

Congenital Triangular Alopecia - Report of a Case and Review of the Literature

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Abstract

We report a 3-year-old Chinese girl who presented with an 18-month history of a localized asymptomatic triangular area of hair loss in the right frontotemporal area. There was no history of trauma. The parents were non consanguinous and there was no family history of a similar lesion. Her 42-year-old father had androgenic alopecia that was first noted 3 years ago. Physical examination revealed a patch of alopecia 3.5 x 4 cm over the right frontotemporal area, with its apex towards the vertex. The overlying skin was otherwise normal. In particular, there was no scaling, scarring, erythema, or induration in the alopecic area. Dermoscopy showed normal follicular openings with thin vellus hairs over the affected area and terminal hairs in the adjacent scalp. Based on the characteristic history and physical findings, a diagnosis of congenital triangular alopecia was made. Awareness of this condition is important so that it will not be overlooked or misdiagnosed.

Keywords: Triangular; Hair Loss; Frontotemporal; Asymptomatic; Benign

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Introduction

Congenital triangular alopecia, also known as temporal triangular alopecia or Brauer nevus, is a developmental anomaly characterized by an asymptomatic bald patch typically involving the frontotemporal region in a triangular shape [1]. It is a non scarring, non inflammatory and circumscribed form of

alopecia [1]. Awareness of this condition is important so that it will not be overlooked or misdiagnosed [2]. A spot diagnosis can usually be made provided one is familiar with the condition thereby unnecessary diagnostic procedures and inappropriate treatments can be avoided. A review of the topic is therefore in order and is the purpose of the present article.

Illustrative Case

A 3-year-old Chinese girl presented with an 18-month history of a localized triangular area of hair loss in the right frontotemporal area. The lesion was asymptomatic. She was born to a 23-year-old primigravida at term. Cesarean section was performed because of fetal distress. There was no history of trauma. She was otherwise in good health and was not on any medications. The parents were nonconsanguinous and there was no family history of similar lesion. Her 42-year-old father had androgenic alopecia that was first noted 3 years ago.

Physical examination revealed a patch of alopecia 3.5 x 4 cm over the right frontotemporal area, with its apex towards the vertex (Figure 1). The overlying skin was otherwise normal. In particular, there was no scaling, scarring, erythema, atrophy, or induration in the alopecic area. The rest of the examination was normal and there were no dysmorphic features. A hair pull test was negative. Exclamation mark hairs were absent. Dermoscopy showed normal follicular openings with thin vellus hairs over the affected area and terminal hairs in the adjacent scalp.

Based on the characteristic history and physical findings, a diagnosis of congenital triangular alopecia was made. The patient and her parents were reassured of the benign nature of the condition and they opted for no treatment.

Figure 1: A 3-year-old girl with congenital triangular alopecia affecting the the right frontotemporal area.



Discussion

Congenital triangular alopecia is a benign, circumscribed, noncicatricial form of alopecia characterized by a triangular bald area that is generally confined to the frontotemporal area, with the exclusive presence of vellus hair [3]. The condition was first described in 1905 by Raymond Sabouraud [4]. In 1926, the lesion was defined again by Brauer as part of Brauer syndrome [5].

Epidemiology

The exact incidence in the general population is not known. In a review of 6200 randomly selected patients who were seen for the first time in a dermatology clinic in Spain, 7 (0.11%) patients were found to have congenital triangular alopecia [6]. Congenital triangular alopecia occurs in all races and skin types but is strikingly common in Caucasians and rare in blacks [6-8]. The condition is usually observed in children between 2 and 4 years of age [3]. The sex ratio is approximately equal [1, 7, 9].

Etiopathogenesis

The exact etiopathogenesis is not known. Most cases are sporadic [7, 10]. Familial cases have occasionally been reported [1, 11, 12]. Congenital triangular alopecia may reflect mosaicism and may be inherited as a paradominant trait where a postzygotic loss of the wild type allele in a heterozygote state leads to the disorder [5, 7, 11, 13, 14]. Some authors suggest that the condition may result from an unknown stimulus that leads to localized miniaturization of the hair follicles with resulting

regression to vellus hair [14, 15].

Histopathology

Histologically, hair follicles are present in normal numbers but are miniaturized, producing sparse vellus hairs or no hair instead of normal terminal hairs in the affected area [8, 16]. The epidermis and dermis are unremarkable [8]. Inflammatory and/or scarring processes are not observed [7].

Clinical Manifestations

Although the condition is congenital, the alopecia is usually not noticed by the family until the child is 2 years and older [5, 10]. In a review of 54 cases, only approximately one-third of cases were noted at birth [1]. Rarely, the condition may arise in adulthood [17]. Clinically, congenital triangular alopecia presents as an asymptomatic, triangular, and less often, oval or lancet-shaped patch of hair loss [14]. Typically, the hair loss affects the frontotemporal area and rarely the temporoparietal or occipital area [14]. The lesion is circumscribed, nonatrophic, nonscarring, and noninflammatory [5, 18]. The condition is usually unilateral but may be bilateral in up to 20% of cases [5, 6, 16]. For unilateral cases, the left side is slightly more commonly affected than the right side [1, 18].

Congenital triangular alopecia is usually an isolated finding in the majority of cases. The condition may also be a feature of Brauer syndrome, Down syndrome, Turner syndrome, Klippel-Trenaunay syndrome, LEOPARD syndrome (multiple lentiginos, electrocardiographic conduction defects, ocular hypertelorism, pulmonary stenosis, abnormalities of genitalia, retardation of growth and sensorineural deafness), Pai syndrome, cerebellar-trigeminal-dermal dysplasia (Gómez-López-Hernández syndrome), and phakomatosis cesioflammea (phakomatosis pigmentovascularis type II) [5, 10, 13, 14].

Diagnosis

The diagnosis is usually clinical, based on its distinct clinical appearance and typical location. Dermoscopy reveal normal follicular openings, thin vellus hair with varying hair length diversity in the affected area, and normal terminal hair in the adjacent scalp [3]. Biopsy of the lesion is generally not warranted.

Differential Diagnosis

The main differential diagnosis is alopecia areata; many cases of congenital triangular alopecia have been misdiagnosed as

alopecia areata and treated as such [2, 5, 13, 17]. Alopecia areata most commonly manifests as asymptomatic sudden loss of hair in a well demarcated, often round or oval area [19]. The hair loss is usually limited to a single patch, although it can present as multiple patches. The scalp usually has a normal appearance. "Exclamation point hairs" (short broken hairs that are broader at the distal end than at the scalp level) are frequently seen at the periphery of the lesion [19]. Nail involvement, particularly nail pitting may be observed [19]. Other differential diagnosis include physiological shedding of newborn hair, tinea capitis, aplasia cutis congenita, traction alopecia, trichotillomania, pressure alopecia, lichen planopilaris, pityriasis amiantacea, and androgenic alopecia [14, 18, 20].

Complications

Although the lesion is benign and asymptomatic and does not cause physical disabilities, the cosmetic concern can be problematic.

Prognosis

Congenital triangular alopecia is a nonprogressive disorder but the lesion tends to persist throughout life [3, 8].

Management

Treatment is usually not necessary other than reassurance of the benign nature of the condition and watchful observation. For those patients who prefer treatment for cosmesis, treatment options include surgical excision of the lesion and follicular unit hair graft transplantation [9, 15]. A case report suggests that topical application of minoxidil might be useful in the treatment of congenital triangular alopecia [21]. Further studies are necessary before such treatment can be recommended.

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