cases of LAHS with concurrent UHS reported in the literature, although there are some notable differences. In contrast to Lee et al,[4] examination of this patient's hair samples with polarized filters did not show light and dark horizontal banding alternating but, rather, symmetric and normally appearing shafts. Lee et al note that the banding pattern they described is not characteristic of either LAHS or UHS.

Although the incidence of patients meeting the criteria for both of the mentioned conditions is rare, it may be underdiagnosed or underreported in the literature. It is important to consider rare entities and concurrent overlap of syndromes when caring for patients. If criteria for 1 syndrome are met, other diagnostic clues should not be ignored because that may reveal a more complex case. It is possible that there is a link between LAHS and UHS, either genetic or through a shared downstream pathway, but further study and input from the literature are necessary. Increased awareness of patients with concurrent syndromes such as this will allow more providers to recognize and report similar situations, eventually leading to an insight into the mechanism of these diseases.

CONCLUSION

The presented case represents an unusual condition characterized by features of both LAHS and UHS. The patient's scalp has patchy hair loss and easily extracted anagen hairs seen under light microscopy. This is characteristic of LAHS. However, the patient also has an unruly texture to her hair, characteristic of UHS. It is important to consider multiple syndromes and rare entities when evaluating a patient.

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