Aplasia Cutis Congenita Type V: a case report and review of the literature

Benjamin M. Perry, DO, PGY-3; Cory Maughan, DO
Western University of Health Sciences- Silver Falls Dermatology

Abstract

Aplasia cutis congenita is a relatively rare congenital anomaly that most commonly occurs as a solitary cutaneous defect on the scalp. Depth of involvement varies and involvement of deeper calvarium and dural structures can be seen in more severe cases. Multiple classification systems have been devised with the Frieden Classification System being the most widely adopted. Using this system, we describe a patient that developed Type V aplasia cutis congenita (ACC) with associated fetal papyraceous. The child healed remarkably well with application of petrolatum impregnated gauze and topical silver sulfadiazine twice daily for approximately 4 weeks. The child was noted to have no significant contractions or complications at 6 months and 1 year follow-up exams.

Case

A 37.4/7’d male was born to a G2P1 mother after a complicated, monochorionic twin pregnancy. At 16 weeks gestation mother experienced fetal demise of one fetus. The surviving fetus developed hydrops fetalis and severe anemia requiring a fetal blood transfusion. At the time of delivery the male infant was noted to have significant absence of skin on the lateral torso and vertex scalp (Image 1). There was also apparent stellate scarring on the elbows, knees and hips. This was presumed to have been areas of aplasia cutis that had begun the healing process in utero. The remaining open wounds on the torso were initially dressed with petrolatum impregnated gauze and then a regimen of topical silver sulfadiazine was implemented (Image 2). The parents applied these dressings twice daily for approximately 4 weeks. At two weeks postpartum the infant developed a fever of unknown origin. Bacterial cultures from the healing wounds were taken and returned negative. At 4 weeks postpartum the areas of involvement demonstrated significant healing (Image 3). No significant contractions or complications were noted with routine exams at 6 months, 12 months and 2 years (Image 4).

Discussion

In contrast to the other types of Aplasia cutis congenita, Type V most commonly affects the trunk and is often symmetric (Table 2). 1,4 ACC with fetus papyraceus is typically observed in association with monochorionic twin pregnancies (90% of cases). 1 It has been noted that if demise occurs prior to 34 weeks gestation, ACC typically will develop on the trunk versus the extremities with fetal demise after 14 weeks. 6,7 Following twin fetal demise, some believe that an ensuing transient hypoplasia leads to ischemia of watershed areas of the skin 8,9 while others have suggested that thrombosis formation in the setting of disseminated intravascular coagulation of the dying fetus embolizes to the healthy twin. 9 An exact pathogenesis has not been proven but it is almost certain that a transient vascular process is responsible for the clinical findings observed in type V ACC.

One of the greatest risks in type V ACC is neonatal development of infection. As a result, early intervention is directed towards minimizing this risk. Depending on the size of the defect, treatment using surgery, skin grafts (flaps) or conservator wound care is often employed. Surgical risks include potentially fatal hemorrhage, infection, and anesthesia complications. Supportive wound care carries risk of hemorrhage, sagittal sinus thrombosis, wound bed necrosis, and infection. Because both carry significant risk of complications a definitive consensus for treatment has yet to be achieved. Most experts agree that conservative wound care is appropriate in the majority of cases with size of defect and location the most important factors to consider. General skin grafting is often reserved for defects larger than 2-4 cm; particularly with scalp defects. 9, 10 One review of 11 cases of type V ACC associated with twin loss demonstrated successful reepithelialization and later scar formation in 10 of the cases. 9 In the remaining case, skin graft became necessary due to the development of bacteria. 9 Conservative wound care has found similar success in other cases including our patient. As a result, we feel that any treatment algorithm for type V ACC should rely on conservative wound care and infection prevention with more invasive methods utilized if complications arise. A basic regimen could include sequential application of silver sulfadiazine, petroleum gauze, dry gauze and a self-adherent wrap with dressing changes twice daily. 9 Use of antibiotic ointment can be considered in place of silver sulfadiazine for any concern.

References