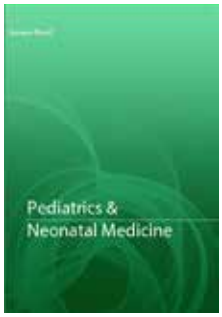
Pediatrics and Neonatal Medicine

Case Report



A Case Report: Aplasia Cutis Congenita Secondary to Fetus Papyraceus

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Introduction

Aplasia cutis congenita (ACC) is a rare skin condition that presents with absence of fetal skin at the time of birth. It presents most commonly with an isolated lesion on the scalp. Non-scalp lesions usually involve the trunk and lower extremities and are often bilateral and symmetrical [1] (Figure 1). Fetus papyraceus (FP) is a known, yet rare cause of ACC. The incidence of ACC in association with FP is estimated to be 1 in 12,000 live births [2]. FP is a condition that occurs in multiple gestations where one fetus is demised, usually in the second trimester. The non-viable fetus becomes a mummified, parchment-like fetus [3]. The cause of ACC secondary to FP is unknown but theories have been proposed. The vascular theory suggests the death of one fetus allows for passage of thrombogenic agents to the living twin through placental anastomosis. This, in turn, activates the coagulation cascade resulting in disseminated intravascular coagulation (DIC). DIC results in ischemic changes to the developing skin and, rarely, to the underlying tissue [4-8]. Another proposed mechanism is when one twin dies it leads to a drastic relaxation of the vasculature in that twin causing blood to shunt to the low resistance system leading to acute hypovolemia in the living twin. This results in ischemia of the skin and other organs [5,6,9,10]. The last proposed theory is disturbance of feto-fetal transfusions [7,10].

This theory states ischemia occurs in both twins and only one twin can survive with aplastic scarrin g due to restricted blood flow [5,11]. As ACC is primarily limited to the skin, it is not detectable on ultrasound and is usually diagnosed at time of delivery. We present a case of symmetric aplasia cutis congenita secondary to fetus papyraceus.

Case Report

29 year old G5P2022 with dating ultrasound demonstrating a monochorionic/diamniotic twin gestation at 11 weeks gestation. A referral to Maternal-Fetal Medicine (MFM) was placed with a repeat ultrasound demonstrating one viable fetus consistent with dating at 13 weeks gestation and one non- viable fetus measuring 12 weeks gestation. The remainder of her antepartum course was unremarkable. Repeat ultrasounds during the pregnancy were unremarkable with the demised fetus noted on the maternal left. She underwent an uncomplicated spontaneous vaginal delivery at term. At the time of birth the neonate had extensive bilateral skin lesions on the abdomen and legs (Figures 2 and 3).

The neonate was admitted to the neonatal intensive care unit where dermatology, and plastic surgery were consulted. The infant received a chest radiograph and an ultrasound of the head and abdomen. Imaging demonstrated appropriate bone development and did not identify any underlying pathology. Fetal echocardiogram demonstrated normal cardiac anatomy. Dermatology recommended conservative management of the skin lesions with xeroform petrolatum dressings with daily changes. In addition, they recommended topical mupirocin ointment for infection prophylaxis. Once discharged, plain white petrolatum and telfa non-stick dressings were used. Plastic Surgery agreed with

Dermatology's recommendations to pursue conservative management with the addition of Promogran Prisma dressing to improve wound healing and decrease dressing changes to every three days. Plastic Surgery also recommended a follow up in 6-12 months to determine the need for scar revision. It was noted that there were signs of epithelialization at the edges of the atrophic area on day of life one.

Ophthalmology reported a normal ophthalmic exam and recommended routine vision screening.

The placenta was sent for analysis and appeared to be within normal limits measuring small with a weight of 410 g and attached fetal twin identified within the fetal membrane.

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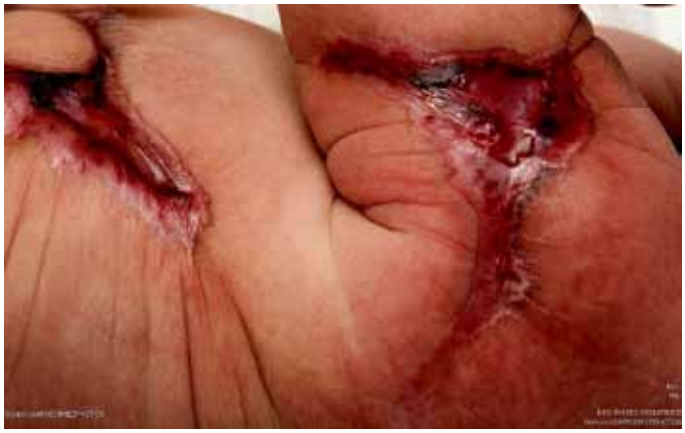

Figure 1. *Lesions on the abdomen and trunk at time of birth.*

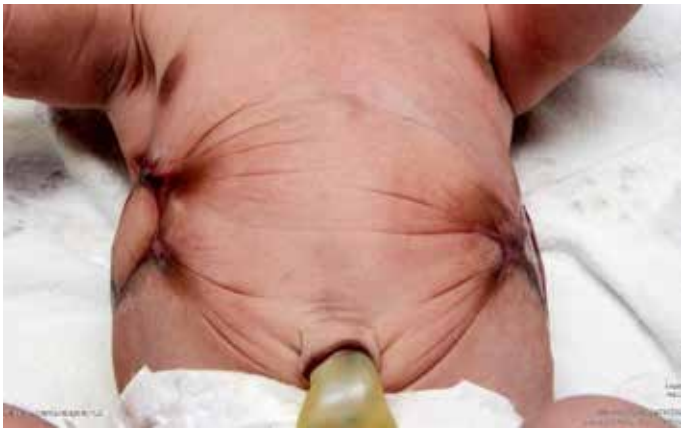

Figure 2. *Symmetrical truncal lesions noted at time of birth.*

Figure 3. *Aplastic stellate lesion on the infant's abdomen.*

Whole genome oligonucleotide array was performed and was negative for other causes of ACC.

The infant was discharged on day of life six with close follow up with Pediatrics and Dermatology. At follow up appointments, the infant appeared to be growing appropriately and the atrophic lesions were healing well. No scar revision was required.

Discussion

ACC-FP is a condition in multiple gestations where one fetus is lost and becomes mummified and parchment-like; the surviving twin has areas of absent skin at the time of delivery. FP usually occur s in the early second trimester [4,5]. If the demise happens earlier, the fetus will be completely reabsorbed, as in the case of a vanishing twin syndrome [5,6].

ACC is not detectable on ultrasonography and though rare it is usually found at the time of birth. There are no evidence based guidelines on management of ACC-FP [12,13]. Treatment is generally tailored to the extent of the disease. For large, non-scalp lesions like in our case of ACC-FP, the defects can usually be managed conservatively [10,14]. Early diagnosis of the condition by the obstetrician or neonatal provider at the time of birth is imperative for proper management. Conservative options include adhering and non-adhering dressings with emollient solutions. Topical antibiotics agents may be added. For small scalp lesions and large scalp lesions without bone involvement, it can also be managed conservatively. Large scalp lesions and skull defects involving underlying bone may require surgical intervention with skin grafts or skin flaps for soft tissue coverage. Rarely, reconstructive cranioplasty with split-rib grafts or cranial vault splitting may be indicated. Usually, if these procedures are indicted, they are performed at between 2 to 4 years of age to allow spontaneous bone growth [12,14].

For most infants affected by ACC-FP, the prognosis is good. Most cases resolve with conservative management within the first weeks to months. For large lesions involving the underlying bone, vascular and nervous system, the mortality rate is as high as 50 percent. This is often due to increased risk of hemorrhage, thrombosis, and infections. Early recognition will allow for appropriate care of the neonate and counseling of the parents. Our case report aims to educate the generalist and the neonatal provider in this rare condition and the management options available.

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Conflicts of Interest: None

Acknowledgements: None

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