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CASE REPORT

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Congenital vertex ulceration in a 3-day-old newborn

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ABSTRACT

Aplasia cutis congenita (ACC) is a rare congenital malformation characterized by localized or generalized absence of the skin. It can occur on any part of the body but primarily on the scalp. Varying degrees of severity are observed, ranging from isolated skin absence to full-thickness defects involving bone and underlying nerve structures. These defects can impact the child's vital and functional prognosis. Management depends on the degree of skin involvement and the presence of underlying issues, mainly neurological. We report the case of a 3-day-old newborn with cutaneous aplasia of the vertex without underlying bone involvement, who was successfully treated with targeted wound healing.

KEYWORDS

Skin ulceration; Aplasia cutis congenita (ACC); Newborn; Congenital; Prognosis

ARTICLE HISTORY

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Introduction

Aplasia cutis congenita (ACC) is a rare congenital malformation characterized by localized or generalized congenital absence of the skin that mainly appears on the scalp. Varying degrees of severity can be seen, ranging from isolated absence of the skin to full-thickness defects involving bone and underlying nerve structures. It can be isolated or part of a heterogeneous group of syndromes [1].

Herein, we present a case of isolated ACC in a 3-day-old newborn.

Case Presentation

Three-day-old, full-term male infant born by vaginal delivery with good adaptation to extrauterine life, weighing 3000g, was sent to our establishment for an opinion on an extensive tissue defect on the vertex. This defect was observed 24 hours prior to the call from the Neonatology department. There was no maternal use of medication except Ibuprofen, alcohol, or drugs, and no history of infection during pregnancy. There was no family history of skin problems or genetic abnormalities.

A dermatological examination revealed an ulcerated midline area on the vertex, measuring 12 cm by 3 cm, with an erythematous border and a crusted surface (Figure 1a). There was no bony defect on palpation nor blood or spinal fluid outflow. The child was otherwise normal, with no other cutaneous lesions or abnormalities of limbs or digits. He was alert and active, with normal vital signs. The physical examination revealed no abnormalities, with all neurological, cardiological, and musculoskeletal systems functioning within normal parameters. Cerebral exploration, including cranial radiography and transfontanellar ultrasound, was normal. A neurosurgical opinion was requested, and the decision was to put the patient under local care and surveillance. The lesion evolved toward healing after twice-daily local care with greasy tulle (Figure 1b). Scarring alopecia of the vertex and a central erythematous area remained (Figure 1c).

Figure 1. (a). Ulcerated midline area with crusted surface. (b). Clinical aspect after one month. (c). Clinical aspect after eight months: Scarring alopecia of the vertex.

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Discussion

ACC is a rare congenital condition characterized by localized or generalized absence of the skin, first described by Cordon in 1767. It is often observed during the first days of life as a single lesion on the vertex on the median line, but reports of lesions appearing on different locations, such as the face, trunk, abdomen, and limbs, have been documented.

Typically, the lesion is small, well-circumscribed, and may exhibit various aspects such as circular, oval, linear, or stellate shapes. Following its healing, the lesion gives way to cicatricial alopecia [2]. Diagnosis of ACC is clinical, and paraclinical examinations aim to identify associated abnormalities, mainly neurological, ophthalmological, and skeletal.

Frieden Proposed in 1986 [3] a classification modified by Sathishkumar in 2020 [4], consisting of nine groups based on the location of lesions and the associated abnormalities leading to various sub-entities, aiding daily practice. Our patient had isolated cutaneous involvement, and we were unable to categorize it into a specific type according to Frieden's classification.

Numerous authors have postulated the precise mechanism underlying ACC, positing that factors such as intrauterine issues, thrombotic events, and vascular alterations may contribute to the development of this condition. Additionally, specific teratogens administered during pregnancy, such as cocaine, methotrexate, benzodiazepines, and valproic acid, have been suggested as potential triggers for ACC. Furthermore, Frieden classification designates Type 7 as drug-induced ACC [3].

The management of ACC remains controversial, with options including nonintervention, conservative treatments, and surgical intervention. Conservative treatment is typically recommended for small lesions, especially if no underlying defects are identified, like was the case of our patient. Nevertheless, close monitoring is recommended to detect infectious and hemorrhagic complications. However, in the presence of osseous or dural involvement, surgical treatment should be initiated early to prevent complications [5].

Conclusions

Despite its exceptional nature, ACC deserves recognition as it can pose a life-threatening prognosis for newborns. Its treatment remains controversial, with proponents of emergency surgery and advocates for medical intervention.

Disclosure statement

No potential conflict of interest was reported by the authors.

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