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Case Report

Congenital Triangular Alopecia Indicating Underlying Disorders: The 128th Case

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ABSTRACT

Congenital Triangular Alopecia (CTA), also known as temporal triangular alopecia or Brauer nevus, is a non-cicatricial pattern of hair loss that can present at birth, in infancy, or later in life. The condition typically affects the frontotemporal region of the scalp and presents as a non-progressive triangular, or lancet shaped patch of alopecia. The exact etiology of CTA is unknown, though it was once considered to be congenital. It appears to be sporadically acquired in most individuals; however, there have been a small number of familial cases. It is postulated that CTA may reflect mosaicism and may be inherited as a para-dominant trait where a postzygotic loss of the wild type allele in a heterozygote state leads to disease. Others have suggested that CTA may be an ectodermal defect and should be included in the group of epidermal nevi, though no consensus has been reached. The estimated incidence in the general population is 0.11%. The disease histologically presents as replacement of normal hair follicles with sparse vellus hair follicles. The total number of hair follicles is usually within the normal range. There is no evidence of inflammatory infiltrate and the epidermis and dermis are unremarkable. Hair follicles are miniaturized with an increased proportion of vellus or intermediate hairs. CTA is associated with other conditions and it can be a useful signal to clinicians to look for other disorders that may be present in the patient. CTA has been associated with bone abnormalities, café-au-lait patches, congenital heart defects such as atrial septal defects and mitral regurgitation, Down syndrome, Dandy-Walker malformation, and phakomatosis pigmentovascularis. These are a few of the many conditions associated with CTA. When concerned parents bring their child to their clinician for temporal balding, this may help prompt a more detailed examination of the infant to search for associated conditions.

Keywords: Congenital triangular alopecia; Triangular temporal alopecia; Alopecia; Non-cicatricial; Hair loss; Intralesional Kenalog©; Underlying disorders

INTRODUCTION

Congenital triangular alopecia (CTA) or triangular temporal alopecia (TTA) also known as a Brauer nevus is a benign non-cicatricial, localized pattern of hair loss that typically affects the frontotemporal region of the scalp. Although, there have been a few cases reported where the occipital aspect of the scalp was also involved. It is normally a non-progressive disorder, which presents as a triangular, oval, or lancet shaped patch of hair loss. CTA was first described by Raymond Sabourad in his "Manuel élémentaire de dermatologie topographique régionale", and was labeled as "alopecia triangulare congenitale de la temp."

The majority (58%) of cases present in children from ages two through nine, a third of cases (36.5%) present at birth, while a tiny (3.8%) percentage present in adulthood. An overwhelming majority (79%) of patients present with unilateral hair loss, while only 18.5% of patients had bilateral involvement. Only 2.5% of cases presented with occipital alopecia. There was an even distribution of cases between the two genders with a slightly higher (3.2%) prevalence in males. Although most patients that

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present with CTA are Caucasians, a small percentage of patients are African-American and Asian [1].

Patients are commonly brought to their pediatricians by their parents after the first year of life, likely because the sparse hair present on an infant's scalp prevents parents from realizing there is a lack of hair growth in the above-mentioned regions of the scalp [1]. It is fortunate that parents eventually present this finding to their child's doctor because 15% of cases of CTA are associated with other, more severe congenital anomalies.

CASE

At their first follow-up visit, the diagnosis of TTA was discussed with our patient and the specialist suggested intralesional Kenalog© injections (ILK). Our patient was initially hesitant on receiving the injections into her scalp, so they came to a compromise where they would inject the left frontotemporal aspect of the scalp first and follow-up in 6 weeks. A physical examination of the patient's scalp performed during this appointment showed that the patient's hair was subjectively thicker and longer than at baseline. This improvement was seen with the addition of the Clobetasol solution to our patient's hair growth regimen, prior to ILK administration.

During the second follow-up visit with the dermatologist, the patient reported slight improvement of hair volume to the injected side. She endorsed noticing fine hair appearing in the injected area. Although, on physical examination, the left scalp did not show significant response to the ILK which suggested, according to the specialist, permanent scarring to the majority of the area. Physical exam also revealed that hair growth seems thicker and longer than at baseline especially at the right side of the scalp. There were a few scattered miniaturized hairs still present at center of the affected area on the left side of the patient's scalp. The patient had decided to receive ILK to both sides of her scalp. The specialist then began intermittently administering 10mg/ml Kenalog injections to the affected areas and instructed her to continue using her topical medications.

Histologic sections of a punch biopsy of both the left and right temporal scalp demonstrated an unremarkable epidermis, dermis, and subcutaneous fat, which presented with orthokeratosis, no evidence of band-like inflammation, basal layer damage, psoriasiform hyperplasia, or spongiosis. The terminal hair follicles included showed no evidence of perifollicular inflammation or fibrosis. There was no evidence of miniaturization, peribulbar inflammation, or periappendageal inflammation. Sebaceous lobules appeared intact and without atrophy. A single catagen phase follicle was present and cross sections showed mature terminal hair follicles without evidence of significant inflammation.

After receiving ILK on a few subsequent follow-up visits, the dermatologist noticed that hair regrowth was minimal and that the effect of the ILK was plateauing. The dermatologist decided to discontinue ILK injections and our patient was referred for cosmetic hair implantation.

Figure 1: Bilateral non-traumatic, non-cicatricial hair loss along the frontal, temporal, and occipital aspects of the patient's scalp.

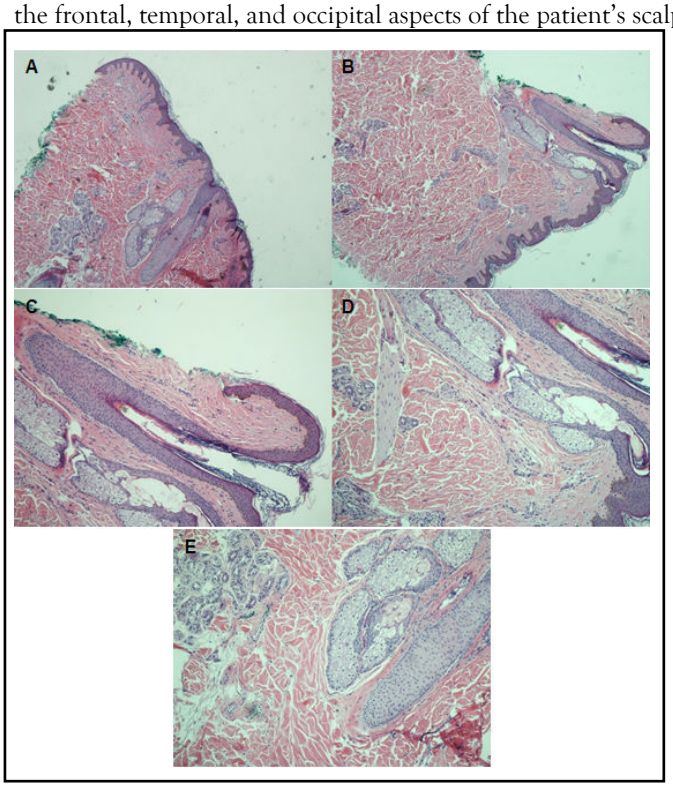

Figure 2: Histological Presentation. [A] Low power H&E (4x) photo demonstrating a normal epidermis, dermis, a terminal anagen hair follicle, intact collagen bundles, a normal distribution of adnexal structures and sebaceous glands, and vellus hair follicles. [B-D] Low power H&E (4x) and intermediate power H&E (10x) photos demonstrating a normal epidermis, dermis, erector pili muscle, sebaceous glands, and a terminal catagen hair follicle with a grayish inner root sheath that is crumbling, disappearing, and being replaced by red staining trichilemmal keratin from the outer root sheath. [E] Intermediate power H&E (10x) photo of unremarkable sebaceous and eccrine glands, and an anagen hair follicle in a sea of dermal collagen bundles that is devoid of any evidential source of follicular atrophy and loss.

DISCUSSION

Congenital triangular alopecia is a rare variant of alopecia, with only 127 cases presented in the literature thus far [2]. Our patient has moderate bilateral temporo-occipital hair loss, which, according to the literature, is exceedingly rare. It is presumed that because of the benign nature of this condition, most parents do not present their child to their clinician with a complaint of hair loss [1]. By the time parents present their child to their doctor with this concern, most patients are already 3 years of age [1]. Most CTA cases are benign and a cosmetic issue,

but for the 15% of CTA cases that are linked with other congenital anomalies, a presentation and diagnosis at an earlier age can help prompt a thorough evaluation of the child to look for other congenital diseases, and possibly prevent poor outcomes.

The diagnosis of TTA is made by combining the clinical, trichoscopic, and pathologic features of the disease. Clinically the patient presents with temporal hair loss with no lesions or scarring in the area to indicate a cause for the hair loss. Dermoscopy provides useful clues such as, short vellus hair of varying lengths, white hairs, epidermal scaling, interfollicular features like arborizing red lines and a honeycomb pattern. Histologically, normal terminal hair follicles are replaced by sparse vellus hair with a normal number of hair follicles. The most important finding is the absence of follicular stelae, indicating the follicles were always small, never having cycled from a location in the subcutis (where normal terminal follicular bulbs reside). The only other mentionable feature is that hair follicles are miniaturized, otherwise the entire epidermis and dermis is devoid of inflammation or abnormalities [3-6].

CTA has been reported to be associated with a handful of congenital syndromes that include Down's syndrome, Klippel-Trenaunay syndrome, LEOPARD syndrome, Pai syndrome, phakomatosis pigmentovascularis, and Turner syndrome. There are several disorders reported to be associated with CTA such as bone abnormalities, café-au-lait patches, congenital hip dislocation, and congenital heart disorders including, atrial septal defects, mitral regurgitation, pulmonary regurgitation, pulmonary stenosis, tricuspid regurgitation, and ventricular septal defects. CTA is also associated with Dandy-Walker malformations, dysesthesia within hairless areas, epilepsy, hydronephrosis, hypospadias, iris nevus, leukonychia, intellectual disability, multiple lentigines, recurrent bronchiolitis, spina bifida, dental abnormalities, tracheo-esophageal fistulas, woolly hair nevus, and Wormian bones [1]. Therefore, prompt pediatric evaluation is recommended if a child is exhibiting temporal hair loss.

CONCLUSION

CTA is a difficult to treat condition that is commonly asymptomatic, but it may generate distress and dysfunction in a patient's life. The patient may experience inter-personal difficulties that may negatively affect their mental, social, and even their physical health. Even after extensive therapeutic treatment with a variety of modalities, our patient's condition only minimally improved. Intralesional Kenalog, topical clobetasol, Rogaine©, and vitamins used in conjunction produced little improvement for our patient. Due to low treatment efficacy, our patient was recommended to undergo hair transplantation.

The benignity of CTA may not elicit a sense of urgency or further screening that may lead to poor patient outcomes. The condition is associated with numerous, underlying systemic conditions, which may have disastrous consequences. For example, if a ventricular septal defect goes unnoticed, cumulative damage to myocardial tissue can cause lead to Eisenmenger Syndrome (shunt flow reversal), which is indicative of pulmonary hypertension and imminent cardiac failure. The sparsity of documented CTA cases, in addition to the benign nature of the condition leads to limitations in our understanding and an inappropriate perception of CTA with respect to its association with congenital disorders. More research and further consideration should be taken for CTA because increasing awareness may lead to a positive shift in our patient's outcomes.

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