

# Congenital Temporal Alopecia

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A 26-year-old male physician had congenital temporal alopecia. The histological findings of this type of alopecia are reported for the first time, to our knowledge, and they are compared with those found in alopecia areata. The clinical and microscopic findings of this rare form of alopecia are distinct and allow easy separation from other forms of alopecia.

Congenital temporal alopecia, also known as alopecia triangularis congenitalis, was originally described by Sabouraud in 1906 as a rare form of alopecia occurring on the temples.<sup>1</sup> Galewsky<sup>2</sup> reported another case in 1932, and since that time only two other cases<sup>3,4</sup> have been described. None of the previous investigators have used histopathological findings to describe this disease. Originally reported as an oval patch, the lesion was subsequently described<sup>2</sup> as resembling the shape of a triangle, with the base at the junction of the frontal and temporal hairline. The sides of the oval or triangle extend 2 to 4 cm onto the scalp, pointing toward the vertex. The surface of this area is devoid of almost all terminal hair.

This report describes another case and the biopsy findings of this rare form of alopecia.

## Report of a Case

A 26-year-old white male physician was seen at the dermatology clinic of the Medical College of Virginia in July 1973 for consultation concerning areas of bilateral alopecia present since birth. His mother had noted these areas in an otherwise normal scalp when the patient was an infant. All other epidermal appendages, teeth, nails, and other hair, developed normally. The patient's general health was excellent, and no other family member was affected by this type of alopecia. The mother has psoriatic arthritis, and the father has male-pattern baldness.

The results of a complete physical examination were normal, except for two sharply outlined areas of alopecia in the hairline at the junction of the frontal and temporal portions of the scalp. The lesion on the right temple resembled a triangle. Its base on the anterior hairline was 2.5 cm long, and its sides were 4 cm long and directed toward the vertex. Except for a few terminal hairs present in the center of the lesion, it was devoid of all hair (Fig 1). The lesion on the left temple was similar in shape, except the anterior one third was not devoid of all hair. The skin in both areas was normal in appearance.

## Histological Findings

Serial sections from a 4-mm punch biopsy were stained with hematoxylin-eosin, PAS, Alcian blue, Masson trichrome, and Verhoeff-van Gieson and examined under light microscopy. The epidermis appeared normal, except for several follicular keratin plugs. No mature hairs were seen. Vellus-like hair follicles were present in the upper dermis (Fig 2 and 3). Normal appearing sebaceous glands

and arrectores pilorum muscles were associated with these hair follicles. The dermis contained apocrine and eccrine sweat glands and was free of inflammatory infiltrate. No connective tissue abnormalities were present on special staining.

## Comment

At our present state of knowledge, congenital temporal alopecia is of no special medical importance. It is apparently not inherited and occurs spontaneously at birth in male and female infants. The alopecia is occasionally bilateral and is seen as soon as hair growth on the scalp is noticed. There are no nevus structures present. No skin changes (erythema, atrophy, follicular plugging) precede the onset of alopecia, nor does scarring follow it. The condition has been found to be unresponsive to therapy,<sup>1,6</sup> and no other ectodermal defects have been associated with it. It persists but does not increase in size. The cause of this alopecia is unknown, and it has been incorrectly attributed to the application of forceps at birth.

Alopecia areata and congenital temporal alopecia share one interesting finding; they are both diseases in which sharply outlined areas of nonscarring alopecia occur, but here the similarity ends. Van Scott considers the primary lesion of alopecia areata to be in the connective tissue surrounding blood vessels supplying the dermal papillae.<sup>7,8</sup> He found degenerative changes in this tissue, with a dense cellular infiltrate surrounding

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Fig 1.—Triangular area of alopecia is seen at anterior hairline pointing toward vertex. Normal male hairline pattern is seen above area of alopecia.



Fig 2.—Biopsy specimen from lesion showing follicular plug, vellus-like hair follicle in upper dermis, and normal skin appendages (hematoxylin-eosin, original magnification  $\times 56$ ).



Fig 3.—Close-up view of biopsy specimen demonstrating vellus-like hair follicle. Notice small dermal papilla invaginating into matrix (hematoxylin-eosin, original magnification  $\times 160$ ).

the hair bulb. From those findings, he concludes that the dermal hair papillae are severely damaged but are capable of regenerating and producing hair, as shown by the clinical regrowth of hair following systemic therapy with adrenal corticoids.

Other dermal papillae abnormalities mentioned by Van Scott include the presence of cells in mitotic division and the presence of large amounts of melanin in the dermal papillae and in the underlying corium of hair roots in the anagen phase of the growth cycle.

In our patient, these dermal papillae abnormalities were not present. The papillae were small and contained no evidence of mitoses or the presence of melanin. The dermis was free from inflammatory infiltrate and did not have any connective tissue

changes. The only abnormal findings present were vellus-like hair follicles.

It is also necessary to differentiate congenital temporal alopecia from long standing alopecia areata. Van Scott<sup>7</sup> described a case of persistent alopecia areata of the scalp in a 23-year-old man. He found that rudimentary hair roots were detectable as local buds of epithelial cells extending downward from the base of the follicular neck. Dermal papillae were not seen beneath these buds. Our patient had hypoplastic dermal papillae.

One must be aware of the characteristics of congenital temporal alopecia in order to distinguish it from the relatively common condition of alopecia areata. Both types of alopecia can appear identical on gross examination, but it is possible to distinguish the two disorders on the

basis of the patient's history, the location of the lesion, and the histologic findings.

#### References

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