Portuguese Journal of Dermatology and Venereology

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CASE REPORT

Congenital triangular alopecia: case report

Alopecia triangular congênita: relato de caso

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Abstract

The authors present a case of congenital triangular alopecia (CTA), also known as temporal triangular alopecia, and debate its clinical presentation and diagnostic considerations. The patient, a 15-year-old male without comorbidities, exhibited left temporal alopecia since birth, seeking medical attention due to esthetic concerns. Dermatological examination revealed a well-defined area of alopecia, and trichoscopy exhibited vellus hairs of varying lengths, surrounded by terminal follicles and white hairs which are characteristic findings of the disease. The discussion explores the possible congenital nature of CTA, its clinical and trichoscopic features, histopathological characteristics, and the necessity for accurate differential diagnosis.

Keywords: Congenital alopecia. Non-cicatricial alopecia. Congenital triangular alopecia. Alopecia areata.

Resumo

Os autores apresentam um caso de alopecia triangular congênita (ATC), também conhecida como alopecia triangular temporal (ATT), debatem sua apresentação clínica e considerações diagnósticas. O paciente, um adolescente do sexo masculino de 15 anos sem comorbidades, referia alopecia temporal esquerda desde o nascimento e procurou ajuda médica devido ao impacto estético. O exame dermatológico revelou área de alopecia temporal bem definida e a tricoscopia exibiu pelos velus de comprimentos variados, cercados por folículos terminais e pelos brancos, que são achados característicos da doença. A discussão explora a possível natureza congênita da ATC, suas características clínicas e tricoscópicas, características histopatológicas e a necessidade de um diagnóstico diferencial preciso.

Palavras-chave: Alopecia congênita. Alopecia não-cicatricial. Alopecia triangular congênita. Alopecia areata.

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Received: 15-01-2024
Accepted: 01-06-2024
DOI: 10.24875/PJDV.24000008

Available online: 15-07-2024
Port J Dermatol and Venereol. 2024;82(4):262-265
www.portuguesejournalofdermatology.com

Introduction

Congenital triangular alopecia (CTA), also known as temporal triangular alopecia (TTA), is a rare form of scalp alopecia. It typically manifests at birth or in early childhood, with no gender predilection. Sabouraud called attention to this condition in 1905 and in 1926 considered it as part of the Brauer syndrome[1,2]. Also referred to as Brauer's nevus, CTA can occur as an isolated condition or have an autosomal dominant inheritance[2].

CTA often presents as a triangular-, oval-, or spear-shaped zone, measuring up to 4 cm in diameter, with fine hairs within the area of alopecia[3,4]. The most common presentation is unilateral, however, bilateral cases have been described[4]. Although its etiology remains unknown, associations with congenital anomalies such as pigmentovascular phakomatosis and Down syndrome have been reported[5]. CTA is a form of non-scarring alopecia and typically remains stable throughout life. Although not harmful or symptomatic, esthetic concerns may arise due to its distinctive shape and location.

Understanding the characteristics of CTA and exploring potential treatment options are crucial for providing appropriate support to patients.

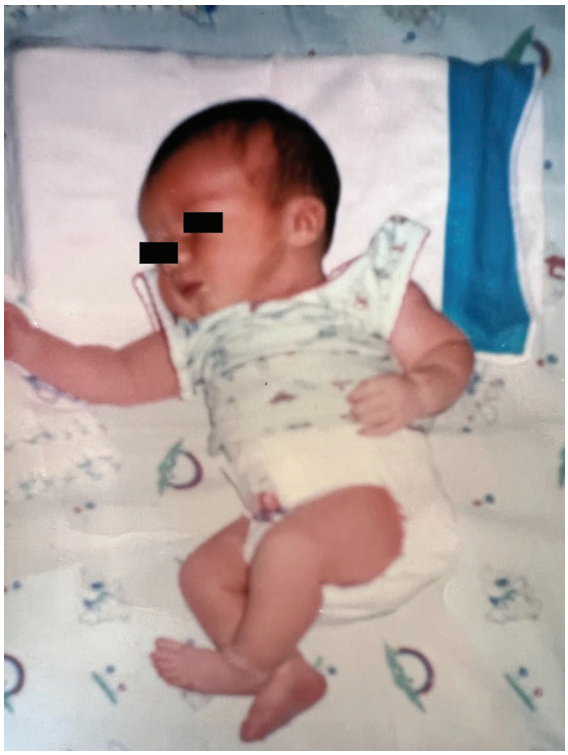

Figure 1. The alopecic area was observed in the patient as a newborn.

Case report

A 15-year-old Caucasian male student, born in Rio de Janeiro, without comorbidities or use of chronic medications, was observed in the dermatology department due to 'an area without hair'. He reported alopecia in this area since birth and denied any periods of worsening or improvement (Fig. 1). He previously received a diagnosis of alopecia areata and underwent topical corticosteroid therapy for a brief period but discontinued treatment due to a lack of clinical response.

Upon dermatological examination, the patient exhibited alopecia in the left temporal region, measuring approximately 3 cm by 2 cm, with a negative hair pull test (Fig. 2). Simultaneously, scalp trichoscopy revealed vellus hairs of varying lengths, surrounded by terminal follicles, white hairs, normal follicular openings, and an absence of erythema, yellow dots, or black dots (Fig. 3).

With the diagnosis of CTA, the patient received guidance about the natural course of the disease, which is characterized by stability. Furthermore, details were provided concerning potential poor responses when compared to treatment modalities utilized in different types of alopecia, such as minoxidil and topical corticosteroid therapy. Therefore, the patient opted not to undergo further therapeutic proposals.

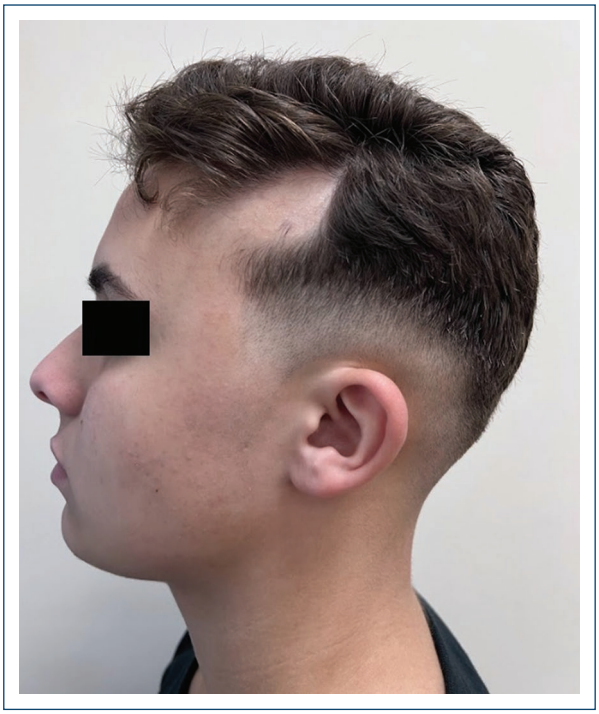

Figure 2. A triangular-shaped alopecic area with 3 cm in its largest diameter in the left temporal region. The hair pull test was negative.

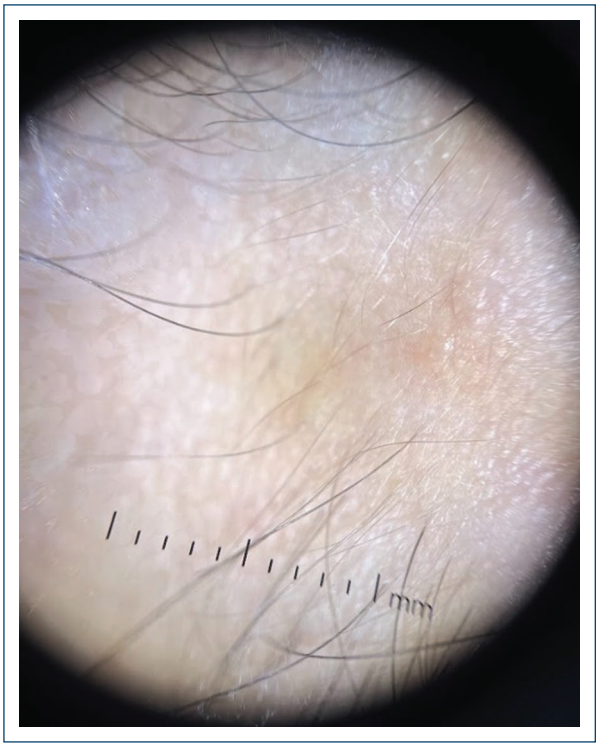

Figure 3. Scalp trichoscopy shows vellus hairs of varying lengths, surrounded by terminal follicles, white hairs, and normal follicular openings.

Discussion

CTA is considered congenital by most authors, although there is disagreement as its manifestation typically occurs between 3 and 6 years of age, and in some cases, it may present in adulthood. This discrepancy has led some authors to prefer the term 'TTA[2,6].' CTA manifests equally in both sexes, with most cases observed in Caucasians or Asians[7].

Pathogenesis remains unknown, and the genetic basis is not clearly understood. It is believed that the gradual transition from vellus to terminal hairs does not occur in the alopecic area.

Diagnosis is predominantly clinical[3,4] and certain trichoscopic findings contribute to confirmation in suspected cases. These findings include short vellus hairs of varying lengths, white hairs, typical location, the absence of change in size and typical shape of the alopecic area, and numerous vellus hairs in trichoscopy. Lack of response to intralesional/topical corticosteroids is a difference regarding confusion with alopecia areata[2,8].

Characteristic histopathological features include the absence of inflammatory cells in the dermis, unaltered collagen, and normal sweat glands. The epidermis appears normal with follicular keratosis. Hair follicles are reduced in size with preserved density, and there is a replacement of terminal hairs by vellus hairs, resembling androgenetic alopecia. The absence of fibrous scarring tissue adjacent to the outer root sheath distinguishes CTA from androgenetic alopecia[5,9].

The primary differential diagnosis is alopecia areata. Although both manifest as a patch of alopecia, these entities differ in terms of symptoms, clinical signs, trichoscopic features, causes, and progression[10]. Both conditions (CTA and alopecia areata) can have a significant emotional impact due to hair loss, affecting the self-esteem and body image of affected individuals. However, it is crucial to differentiate between these conditions for accurate diagnosis and appropriate treatment. CTA is a stable and non-progressive condition, whereas alopecia areata may have a more variable and unpredictable course[7,10].

Furthermore, other differential diagnoses to consider in the clinical presentation of patchy alopecia include aplasia cutis, chemical or physical trauma, end-stage lichen planopilaris, traction alopecia, trichotillomania, and tinea capitis[6].

In the treatment of CTA, options include esthetic camouflage, hair transplantation, and surgical excision of the affected area, particularly in those patients with esthetic/emotional concerns[6,7].

Knowledge of the disease and its associations enables precise diagnosis, avoiding unnecessary and traumatic treatments.

Conclusion

Congenital triangular alopecia (CTA) is a rare, benign condition of non-scarring alopecia that usually presents at birth or in early childhood. Accurate identification of this condition is essential to avoid erroneous diagnoses and unnecessary treatments. Understanding CTA and its specific clinical manifestations allows dermatologists to provide appropriate guidance and emotional support to patients, helping them manage their expectations regarding treatment and the prognosis of the disease. Patient education about the benign nature and stability of CTA can reduce anxiety and improve satisfaction with clinical management.

Funding

None.

Conflicts of interest

None.

Ethical disclosures

Protection of human and animal subjects. The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

Use of artificial intelligence for generating text. The authors declare that they have not used any type of generative artificial intelligence for the writing of this manuscript or for the creation of images, graphics, tables, or their corresponding captions.

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