Dermoscopic Findings of Scalp Aplasia Cutis Congenita

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Keywords

Aplasia cutis · Scalp diseases · Alopecia

Abstract

Aplasia cutis congenita (ACC) is a rare disease characterized by congenital absence of skin, affecting preferentially the scalp. Diagnosis is made clinically; however, recent studies have shown that dermoscopy can be a useful tool for the diagnosis and differentiation from sebaceous nevus. The clinical findings include a shiny atrophic alopecic patch associated with dermoscopic findings of absent follicular openings, thicker vessels and a distinct collar hypertrichosis. We report 2 cases of alopecia presenting from birth. At dermoscopy, the absence of follicular openings and the increase in the caliber of vessels led us to establish the diagnosis of ACC.

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Introduction

Aplasia cutis congenita (ACC) is a rare malformation [1] characterized by a congenital absence of small or extensive areas of skin usually affecting the scalp [2]. As a result of its causal heterogeneity, the clinical appearance of ACC at birth is extremely variable, ranging from an erosion or deep ulceration, through a scar to a discrete ovoid defect covered by a membrane [2, 3]. There are 9

subtypes of ACC; the one referred to as membranous aplasia cutis is the most common [3]. This article aims to report the main dermoscopic findings of 2 cases of membranous aplasia cutis on the scalp.

Case Report

We describe 2 cases of membranous aplasia cutis on the scalp in a boy and a girl born by cesarean section without complications. They were referred to our dermatology clinic with an alopecic patch located in the vertex of the scalp since birth. The physical examination showed a single alopecic area measuring 8 cm in diameter in the boy (Fig. 1) and 3 cm in the girl (Fig. 2), with yellow dots, and a membrane showing underlying vessels. Dermoscopy revealed the absence of follicular structures and an increase in caliber and quantity of vessels (Fig. 3, 4). These findings led to the diagnosis of ACC.

Discussion

ACC has an incidence of approximately 1/10,000 births [1, 3]. It is usually located on the scalp (in 85%) [2, 3] but can occur anywhere on the body including the face,

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Fig. 1. Single alopecic area measuring 8 cm in diameter.

Fig. 2. Single alopecic area measuring 3 cm in diameter.

Fig. 3. Absence of follicular structures and increase in caliber and quantity of vessels.

Fig. 4. Absence of follicular openings and visible vascular network.

Table 1. Dermoscopic features of sebaceous nevus and ACC

Sebaceous nevus	ACC on the scalp
Yellow dots not associated with hair follicles	<p>Margin of the alopecic plaque:</p> <ul style="list-style-type: none"> – hair collar sign: hair shafts are arranged radially, forming a ring of hypertrichosis – elongated and pigmented hair bulbs visible through translucent epidermis <p>Center of the alopecic plaque:</p> <ul style="list-style-type: none"> – absence of follicular openings – lack of skin appendages – translucent appearance – visible vascular network, corresponding to skin atrophy in this area

trunk, and extremities. ACC may occasionally be associated with other anomalies [3]. Most ACC lesions occur on the region of the fontanel, lateral to the midline, and extended to superior sagittal sinus, and also in the parietal and retroauricular regions [1]. The defects usually measure 1–2 cm in diameter, although the size can vary from 0.5 to 10 cm, and can achieve different depths, even involving the periosteum, skull, and dura mater [3]. The cases reported here presented with an area of shiny alopecia covered by translucent and vascularized skin, resembling a parchment [4]. The dermoscopic images revealed that at the hair-bearing margin, the hair shafts were arranged radially, forming a ring of hypertrichosis (“hair collar sign”) [5]. Elongated hair bulbs with dark pigmented proximal ends, typical of the anagen phase, were visible through the translucent epidermis [5]. The midpart of the lesion showed no follicular openings [4, 5]. A vascular network was visible, which was due to the skin atrophy in this area [4, 5]. The constellation of these findings is not detected in other scalp disorders.

Sebaceous nevus may also present as a smooth patch of alopecia at birth, but it shows distinct dermoscopic features (Table 1). Histology is rarely performed, and it shows absence of epithelium and adnexal structures and a thin layer of collagen in the dermis. Systemic associations are excluded by imaging tests and electroencephalography. Due to several distinct clinical subtypes, the lesion repair may involve excision with subsequent reconstruction using bone grafts and muscle flaps [1, 3].

Statement of Ethics

Patient consent was obtained for publication of this report.

Disclosure Statement

The authors certify that they have no affiliations with or involvement in any organization or entity with any financial interest.

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