BILATERAL CONGENITAL TRIANGULAR ALOPECIA ASSOCIATED WITH ALOPECIA AREATA: REPORT OF TWO CASES

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Abbreviations **AA**= alopecia areata; **CTA** = congenital triangular alopecia.

Case 1. An 8-year-old girl with normal psychomotor development and up-to-date vaccinations presented for evaluation of two large patches of hair loss on both sides of the scalp, present since birth. For the past two years, she had also noted partial eyebrow alopecia and complete eyelash loss. The patient had not received any specific treatment and reported no systemic symptoms or associated comorbidities.

Her mother had three patches of scalp alopecia areata. There was no history of maternal drug intake during pregnancy; delivery was vaginal, and there were no complications during childbirth.

Cutaneous examination revealed well-demarcated triangular alopecic patches measuring 13 × 10 cm, bilaterally located in the temporoparietal regions of the scalp (Fig. 1, 2). The hair pull test at the margins of both patches was negative.

The medial portion of both eyebrows showed an ill-defined alopecic patch measuring 0.5 × 0.5 cm, and the eyelashes of the lower eyelids were bilaterally absent (Fig. 3); the hair pull test was also negative at these sites. Both cheeks displayed ill-defined light-brown pigmentation. The remainder of the mucocutaneous examination, including teeth and nails, was unremarkable.

Dermoscopy of the congenital alopecic patches of the scalp revealed numerous empty follicles and occasional vellus hairs, findings suggestive of congenital triangular alopecia. Dermoscopy of the eyebrow alopecic patch showed hair rarefaction, empty follicles, and vellus hairs. Dermoscopy of the left lower eyelid demonstrated complete hair loss, numerous empty follicles, and linear and branching vessels at the periphery.

Based on the clinical and dermoscopic findings, a diagnosis of congenital triangular alopecia (CTA) associated with bilateral alopecia areata of the eyebrows and eyelashes was made.

The patient was treated with topical minoxidil 2% once daily on the scalp lesions, mometasone furoate 0.1% for the eyebrow patch, and desonide 0.05% for the lower eyelids, in combination with oral hair supplements. After one month, mild hair regrowth was observed in the CTA patches, with no regrowth of the eyebrows or eyelashes.

Case 2. A 9-year-old girl with normal development and up-to-date vaccinations presented for evaluation of alopecic patches on both sides of the scalp present since birth, as well as a patch in the occipital region that had developed one month earlier. Her mother also reported patchy hair loss in both eyebrows for one month, without hair loss at other sites.

The patient had no systemic symptoms or associated comorbidities and had not undergone any specific treatments. Family history was negative for similar conditions.

Cutaneous examination revealed triangular alopecic patches measuring 5 × 6 cm, bilaterally located in the temporoparietal regions of the scalp, more evident on the left side. The hair pull test at the margins of the patches was negative.The bilateral medial portion of the eyebrows showed an ill-defined

Fig. 1

Fig. 2

Fig. 3

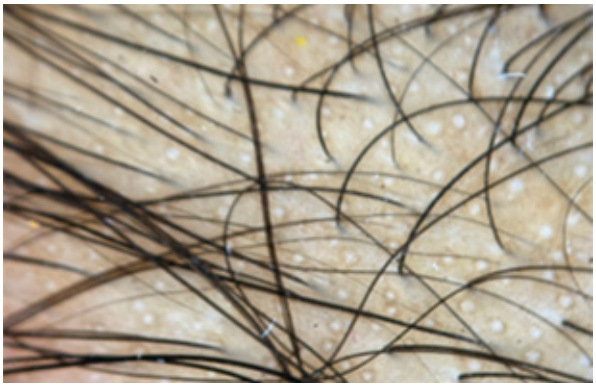

Fig. 4

Fig. 1, 2, 3, 4: Bilateral congenital triangular alopecia associated with alopecia areata of the eyelashes and eyebrows (case 1, Fig. 1, 2, 3). Figure 4 shows trichoscopy of a patch of congenital triangular alopecia (case 2) with empty follicular openings and a few vellus hairs.

alopecic patch measuring 0.5 × 0.5 cm with eyebrow rarefaction, and a round alopecic patch measuring 4 × 4 cm was observed in the occipital scalp, also with a negative hair pull test. The remainder of the mucocutaneous examination, including teeth and nails, was normal.

Dermoscopy of the triangular alopecic scalp patches showed numerous empty follicles and occasional vellus hairs (Fig. 4), consistent with CTA. Dermoscopy of the eyebrows revealed hair rarefaction, empty follicles, and vellus hairs, compatible with alopecia areata. Dermoscopy of the occipital alopecic patch showed empty follicles and some broken hairs, findings suggestive of alopecia areata. Based on the clinical and dermoscopic findings, a diagnosis of CTA associated with bilateral alopecia areata of the eyebrows and scalp was established.

The patient was started on topical minoxidil 2% once daily on the CTA lesions and mometasone furoate 0.1% for the eyebrow and scalp patches.

Discussion. Congenital triangular alopecia (CTA), also known as Brauer nevus or hypotrichotic nevus, is characterized by circumscribed miniaturization of hair follicles resulting in the presence of vellus hairs. The temporal region is most commonly affected – 54.6% – , followed by the frontotemporal region – 28.5% – and the occipital region – 2.5% of cases – (1). The lesion is characteristically triangular with an anterior apex; it is unilateral in 80% of cases, more commonly described on the left side, and only rarely bilateral.

The incidence is 0.1%; 36.5% of cases are present at birth, more than half (55.8%) are noted between 2 and 9 years of age, and only 3.8% are diagnosed in adulthood (2). There is no gender predilection, and the condition predominantly affects individuals of Caucasian ethnicity. Once established, it remains unchanged throughout life, in the absence of inflammation and scarring.

The cause of CTA remains unknown. It is a developmental defect initially considered congenital but currently thought to be acquired. Familial cases have been reported; CTA has been hypothesized to be transmitted as a paradominant trait, expressed only after postzygotic mutation of the corresponding wild-type allele at an early stage of development (2).

The diagnostic criteria for CTA are shown in table 1.

TABLE 1: Diagnostic criteria for CTA (2).

1. Triangular- or lance-shaped alopecic patch involving the frontotemporal scalp.
2. Trichoscopically normal follicular openings with vellus hairs, surrounded by an area of normal terminal hairs.
3. Trichoscopic absence of broken hairs, tapered hairs, black dots, yellow dots, or loss of follicular openings.
4. Persistence without significant hair regrowth for at least 6 months after clinical or trichoscopic confirmation of vellus hairs.

The differential diagnosis of CTA includes alopecia areata and trichotillomania.

Trichoscopy of CTA shows normal follicular openings with vellus hairs surrounded by normal terminal hairs, in the absence of broken hairs, tapered hairs, black dots, and yellow dots, which are instead typically observed in alopecia areata (3) and reflect disease activity.

No treatment is required, and counseling is generally sufficient. Therapeutic approaches that have been attempted include topical minoxidil, surgical excision, and hair transplantation. Hair transplantation and surgical excision represent the main therapeutic options in cases with significant cosmetic and emotional impact. Complete excision may be considered for small lesions, whereas in other cases hair restoration surgery is required (4).

Trichoscopic features of eyebrow and eyelash alopecia areata (AA) include uniform miniaturization of hair shafts, which is the most common finding during disease remission. Black dots, broken hairs, and exclamation mark hairs are predictors of disease activity (5).

The prevalence of eyebrow and eyelash involvement in AA is not precisely known. In studies with larger case series, eyebrow involvement ranges from 19.8% to 62.8%, and eyelash involvement from 10% to 56.4% (6, 7).

Medium-potency topical corticosteroids and intralesional steroids have long been used to treat eyebrow alopecia. Intralesional triamcinolone acetonide (2.5 mg/mL; 0.5 mg per eyebrow) may be injected every 4-6 weeks for up to 6 months. Topical minoxidil 5% may also be used on the eyebrows, although studies on its efficacy are lacking (8).

In a retrospective study including 119 patients treated with topical tofacitinib for AA, complete regrowth of eyebrows and eyelashes was achieved in 34.5% and 38.7% of patients, respectively, after ≥6 months of treatment (8). Topical tofacitinib 2% gel twice daily for the eyebrows and 0.005% ophthalmic solution once daily for the eyelashes proved effective, resulting in partial or complete regrowth in 66.7% of cases of eyebrow AA and 100% of cases of eyelash AA. Complete regrowth of the upper eyelashes in localized AA has also been reported with tofacitinib 2% solution applied to the upper eyelid once or twice daily for 7 months (8).

The JAK inhibitor baricitinib has been approved by the U.S. Food and Drug Administration (FDA) for the treatment of AA. In a baricitinib study that included 36.2% of AA cases, 1,200 patients were randomized to receive baricitinib 4 mg, 2 mg, or placebo for 36 weeks; eyebrow and eyelash outcomes were assessed using the Clinician-Reported Outcome (ClinRO). Among patients treated with baricitinib 4 mg, 36.2% and 36.8% achieved, respectively, a terminal score of 0-1 and a reduction of at least 2 points in the eyelash ClinRO score from baseline, compared with 4.4% and 6.9% in the placebo group. Fewer studies have evaluated ruxolitinib; complete eyebrow regrowth was reported in a single patient after 12 weeks of treatment with topical ruxolitinib 0.6% cream (8).

Among prostaglandin analogs, bimatoprost is one of the most commonly used treatments. In a study of 41 subjects with alopecia universalis, application of topical bimatoprost 0.03% to the eyelid margin once daily for one year resulted in mild, moderate, or complete eyelash growth in approximately 70.3% of patients. Other experimental studies have instead shown a lack of efficacy of latanoprost (8).

In a clinical study of three patients with refractory eyebrow alopecia areata, four sessions of treatment with a 904-nm pulsed diode laser resulted in complete eyebrow regrowth in five of the six treated patches (8).

Conclusions. Congenital triangular alopecia should be considered as a possible diagnosis in any patient presenting with non-scarring alopecia. Accurate diagnosis of CTA is essential to prevent unnecessary investigations and treatments.

Conflicts of interest

The authors declare that they have no conflicts of interest.

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