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Congenital temporal triangular alopecia: A typical Brauer nevus

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To the Editor:

A 9 year-old-boy presented with a localized area of diminished hair over the left temporoparietal region. The lesion had already been noticed at birth because he was born with terminal scalp hair. Since then, there had been no change in the appearance of the hairless area. The patient was evaluated in the past by two general practitioners, who diagnosed it as a case of alopecia areata (AA). There was no history of any intrauterine pressure, or of obstetric or acquired trauma. The family history was unremarkable. No cutaneous defect was seen at the site of the alopecia. Upon dermatological examination, there was a 6 × 9 × 10 cm less hairy and fine hairy triangular alopecic patch over the left temporoparietal region (Fig. 1). On the alopecic area there were mostly vellus hairs and a small number of terminal hairs. Between the triangular patch and left frontal area, there was a hair fringe. No scaling, erythema, scarring, or induration was seen in the alopecic area. A hair pull test was negative. Exclamation mark hairs were absent. Other dermatological examinations, including the nails and mucosa, and the remaining physical examination were normal. Routine laboratory tests were within normal limits. A trichogram or scalp biopsy could not be performed because the patient's parents did not give consent. All of the typical history and clinical findings of the patient suggested a diagnosis of congenital temporal triangular alopecia.

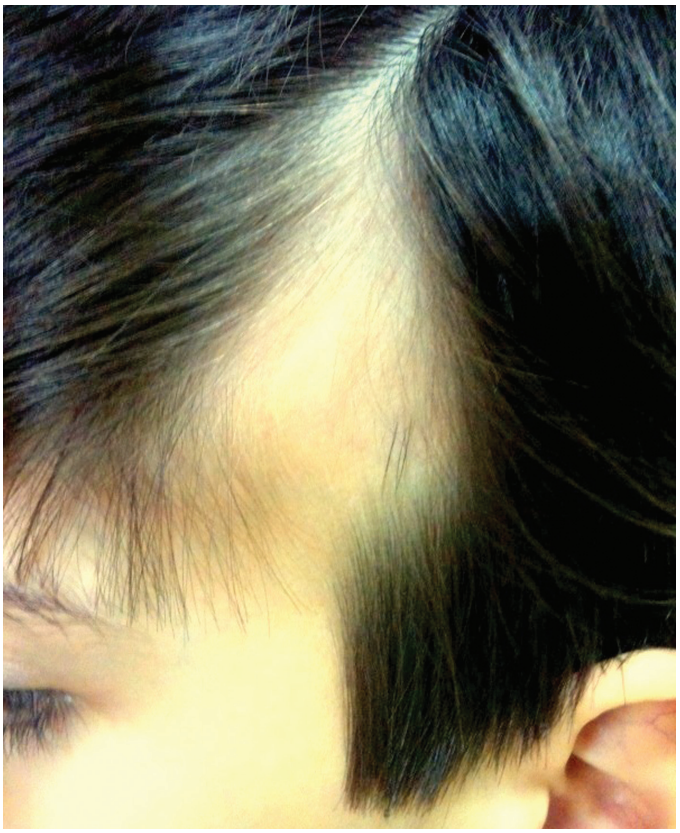

Figure 1 | Clinical view of the typical triangular alopecic patch with a small fringe of terminal hair.

Temporal triangular alopecia (TTA) is a well-circumscribed triangular or lancet-shaped area of non-cicatricial hypotrichosis in the frontotemporal area (1–3). Sometimes the anterior margin is separated from the lesion by a small fringe of normal hair (4). This condition was first described by Sabouraud in 1905 (3, 5). In 1926, the lesion was defined again by Brauer as a part of Brauer's syndrome (focal facial dermal displasia type 1 = hereditary symmetrical aplastic nevi of the temples, bitemporal aplasia cutis congenita), which is characterized by temporal skin depressions that resemble "forceps marks" (6). Around 54 cases have been reported so far (3, 5). The cause is unknown. It is unilateral in 80% of cases (1) and it usually appears sporadically (7). It usually manifests after 2 years of age. Parents become aware of the disease when the vellus is replaced by terminal hair. However, it may also occur within families as a paradominant trait (3). In addition, it has been argued that TTA may reflect a mosaicism. This entity usually occurs as an isolated anomaly but it may also be a part of multisystemic birth anomalies such as cerebello-trigeminal-dermal dysplasia or phakomatosis pigmentovascularis type 2 (5). On the other hand, the condition may occur as part of a syndrome with autosomal-dominant inheritance (7). Vellus hairs are present in the affected area and occasionally a few terminal hairs are retained (1, 7). The skin is normal in other respects (4). Exclamation mark hairs are absent and the hair pull test is negative (7). Histopathologically, hair follicles are miniaturized (5, 7) and the hair density of vellus hairs is normal (7). The diagnosis of TTA was made based on clinical features and typical location, and not on biopsy results (5). It has been confused with AA, but typical history and clinical findings help distinguish it from AA and other patterned circumscribed alopecias (1, 3). In particular, the unresponsiveness to topical steroids, lack of exclamation mark hair, and negativity of the hair pull test help rule out AA (5, 7). In addition, trichotillomania, tinea capitis, androgenetic alopecia, and aplasia cutis should be considered in differential diagnosis (5). Our patient's lesion was noticed at birth because he was born with terminal scalp hair. Our patient had typical features of TTA: persistence of the lesion since birth, a typical triangular shape of the lesion, a fronto-temporal location, the presence of a small terminal hair fringe on the border of the frontal region, absence of exclamation mark hairs, normal appearance of the skin in the affected area, absence of any inflammation, scarring, erythema, or induration on the lesional skin, and the presence of mostly vellus hairs on the lesion. There is no specific treatment for this condition but the patient can benefit from hair transplantation in the future (3, 5, 8). Our case was presented to raise awareness of this very rare and unique lesion that very closely mimics AA. Patients may thus be spared from redundant intervention and unnecessary treatment.

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