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CASE REPORT

Clinical and Trichoscopic Characteristics in a Case of Congenital Triangular Alopecia

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Abstract:

Congenital triangular alopecia (CTA) is a rare, asymptomatic, non-inflammatory and non-scarring form of alopecia with triangular, ovoid or lancet shape that is generally localized at frontotemporal region of scalp. It mostly appears at birth or within the first 9 years of age. Here, we presented a 9-months-old boy who had triangular-shaped, congenital alopecia involving the right fronto-temporo-parietal region and diagnosed as congenital triangular alopecia considering clinical and trichoscopic features that did not respond topical steroids and traditional treatment modalities. With this case, we wanted to remind how CTA is diagnosed in trichoscopic examination using a dermatoscope, accompanying symptoms and syndromes and treatment approaches in patients with CTA.

Keywords: *Congenital triangular alopecia, non-scarring alopecia, trichoscopic, dermatoscope*

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Introduction

Congenital triangular alopecia (CTA) is a rare disorder. It's a form of congenital, asymptomatic, non-inflammatory and non-scarring alopecia having a triangular, ovoid or lancet shape that is confined to the scalp. The lesion is often seen at the frontotemporal region of the scalp. The disease is also termed Brauer nevus. Its incidence is reported as 0.11% [1]. Its etiopathogenesis is unknown. Here, we aimed to present a case of congenital triangular alopecia and related trichoscopic findings.

Case Report

A 9-months-old boy presented to our outpatient clinic with a triangular, hair-free area at the scalp. In history, it was found out that the lesion was present since birth without enlargement in size and that hair in the area remained as thin hair without thickening. There was no history of trauma in the patient. It was also found out that the patient was treated with topical steroid ointment/lotion and traditional therapies (topical garlic administration) but did not respond. In dermatological examination, a triangular alopecia area was observed at the right fronto-temporo-parietal region (Figure 1). No erythema, induration or scarring was detected in the lesion area. In trichoscopy, terminal hair was seen in the areas surrounding alopecia area while vellus hair was observed in follicular ostia in the alopecia area (Figure 2). There was no yellow or black dot or dystrophic hair. Physical examination and development were normal. As parents declined biopsy, CTA was diagnosed by clinical and trichoscopic findings.

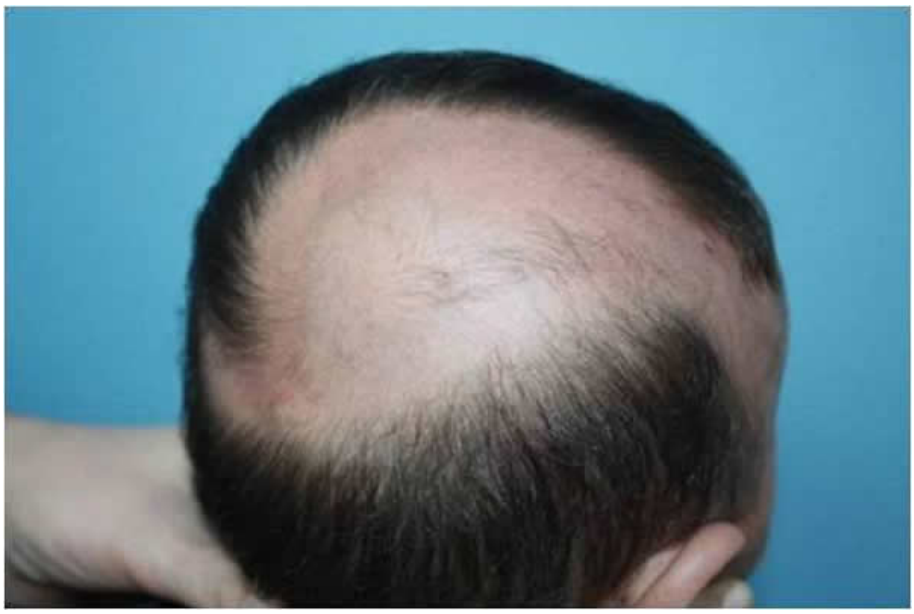

Figure 1. *A triangular, non-inflammatory, non-scarring alopecia in right fronto-temporo-parietal area at the scalp.*

Figure 2. *Vellus hair seen in normal follicular ostia in trichoscopic examination (Digital dermatoscope [Mole Max II] original magnification x20).*

Discussion

CTA generally appears at birth or within the first 9 years of life; however, there're cases of adult onset[2]. There's no gender preponderance. Although it is generally sporadic, there are reports of familial cases[3]. There are publications proposing that it's caused by a neuroectodermal disorder during embryonic period or suggesting that it is due to post-zygotic mutation and should be classified within epidermal nevus group[4]. There's localized miniaturization in follicles. However, the stimulus causing miniaturization is unknown[5].

Although it's typically localized at the frontotemporal region, it may be rarely seen at frontoparietal and occipital region. It's generally unilateral; however, it may also be bilateral[1]. The alopecia area is stable without progression and remains lifelong. There are no concurrent skin findings such as erythema, squamous changes, follicular pustules, discoloration or atrophy[4].

Trichoscopic examination using a hand-held polarized light dermatoscope or digital dermatoscope reveals short vellus hairs of varying length and white hairs in the normal follicular ostia in CTA[6, 7]. Vellus hairs are a non-specific but highly sensitive marker for CTA. In the interfollicular area, the presence of arborizing vessels and honeycomb pigment pattern has been reported as additional findings observed in some cases. The hair pull test is negative in CTA. Inui et al., proposed 4-item diagnostic criteria for CTA; i) triangular or lancet-shaped patch of alopecia on the frontotemporal scalp; (ii) vellus hairs surrounded by a normal terminal hair area with trichoscopically normal follicular openings; (iii) trichoscopically the lack of yellow dots, black dots, tapering hair, broken hair and loss of orifice; and (iv) persistence without significant hair growth for 6 months after

clinically or trichoscopically confirming the presence of vellus hair. It has been reported that using these diagnostic criteria, unnecessary biopsy from children with CTA can be prevented[7].

In histopathological examination, epidermis, dermis and total number of follicles are normal. However, terminal hair is replaced by vellus hair with miniaturization of follicles[2, 6]. Peribulbar inflammation is seen in alopecia areata which should be kept in mind in differential diagnosis while no inflammation is observed in congenital triangular alopecia[5].

In differential diagnosis, alopecia areata, alopecia mucinosa, tractional alopecia, trichotillomania, tinea capitis, primary cicatricial alopecia, aplasia cutis, androgenic alopecia, pressure alopecia and nevus sebaceous should be considered[1, 8].

CTA may be associated with syndromes such as phacomatosis pigmentovascularis, Klippel-Trenaunay syndrome, LEOPARD syndrome, Pai syndrome, Turner Syndrome[1, 9, 10]. In addition, in CTA, caf***é***-au-lait patches, multipl lentigines, mental retardation, epilepsy, Dandy-Walker malformation, spina bifida, cardiac, osseous and dental abnormalities, congenital dislocation of the hip, hydronephrosis, hypospadias, tracheo-oesophageal fistula, iris naevus, leuconychia, dysaesthesia within hairless areas can be found[1, 3, 9, 10].

CTA is an asymptomatic disease with a stable course, there's no effective treatment and in most cases therapeutic intervention is not required, unnecessary treatments such as steroids should be avoided. In the literature, cases with better cosmetic outcomes were reported following hair implant[8] and surgical resection[1]. Partial success has been reported with topical minoxidil[1, 5]. A patient who benefited from minoxidil treatment and experienced recurrence of alopecia after treatment discontinuation was reported. It has been suggested that minoxidil treatment may be effective by preventing follicle miniaturization in CTA[5]. We also started 2% topical minoxidil therapy in our patient.

With this case, we wanted to remind that it's easy to diagnose CTA with trichoscopic examination using a dermatoscope, patients with CTA should be examined in detail in terms of accompanying findings and syndromes, and steroids are not used in the treatment.

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