Central Trichoptilosis Associated with Trichorrhexis Nodosa and Pili Torti

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A 12-year-old girl presented with a 1-week duration of hair loss associated with splitting of the hair ends and whitish dots on the occipital hairs. On microscopic examination, a longitudinal splitting of the hair shaft with reconstitution of the normal hair distal to the fracture, nodular swellings, with the appearance of broomsticks pushed into one another, at the site of whitish swellings, and the flattening and twisting of the hair shaft around the long axis were demonstrated.

Minor trauma to injury-prone hair is a common cause of hair shaft defects, however the reports with the combined conditions are insufficient in the literature. We describe a patient with central trichoptilosis associated with localized trichorrhexis nodosa and pili torti. (Ann Dermatol 16(2) 57–60, 2004)

Key Words: Central trichoptilosis, Trichorrhexis nodosa, Pili torti, Hair shaft defect

INTRODUCTION

Hair shaft defects are common and often result in varying degrees of hair loss. It can be inherited or acquired and can sometimes be useful indicators of an underlying disease.1,2 Minor trauma to injury-prone hair is a common cause of hair shaft defects and the clinical features are usually subtle. The classification of hair shaft defects includes fractures of the hair shaft such as trichorrhexis nodosa, trichoclasis, trichoschisis, trichorrhexis invaginata, trichoptilosis; hair shaft coiling and twisting such as pili torti, trichonodosis, circle hair; irregularities of the hair shaft such as longitudinal ridging and grooving, pili bifurcati, pili annulati, monilethrix.

Central trichoptilosis, a longitudinal split in the hair shaft without involvement of the free end of the hair, sometimes occurs1. However our review of the literature revealed a dearth of information about central trichoptilosis but only two well-documented cases were reported, to our knowledge3,4. Here, we report a 12-year-old girl with central trichoptilosis associated with localized trichorrhexis nodosa and pili torti.

CASE REPORT

A 12-year-old girl presented with a 1-week duration of hair loss. She complained her hair was coarse and easily broken. The patient reported to have carried out tight ponytailting of her hair since a month before and to have done back combing habitually. She denied the use of curling irons, hot combs, or chemical agents for styling, except for a single use of dye coloring treatment a week previously. She washed her hair two to three times a week. There were no associated symptoms, no significant medical history, and no family history of similar complaints.

Clinical examination revealed that the distal ends of hair were split and several hair shafts were broken.
The hair shafts showed to be irregular in diameter. There were minute whitish nodules in some hair shafts (Fig. 1). Those clinical findings were localized to the occipital scalp hair and areas of frank alopecia were not seen. A hair pull test detected two to three hairs falling out.

By light microscopy, a longitudinal splitting of hair shaft with reconstitution of the normal hair distal to the fracture was detected (Fig. 2). Split ends of the hair shaft with curricular loss was also revealed by scanning electron microscopy (SEM) (Fig. 3A). SEM also demonstrated nodular swellings with longitudinal fissuring of the cuticles and the appearance of broomsticks pushed into one another at the site of whitish swellings (Fig. 3B), which were the characteristic findings of trichorrhexis nodosa. Flattening and twisting of the hair shaft around the long axis (Fig. 3C), which were the classical findings of pili torti, and focal different widths of the hair shafts were also revealed (Fig. 3D). Those changes were found at different hair shafts, not in one shaft.

The patient was advised to handle her hair gently and to avoid physical and chemical trauma to the hair, especially excessive combing, traction due to a ponytail, and the use of dye coloring treatment.

**DISCUSSION**

Trichoptilosis, first termed in 18721, is a longitudinal splitting of the distal end of hair shaft, and a common fracture caused by chemical and physical trauma2. Usually, it denotes distal trichoptilosis. Following cuticular loss from weathering, the cortical fibers separate like the frayed end of a rope, resulting in 'split ends'. This can occur in normal hair from excessive damage due to hair styling, trichotillomania, or pruritic dermatoses, and can be associated with any congenitally brittle hair in monilethrix, trichothiodystrophy, pili torti, Netherton's syndrome, etc3. It may be seen with trichorrhexis nodosa4. Once a trichorrhexis node has broken, the proximal element may proceed to split further resulting in trichoptilosis. The patients with trichoptilosis usually complain that their hair is dry and brittle. Pathologically, the distal end of the hair shaft is split longitudinally into two or several divisions, which is often easy to see grossly. Avoidance of further chemical or physical trauma and gentle hair care are required to control this condition.

Not only distal but central trichoptilosis also sometimes occurs1. Central trichoptilosis is a longitudinal split in the shaft without involvement of the free end of the hair. Our review of the literature revealed a dearth of information about central trichoptilosis, however, only two well-documented cases were reported to our knowledge5,4. Burkhart et al reported a case with this condition in 19811, and previous to it, Halloran described a case in which central trichoptilosis was noted in between two nodes of trichorrhexis nodosa3. In differential diagnosis, there is pili bifurcati, the other form of
Fig. 3. A. The end of the hair shaft showed cuticular loss and splits in the cortex (SEM, ×80). B. Longitudinal fissures and fractures in the cortex with loss of cuticular scales (SEM, ×400). C. Flattening and twisting of the hair shaft around the long axis (SEM, ×150). D. Hair shafts with different focal width (SEM, ×70).

split hair. It is different from central trichoptilosis. In the former, each bifurcated segment is circumferentially invested with its own cuticle, which is not seen in the latter. We consider that it is difficult to give a diagnosis of central trichoptilosis clinically. Our described patient, here, showed only local thickening of the hair shaft clinically but light microscopy revealed the finding of central trichoptilosis. Our patient showed both central and distal trichoptilosis, and trichorrhexis nodosa was associated as in the previously reported two cases. Areas of pili torti were also noted.

Trichorrhexis nodosa is the most common defect of the hair shaft and known to be a distinct response to injury. It appears along the hair shaft as node-like swellings associated with the loss of the cuticle, and the affected hairs are fragile. It may affect a normal hair following excessive or repeated trauma, and may also occur after minimal trauma in brittle hair caused by an inherent weakness of the hair shaft.
Electron microscopic examination shows longitudinal fissures and fractures of the cortex with loss of cuticular scale. Localized trichorrhexis nodosa is relatively rare and found in patches of trichotillomania, lichen simplex chronicus, and other pruritic dermatoses, but it occasionally appears spontaneously. Our patient presented with localized trichorrhexis nodosa on the occipital scalp hairs.

Pili torti is a rare hair abnormality in which the hair is flattened and completely rotated through 90 to 360 degrees around the long axis at irregular intervals. The affected hairs are coarse, fragile, dry, and fail to attain normal length. A local inflammatory process which distorts the follicles can induce twisted hairs, however the exact etiology has not yet been revealed. Pili torti is usually congenital but it is sometimes acquired. Congenital pili torti may occur as an isolated condition or may be associated with other abnormalities, such as Bjornstad syndrome, Menkes syndrome, ectodermal dysplasia, Crandall syndrome, pseudonelphthria, etc. In cases where it appears as an isolated congenital defect, an autosomal dominant inheritance is suggested. Pili torti may be replaced by normal hair in later childhood and improvement is common after puberty.

Pili torti can be acquired as in our patient, but the abnormality is localized or patchy rather than diffuse, and trauma to the hair or some kinds of scarring process in the scalp such as scarring alopecia can be the cause of this condition. No effective treatment for pili torti is known, however the hair may grow longer if trauma to the hair is minimized. There were no associated congenital disorders or family history in our patient and the physical trauma to the hair could lead to pili torti. Interestingly, in our case, several affected hairs showed different local widths of the shaft (Fig. 3D).

In summary, our described patient here presented with central and distal trichoptilosis associated with localized trichorrhexis nodosa and pili torti on the occipital scalp hair and with mild traction alopecia. It seems the physical trauma to the hair due to a tight ponytail was the possible cause. We consider this is an interesting case which showed various hair shaft defects at the same time. Actually, more often than not, several different defects of the hair shaft may be presented at one time, in the same patient, as in our case.

REFERENCES