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CASE REPORT

Aplasia cutis congenital. A case report

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Abstract

Aplasia cutis congenita (ACC) is a rare congenital disorder that commonly affects the scalp. In this disease, some parts of the skin with or without underlying structures were not formed at birth. The aplastic lesion always presents as a solitary lesion; however, multiple lesions also were reported. In more severe cases, deeper structures, such as the subcutaneous tissue, bone, and dura, can be affected. In this study, we report a case of ACC and its management.

Key words: Aplasia cutis congenita, Ectodermal dysplasia, Nonsyndromic

Introduction

Aplasia cutis congenita (ACC) is a rare congenital disorder that commonly affects the scalp; however, the involvement of other parts of the body has been reported in this regard (1).Some parts of the skin with or without the underlying structures were not formed at birth which is considered the characteristic of the disease (2). The aplastic lesion always presents as a solitary lesion; nevertheless, multiple lesions were also reported. In more severe cases, deeper structures, such as the subcutaneous tissue, bone, and dura, can be affected (2).In this study, we report a case of ACC and its management.

Cases

A 5-day-old neonate was referred to a surgery clinic for a follow-up of an ulcer that was located in the scalp area. The case was born through a normal vaginal delivery from a gravida-2 para-1 mother. At first, theneonate had no congenital abnormality. There was no history of drug consumption or disease during pregnancy. The grandfather of the patient has had such a history and alopecic area on the scalp.

In the physical examination of the case, there was a 4 × 4 cm ulcer (i.e., aplasia cutis) in the vertex area at the right paramedian location (Figure 1). There was some exudate on the surface

Figure 1: Aplasia cutis congenita of scalp

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of the ulcer; however, the periosteum was intact and there was no bony defect. The patient was evaluated by our pediatrician colleague, and there was noother congenital abnormality. The wound was conservatively managed with petrolatum gauze and daily dressing changes. In addition, the wound was completely epithelialized after 50 days.

Discussion

The ACC is a rare congenital cutaneous disorder that occurs commonly in the scalp area and sometimes in the trunk or extremities. The prevalence of this disorder has been reported in various studies within a range of 0.5-1% per 10,000 live births (1). The genetic pattern of transmission can be sporadic, autosomal recessive, or dominant. Friedenclassified the aplasia cutis into nine groups based on the presence of congenital disorders and location of skin involvement (1).

A wide variety of mechanisms has been proposed for the pathogenesis of the disease, including maternal disease during pregnancy, teratogenicity drugs, intrauterine trauma, and ectodermal or neural tube closure defects. There were several reports about the association of ACC with dermal melanocytosis that can explain the existence of neural tube closure defects as an etiologic factor (2).

The lesion appears as a round or oval skin defect or ulcer, usually adjacent to the midline in the vertex area. It is often isolated; nevertheless, multiple lesions have also been reported. The disease usually involves the skin; however, it can also affect deeper tissues, such as the bone and even dura. Sometimes, a hair collar can be present around the lesion; in these cases, the probability of the involvement of deep tissues, especially the central nervous system, increases (3).In cases with the involvement of deeper tissue, such as the bone and dura, in addition to the scalp, it can lead to infection, bleeding, seizure (1), sagittal sinus hemorrhage, or thrombosis (4) (5).

More extensive cases of ACC with other anomalies have also been reported (6).In small ACCs, themanagement of the wound consists of conservative therapy and prevention of wound complications, such as bleeding and infection. In severe cases, it may be necessary to cover the defect with a skin graft or flaps (7). The application of silver-containing dressings, such asActicoat, and coverage of the lesion with allograft until the completion of epithelialization were also reported (2).

Conclusions

There are different forms of the ACC. It can cause serious complications such as bleeding from the sagittal sinus.So although it is a rare disease, physiciansmust to be familiar with its management.

Conflict of Interest

There is no conflict of interest to be declared.

References

1. Browning JC.Aplasia cutis congenita: approach to evaluation and management. DermatolTher. 2013;26(6):439-44. PMID: 24552406DOI: 10.1111/dth.12106
2. Lonie S, Phua Y, Burge J. Technique for management of aplasia cutis congenita of the scalp with a skin allograft. J Craniofac Surg. 2016;27(4):1049-50.PMID: 27171959DOI: 10.1097/SCS.0000000000002610
3. Takayama E, Harada A, Ikura Y, SetoH.Two cases of aplasia cutis congenita with hair collar signs and macrophage hyperplasia.JDermatol. 2019; 46(8): 734-8. PMID: 31180149DOI: 10.1111/1346-8138.14946
4. Johnson R, Offiah A, Cohen MC. Fatal superior sagittal sinus hemorrhage as a complication of aplasia cutis congenita: a case report and literature review. Forensic Sci Med Pathol. 2015; 11(2):243-8.PMID: 25614301DOI: 10.1007/s12024-014-9645-5
5. Brzezinski P, Pinteala T, Chiriac AE, Foia L, Chiriac A. Aplasia cutis congenita of the scalp--what are the steps to be followed? Case report and review of the literature. An Bras Dermatol. 2015; 90(1): 100-3.PMID: 25672305DOI: 10.1590/abd1806-4841.20153078
6. Zhou J, Zheng L, Tao W. Systemic aplasia cutis congenita: acase report and review of the literature. Pathol Res Pract. 2010; 206(7):504-7.PMID: 20188489DOI: 10.1016/j.prp.2009.12.011
7. Arslanca SB, Arslanca T, Koç A. Aplasia cutis congenita: a case report. J ObstetGynaecol. 2019; 39(6):860-1.PMID: 31023179DOI: 10.1080/01443615.2019.1581753

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