

Extensive Aplasia Cutis Congenita: A Case Report

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ABSTRACT

Aplasia cutis congenita (ACC) is a rare congenital disorder characterized by localized or widespread skin absence. The most common site is the scalp and the main cause is unknown. Here we report a case of extensive ACC with scalp involvement. A female infant with an Apgar score of 9 and weighing 3406 gr was born by cesarean section in Besat Hospital, Sanandaj, Iran. A lesion measuring 5×10 cm with highly prominent vessels was on the forehead and parietal bone without scalp, skull, or bone tissue leading to the brain tissue being covered with a thin membrane. Other clinical examinations were normal. The newborn suffered from a ruptured aneurysm and intracerebral hemorrhage and underwent pharmacologic treatment. She died after 21 days of hospitalization. In order to prevent mortality in infants with skin defects, infection control is a priority. Antibiotics could be administered in cases with extensive lesions to prevent neonatal sepsis and improve scar tissue formation. Moreover, restorative measures should be taken if necessary.

Keywords: Aplasia, Congenital, Cutis, Lack of scalp



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Introduction

Aplasia cutis congenita (ACC) was first reported by Corden in 1767 (1). This rare abnormality is characterized by localized or widespread absence of skin at birth (2). The most common site of ACC is the scalp followed by the forearm, knees, sides of the trunk, and neck (3). The incidence of this disorder is 1-3 per 10,000 live births (4). Although the main cause is unknown, diverse factors, such as placental infarction, teratogens, intrauterine infection, trauma, incomplete neural tube closure, and amniotic membrane adhesion are involved (5). This congenital anomaly is more common in female infants than male newborns (1). Small local lesions are treated conservatively, while larger lesions require surgery (6). We report a case of extensive ACC with scalp involvement.

Case Report

A female infant with an Apgar score of 9 and weighing 3406 gr was born to a mother aged 23 years,

gravidia 1, and gestational age of 40 weeks and 2 days in Besat Hospital, Sanandaj, Iran. The mother underwent a cesarean section due to a fetal facial anomaly. The mother had no history of current illness or family history. Maternal prenatal care had been regular and she had been visited six times representing any particular problems. During pregnancy, she took ferrous sulfate tablets and multivitamin plus mineral capsules.

Clinical examinations performed at birth revealed a round to oval-shaped serrated lesion measuring 5×10 cm with highly prominent arteries on a part of the frontal and parietal bones. There was no scalp, skull, or bone tissue in this area and the brain tissue was covered with a thin membrane. Clinical examinations showed a healthy infant without any other abnormalities. The lesion was covered with vaseline to prevent possible complications. The one-day-old newborn was sent to Mofid Hospital, Tehran, Iran for advanced surgery. The infant had a ruptured aneurysm and intracerebral hemorrhage in Mofid Hospital and was treated with

medication without surgery. The infant died after 21 days of hospitalization.



Figure 1. Image of aplasia cutis congenita. This image shows a lesion measuring 5×10 cm on the forehead and parietal bone.

Discussion

The ACC is a congenital anomaly affecting the scalp in 80%-90% of cases (7). Approximately 15%-30% of cases are associated with defects in the skull or sclera (8). Maternal serum alpha-fetoprotein measurement is used for diagnosis in the first month of pregnancy (9). However, the diagnosis is usually made after birth based on clinical findings. In addition, ACC can be detected by ultrasound before birth (10). In the present case, ACC was reported after birth.

Most skull defects in the midline are on the roof of the skull, which may be associated with the lack of scalp, skull, and sclera (11). Histological examinations of the skin showed no natural structures, such as hair follicles, sebaceous glands, sweat glands, and collagen fibers in the dermis (12). Although skin lesions most often occur spontaneously, they can also be autosomal dominant or autosomal recessive (13). The main etiology is still unknown. However, the two factors of genetics and environment play role in the occurrence of this disorder (5).

The mortality rate due to ACC has been reported as 12.5%-55% with most deaths occurring in the first eight days after birth. However, deaths and near-death events may take place from weeks to months after birth (7). The high mortality rate could be attributed to sagittal sinus bleeding, secondary infection, meningitis, sagittal sinus thrombosis, or a direct result of other serious congenital defects associated with ACC. An important life-threatening factor is a sagittal sinus bleeding, which is

the leading cause of death in 36% of cases (14). It is of high importance to prevent such fatal complications through fast and effective management.

Conservative treatments, including regular wound cleansing and dressings in combination with systemic antibiotics, are applied for small local lesions (6, 14). Wider lesions larger than 15 cm² require surgical treatment (15). Surgical approaches entail skin grafting, bone grafting, as well as local and free flaps (4).

Conclusion

The ACC is a rare congenital disease, the cause of which is still unknown. In order to prevent mortality in infants with skin defects, it is better to prevent infection first. In cases with extensive skin lesions, antibiotics could be used to prevent neonatal sepsis and improve the formation of scar tissue. If necessary, restorative measures should be taken. The present study showed that the treatment applied for this infant with ACC was not successful and she died at the age of 21 days.

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Conflict of Interest

There was no conflict of interest between the authors.

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