Congenital Temporal Alopecia: Dermoscopic Features of Two Cases

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Abstract: Congenital triangular alopecia (CTA), also known as temporal triangular alopecia (TTA) or Brauer nevus, is a well circumscribed, non-scarring form of alopecia present over the temporal region. CTA was first reported by Sabouraud in 1905. It is characterized by triangular area of hair loss, more often occurs unilaterally. Most of the time it is not congenital, usually manifests after 2 years of age. It may rarely appear in adulthood as well. The cause of CTA remains unknown. It usually occurs sporadically, but paradominant trait in families has been reported. Histopathological examination shows normal number of hair follicles, most of it are vellus or indeterminate. CTA is more often misdiagnosed as alopecia areata and mistreated with steroids leading to development of adverse effects of steroids. CTA involves normal skin, thus topical or intralesional steroids are usually not effective. There is no specific treatment of this condition. This is a case report of 2 cases of CTA with clinical and trichoscopic features.

Key words: Dermoscopy, Congenital triangular alopecia, Brauer’s nevus.

Introduction:
Congenital triangular alopecia (CTA) is a uncommon cause of non-cicatrical alopecia.\textsuperscript{1} The incidence in the general population is estimated to be 0.11%.\textsuperscript{2} It is characterized by a non-scarring, circumscribed alopecia often located unilaterally in the frontotemporal region but rarely involves the temporoparietal or occipital scalp\textsuperscript{3}

Alopecia areata is the main differential diagnosis, especially in atypical cases. Dermoscopy is a noninvasive procedure that helps distinguish temporal triangular alopecia from alopecia areata.\textsuperscript{4}

Case History:
Case I
A 6 year old male child visited the dermatology OPD with complaints of patchy hair loss. The patch of hair loss was present since birth and the patient’s informant did not report any presence of a bulla at birth or a significant change in size or texture of the lesion. Further the lesion was not associated with itching, visible scaling or easy pluckability. There was no history of pulling of hair either subconsciously or otherwise. On the positive side, mother reported the reappearance of few light coloured fine hair over the bald patch since the past six months. There was no history suggestive of systemic involvement.

On examination, we found a well defined, lanceolate shaped patch of hair loss on the left fronto-temporal side measuring about 4*5 cm in size. The surface of the lesion appeared normal and there was no visible scaling. The hair were not easily pluckable and the centre of the lesion showed a few scattered lightly pigmented vellus hair. (Figure 1) Head circumference was within normal limits. There were no associated hypertelorism, high arched palate or low set ears. Palms and soles were normal. Systemic examination was within normal limits while KOH was negative for hyphae. A provisional clinical diagnosis of congenital temporal alopecia was made and the patient subjected to trichoscopic evaluation using videodermoscope (Ultracam TLS; Dermaindia, Tamil Nadu, India)

Trichoscopic finding:
The patch showed loss of hair with numerous vellus hair in the bald patch. The patch was surrounded by normal terminal hair. Several empty follicles were also seen. There were no other features of alopecia areata including exclamation hair, yellow dots and broken hair. (Figure 2)
Case II
A 7 month old male baby was brought to our dermatology OPD with complaints of patchy hair loss over left side of scalp. The patchy hair loss was present since birth and the mother did not report any history of bulla at birth or a significant change in size or texture of the lesion. The lesion was not associated with itching, visible scaling or easy pluckability. There was no history suggestive of systemic involvement.

On examination, we found a well-defined, oval shaped patch of hair loss on the left fronto-temporal side measuring about 3*4 cm in size. The skin overlying the patch was normal, without any visible scaling. The hair were not easily pluckable. There were no other markers of congenital anomalies viz, head circumference was within normal limits, there were no associated hypertelorism, high arched palate or low set ears. Palms and soles were normal. Systemic examination was within normal limits while KOH was negative for hyphae. A provisional clinical diagnosis of congenital temporal alopecia was made and the patient subjected to trichoscopic evaluation using videodermoscope (Ultracam TLS; Dermaindia, Tamil Nadu, India)

Trichoscopic finding:
Trichoscopy of the bald patch showed several vellus hair in the involved area surrounded by terminal hair. Several empty follicles were seen. There were no exclamation marks, yellow dots or broken hair which is seen in alopecia areata.

Both patients were counseled regarding the benign nature of the condition and advised to overgrow the hair in the surrounding area to camouflage the defect

Discussion:
Congenital temporal alopecia is a rare cause of non cicatricial alopecia seen in children around the age 2-9 years. The disorder is considered to be a result of genetic defect and though inheritance patterns of CTA are as yet unknown; para-dominant inheritance pattern has been suggested. Other suggested patterns include the possibility of mosaicism and an autosomal dominant inheritance especially when such cases are part of a syndrome.

Most reported lesions of CTA have been described as lacking any hair, although a few normal terminal or vellus hairs may be evident within the affected patch. In addition, CTA may not present as a completely bald patch but instead show a centrally localized hair tuft or as small islands of dark hairs.

The diagnosis of CTA is mainly clinical and based on its distinct clinical appearance and location and is not dependent on histology. Inui et al. has described a series of features that help in diagnosis of CTA. These include (1) A triangular or spear-shaped patch of hair loss located over the frontotemporal region of the scalp; (2) normal hair follicles with vellus-type hairs surrounded by a normal terminal hair area; (3) absence of fractured or exclamation mark hairs and no black or yellow dots with a preserved follicular orifice; and (4) lack of significant hair growth 6 months after confirming the presence of vellus hairs on dermoscopy.

The main differential diagnoses are alopecia areata, trichotillomania, traction alopecia, pressure alopecia, congenital aplasia cutis, tinea capitis and primary cicatricial alopecia.

However, the most significant differential is perhaps alopecia areata especially when it presents in a typical pattern or patients present at a late stage. Many cases of TTA are diagnosed and treated as alopecia areata.

Often the only way of confirmation of diagnosis is by histopathology which is invasive and not easily acceptable by the patients. In the recent times, trichoscopy has been emerging as the non-invasive tool of choice in differentiating the various differentials for this condition. Thus, scalp dermoscopy is an indispensable tool for a correct diagnosis.

Trichoscopy helps differentiate between these two diseases, avoiding the performance of biopsies to confirm the diagnosis.

Dermoscopic findings include normal follicular openings with vellus hairs covering the area of alopecia and terminal hairs on the outskirts of the lesion. Black and/or yellow dots and ‘exclamation mark’ hairs, which are present in alopecia areata, are absent in this dermatosis.

References
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8. Clinica features of CTa
