Adult onset loose anagen hair syndrome with alopecia totalis

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Abstract: A young adult male patient presented with diffuse hair loss of two years duration. Clinical features and investigations were suggestive of loose anagen hair syndrome. He developed alopecia totalis during follow-up. Scalp biopsy revealed perifollicular lymphocytic infiltration. A diagnosis of adult-onset loose anagen hair syndrome with alopecia areata was made.

Introduction

Loose anagen hair syndrome (LAHS) is a rare disorder seen among children [1]. The condition is self-limiting and improves with age. Occurrence of LAHS in adults is rare and most of these cases are diagnosed retrospectively, while screening the parents of the children suffering from LAHS [2]. In such cases, the process usually starts at a younger age and continues till adult life. Very few cases of adult onset-LAHS have been reported in the literature. In the series of patients with LAHS reported by Tosti et al [2], only three patients developed the condition in adulthood.

Case history

A 22-year-old male office worker presented with diffuse loss of scalp-hair for the past two years. It started insidiously as shedding of hairs in tufts while combing and on gentle pulling. In addition, his hair became thinner, shorter, sticky and unruly compared to earlier. One year prior to consultation, he had an episode of sudden total loss of scalp, facial and body hairs. He was diagnosed as a case of alopecia universalis and was advised oral mini pulse therapy with betamethasone (5mg on two consecutive days / week). The hair had gradually re-grown in the affected sites over a period of 3 months.He had normal hair at birth and till 20 years of age as evidenced by earlier personal photographs. There was no history of any major illness or prolonged medication in the recent past. Neither the patient nor any of the family members was atopic. No other family member was affected with similar hair disorder. On examination, the scalp was oily with sticky, black, sparse hairs (Figure:1).

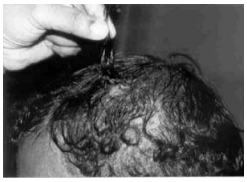

Fig.1 : Sparse, unruly scalp hair

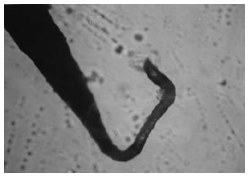

Fig. 2 : Mouse-tail configuration of the hair bulb.

There was no patchy area of alopecia, scarring, atrophy, graying or exclamation mark hairs. A gentle pull to a lock of hair caused painless extraction of the whole tuft comprising more than 40 hairs. Eyebrows, eyelashes, moustache and beard hairs were sparse. Body hairs were normal in density. Other areas of skin, nail, teeth and mucosa were normal. Microscopy of the extracted scalp hairs revealed anagen hairs with fraying of cuticles proximally, ruffling of the inner root sheath and mouse-tail configuration (Figure: 2). A hair-pluck trichogram revealed 95% of the hairs in anagen phase. Routine hemogram, urinalysis and biochemical parameters were normal. ELISA for human immunodeficiency virus was negative. The patient was diagnosed as a case of loose anagen hair syndrome, and was counseled regarding the course and prognosis of the disease.

At follow-up after 3 months, the patient presented with total loss of scalp hairs for the past few days.A punch biopsy was taken from the fronto-parietal region of the scalp. Histopathological examination showed lymphocytic infiltrate surrounding the hair follicles. Fragmentation of the inner root sheath was observed in most of the follicles. The earlier diagnosis was modified. He was diagnosed as a case of loose anagen hair syndrome in association with alopecia areata. The patient was advised oral mini pulse therapy with betamethasone (5 mg on two consecutive days/week).He was lost for further follow-up.

Discussion

The clinical hallmark for the diagnosis of LAHS is easy, painless extraction of hairs by a gentle hair-pull test [1]. The extracted hairs consist predominantly of anagen hairs lacking both outer and inner root sheaths [3]. Microscopically, acute angulation of the distorted hair bulbs to the shafts (mouse-tail configuration) is a characteristic feature of LAHS [3]. The onset is in childhood and there is gradual improvement till adolescence and adulthood. The commonly affected area is scalp but eyebrows and other body parts can also be involved [4]. Clinical features are variable, with fluctuation in the hair-count lost over a given time [1]. Histopathologically, all hair follicles are not involved at one time. Empty follicles, rhomboid or triangular outlines of the hair-shafts [5] and fragmentation of the inner root sheath [2,3] are observed. However, evidence of perifollicular inflammation is never present in LAHS. As there is no characteristic histopathological finding, scalp biopsy is rarely advised[1].Other hair-disorders described in association with LAHS are trichotillomania [1], diffuse partial wooly hair [6] and alopecia areata [2,3]. Tosti et al [2] have reported one child with LAHS in association with alopecia areata. Their patient had patchy loss of hair and developed alopecia totalis during follow-up. Initially our patient had diffuse hair loss. Subsequently he developed one episode of complete hair loss from all body parts which responded to weekly pulse therapy with betamethasone. The second episode of total loss of scalp hair developed during his follow-up with us and a skin biopsy from scalp showed inflammatory infiltrate around the hair follicles, which is a characteristic feature of alopecia areata. Easily pluckable anagen hair is not pathognomonic of LAHS. It can be found in the pull-test and trichogram of 61% of healthy children [7] and some adults [8]. Acquired loose anagen hair is known to occur in patients with AIDS [9]. Pull test may also be

positive at the margins of a patch of alopecia areata indicating very active disease [10]. However, majority of the shedded hairs are telogen or some are dystrophic anagen in alopecia areata. In contrast, more than 90% of the extracted hairs are in anagen phase in LAHS.The initial phase of diffuse hair loss in our patient may well be confused clinically as the rare diffuse form of alopecia areata. However, presence of more than 95% anagen hairs in hair-pull test in association with light microscopic features of frayed cuticles, ruffling of the inner root sheath and mouse-tail configuration confirmed the diagnosis of LAHS. Subsequent histopathological picture of perifollicular lymphocytic infiltrate indicated the development of alopecia areata. Nunez et al [3] have reported a child with alopecia areata who presented initially with clinical and microscopic features suggestive of LAHS. Whether the initial presentation of that child and the index patient with loose anagen hair can be considered as a premonitory sign of alopecia areata remains speculative. The authors have recommended routine scalp biopsy in all patients with an initial diagnosis of LAHS to rule out alopecia areata [3].A combination of adult onset LAHS and alopecia areata in this patient is interesting. Tosti et al [2] and Nunez et al [3] have reported high incidence of inner root sheath fragmentation in their patients with LAHS, which was considered as an artifact due to processing of the specimen. A similar histopathological feature was observed in our patient also. Whether this is a non-significant finding or a histopathological finding of LAHS is to be evaluated.

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