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Bilateral Congenital Triangular Alopecia Associated With Congenital Heart Disease and Renal and Genital Abnormalities

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To the Editor:

Congenital triangular alopecia, also known as temporal triangular alopecia or Brauer nevus, is a nonscarring circumscribed permanent and asymptomatic alopecia that was first described by Sabouraud in 1905. It is usually found on the frontotemporal area and affects only 1 side of the head. Histopathology of the affected area reveals reduced hair follicle size, although hair density remains normal, with no other significant abnormalities.[1] Diagnosis is usually clinical. Other causes of nonscarring circumscribed alopecia must be ruled out, especially alopecia areata, with which it is often confused.[2] In the literature, there are reports of different conditions that coexist in patients with congenital triangular alopecia. We present the association between bilateral congenital triangular alopecia and a multiple malformation syndrome.

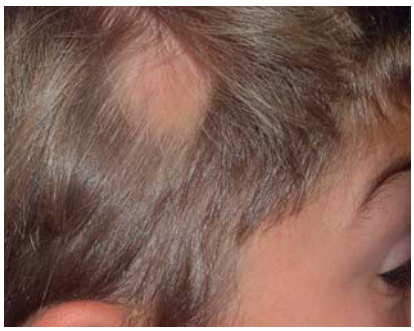

Figure 1. Oval alopecia plaque on the right temporal region.

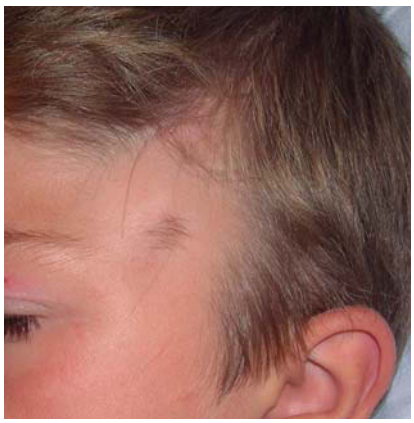

Figure 2. Congenital triangular alopecia on the left frontotemporal region reaching the hairline.

The patient was a 7-year-old boy with a history of congenital heart disease involving a perimembranous ventricular septal defect and an atrial septal defect with no hemodynamic consequences. He also had a history of left hydronephrosis, subcoronal hypospadias, Wormian bones, and recurrent bronchiolitis. The patient was referred because of the presence on the scalp of 2 areas with finer, lighter-colored hair, which his parents remembered as being there since birth. There had never been total hair loss in the area, and the patient

had not responded to topical corticosteroids. There was no history of injury or a family history of similar processes.

Physical examination revealed 2 approximately oval areas on the temporal region. These were well delimited, 2 × 4 cm in diameter, and covered with vellus hair, with no exclamation mark hair (Figures 1 and 2).

The pull test was negative and the surface of the underlying skin was normal (no atrophy, flaking, follicular pustules, or changes in coloring).

Although there have been few published reports of congenital triangular alopecia, it is not uncommon, and some authors have reported a frequency of 0.11%. It seems to affect both sexes equally and has been reported mainly in whites, although there have also been cases among Asians and African Americans.[2]

Clinically, it is characterized by finer hair in a more or less triangular area with blunt angles and the base of the triangle lying towards the hairline, although it can sometimes be oval or round.[3] It is usually unilateral and affects the frontal, parietotemporal area, although there have been reports of lesions in the occipital region and bilateral involvement (20%). Although it is considered congenital, it usually appears between 3 and 6 years of age, and sometimes during adolescence or adulthood. Given the variability in age at diagnosis, some authors prefer to call it temporal triangular alopecia.[3]

The pathogenesis of congenital triangular alopecia remains unknown and its genetic basis is unclear. Paradominant inheritance has been postulated.[5] This would explain the sporadic nature of the process (with very few familial cases[6] and a limited number of members affected), its tendency to be unilateral, and its association with phacomatosis pigmentovascularis as a component of twin spotting.[7] It has been associated with Down syndrome, leuconychia, sectorial iris hyperpigmentation, woolly hair nevus,[8] mental retardation,[6] epilepsy,[6] Dandy-Walker malformation,[6] LEOPARD syndrome,[9] and aplasia cutis congenita. Some authors believe that developmental neurological abnormalities and phacomatosis pigmentovascularis are not simple causal associations.[6,9]

Histopathology reveals normal hair density—although the hair is of the vellus type and not terminal hair—and no added scarring or inflammatory abnormalities are apparent.[1]

Diagnosis is based on its stable nature and characteristic clinical pattern, and a histopathology study is not usually necessary. In addition to the clinical history and location, the normal appearance of the skin in the affected area is very important.[2,4] Differential diagnosis should be made with other causes of nonscarring circumscribed alopecia, especially alopecia areata, which can also appear in childhood, even at birth,[10] but which is associated with a positive pull test, exclamation mark hair, and completely bald areas. In addition, alopecia areata, is not static and often regresses spontaneously or after administration of intralesional or topical corticosteroids.[1,10] In some cases, a histopathology study is necessary to differentiate between the 2 entities, but the result might not always be definitive. Perifollicular inflammation can be observed in alopecia areata, except for some cases of chronic alopecia areata.[1]

There is no effective treatment for congenital triangular alopecia and it is not usually necessary. The nature of the entity should be explained to parents so that they can avoid fruitless or harmful treatments (especially topical corticosteroids).[1,2] The symptoms remain stable for life and both males and females can undergo surgical removal of the affected area. In some cases, hair has been implanted using micrografts.[1]

In conclusion, we present a new case of bilateral congenital triangular alopecia in a patient with multiple malformation syndrome. This may be another illustration that perhaps these associations are not casual.

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