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Case History

Neonate with Aplasia Cutis Congenita associated with Foetus Papyraceus

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Introduction

Aplasiacutis congenita (ACC) is a rare dermatological condition seen in newborns characterized by localized congenital absence of skin. It is subdivided in to many categories out of which ACC associated with foetus papyraceus presents with multiple symmetrical involvement of scalp, chest, flanks, axillae or extremities. We report a case of ACC in a newborn of a twin pregnancy, where the other twin was diagnosed as a 'vanishing twin' with foetal demise in the early 2nd trimester leading to foetus papyraceus.

Case report

A Sri Lankan baby boy weighing 3100g was delivered at term by an elective Caesarean section to healthy non-consanguineous parents. The antenatal ultra sound scan (USS) done at 7 weeks of period of gestation had revealed a mono-chorionic di-amniotic twin pregnancy. However a repeat scan at 16 weeks had revealed spontaneous demise of one twin leading to 'vanishing twin' phenomenon. The anomaly scan done at 20 weeks did not reveal any foetal anomalies in cardiac, renal or gastrointestinal systems of the surviving twin. The pregnancy was not complicated with maternal diabetes mellitus or hypertension.

A foetus papyraceus (Figure 1) was delivered along with the surviving twin who was noted to have lesions with absent skin in the lateral aspects of the trunk and extensor surfaces of lower extremities (Figure 2). The lesions were well demarcated, symmetrical, and involved a large area of bilateral chest, abdominal wall and thighs. These areas consisted of stellate shaped transparent non inflammatory membranes. There was no associated bleeding or herniation from the skin defects. The baby did not have any dysmorphic features and the skin over the rest of the body appeared normal. The hands and feet appeared normal with no evidence of nail dystrophy or clubbing.

A clinical diagnosis of aplasia cutis congenita associated with foetus papyraceus was made. The baby passed meconium on day one itself and there was no evidence of gastrointestinal atresia. 2-Dimensional echocardiography and ultrasound scan of the brain, abdomen and kidneys did not reveal any structural anomalies.

The skin lesions were treated with local application of Fusidic acid and barrier nursing to prevent infection. Dermatological and plastic surgical referrals were arranged and a collective decision was made to manage the lesions conservatively without active intervention. The baby was followed up periodically every week to assess the lesions and spontaneous resolution of the lesions was seen (Figure 3).

Discussion

Aplasia cutis congenita (ACC) is characterized by multiple congenital skin defects, of which 84% are located in the scalp1. Frieden has categorized ACC into 9 distinct subtypes, each characterized by the location, pattern of skin absence, associated malformations, and the mode of inheritance2,3. Although many cases of ACC has been recorded, only 50 cases of ACC associated with foetus papyraceus has been reported up to now6,7.

According to Frieden's classification, ACC associated with foetus papyraceus is classified as 'Type 5.' In this subtype, the lesions are usually large with linear or stellate morphology and distributed bilaterally and symmetrically in the scalp, chest, flanks, axillae or extremities. The common associations in this subtype are gastrointestinal atresia, intracranial haemorrhage, renal cortical necrosis, cardiac and arterial anomalies3. However the baby in this case report did not have any of the above abnormalities.

The precise aetiology of ACC is yet to be determined. Some postulate that midline lesions are due to incomplete closure of neural tube defect, while lateral membranous lesions are a result of incomplete closure of embryonic fusion lines. Type 5 ACC is thought to be due to vascular insufficiency resulting from either thromboplastic material from a foetus papyraceus or due to placental insufficiency2,8. The baby in this case report had clear evidence of foetus papyraceus. Other possible aetiologies include intra uterine infection, vascular coagulation defects, maternal ingestion of certain drugs and amniotic membrane adherence1,2.

There are reports of antenatal identification of ACC by ultrasonography by the absence of the usual strong signal which is generated from normal skin4. But most cases are diagnosed after birth. Early diagnosis may be helpful in antenatal counseling of parents. The optimal management of non-scalp ACC has not been well established. Although most small lesions heal spontaneously, larger lesions with significant deformity may need surgical excision later on5. Very large lesions including stellate scalp lesions may need early surgery and skin grafting3. There are reports of defects as large as 8*12 cm which has been successfully managed conservatively5. Defects associated with fetus papyraceus are known to heal well leaving hypo-pigmented scars2,3. They usually do not require surgical intervention5. Local application of antimicrobial agents and petroleum gauze in affected areas and the use of dry wraps may prevent infection, electrolyte imbalance and ulceration.

Figure 1: fetus papyraceus

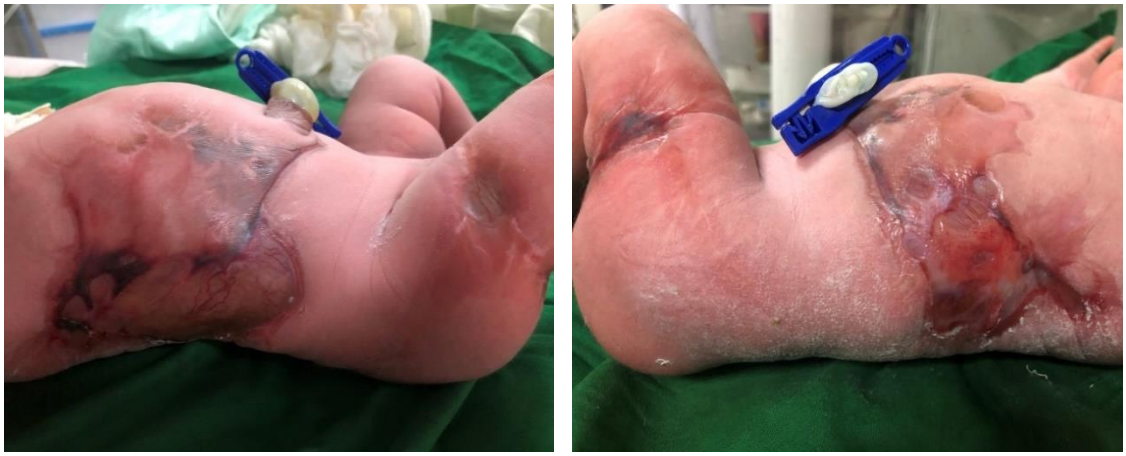

Figure 2: Bilateral symmetrical skin lesions noted on day 1

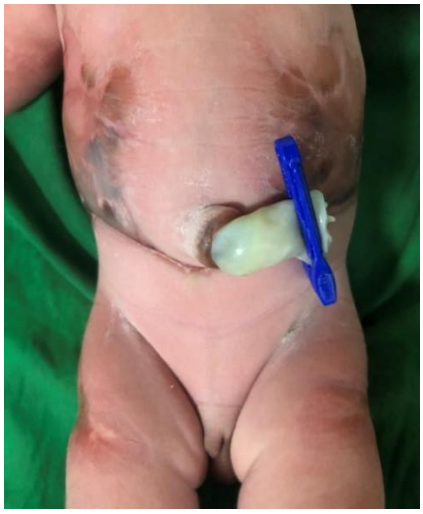

Figure 3: Spontaneously healing skin lesions

References

1. Mannino FL, Jones KL, Benirschke K. Congenital skin defects and fetus papyraceus. The Journal of Pediatrics [Internet]. Elsevier BV; 1977 Oct;91(4):559–64. Available from: http://dx.doi.org/10.1016/s0022-3476(77)80502-7

2. Moss C, Shahidullah H. Naevi and other Developmental Defects. Rook's Textbook of Dermatology [Internet]. Wiley-Blackwell; 2010 May 6;1–107. Available from: http://dx.doi.org/10.1002/9781444317633.ch18

3. Tempark T, Shwayder TA. Aplasia cutis congenita with fetus papyraceus: report and review of the literature. International Journal of Dermatology [Internet]. Wiley; 2012 Nov 21;51(12):1419–26. Available from: http://dx.doi.org/10.1111/j.1365-4632.2012.05545.x

4. Liu F, Chen X, Tu R, Liu S. Prenatal diagnosis of aplasia cutis congenita of the trunk. International Journal of Dermatology [Internet]. Wiley; 2012 Dec 11;53(10):1269–71. Available from: http://dx.doi.org/10.1111/j.1365-4632.2012.05732.x

5. STARCEVIC M, SEPEC MP, ZAH V. A Case of Extensive Aplasia Cutis Congenita: A Conservative Approach. Pediatric Dermatology [Internet]. Wiley; 2010 Aug 27;27(5):540–2. Available from: http://dx.doi.org/10.1111/j.1525-1470.2010.01266.x

6. Mazza JM, Klein JF, Christopher K, Silverberg NB. Aplasia Cutis Congenita in a Setting of Fetus Papyraceus Associated with Small Fetal Abdominal Circumference and High Alpha-Fetoprotein and Amniotic Acetylcholinesterase. Pediatric Dermatology [Internet]. Wiley; 2013 Oct 11;32(1):138–40. Available from: http://dx.doi.org/10.1111/pde.12228

7. Üstüner P, Dilek N, Saral Y, Üstüner I. Coexistence of aplasia cutis congenita, faun tail nevus and fetus papyraceus. Journal of Dermatological Case Reports [Internet]. SpecjalisciDermatolodzy; 2013 Sep 30;7(3). Available from: http://dx.doi.org/10.3315/jdcr.2013.1148

8. Qureshi UA, Ahmed N. Type V aplasia cutis congenita. Annals of Saudi Medicine [Internet]. King Faisal Specialist Hospital and Research Centre; 2010 Mar;30(2):171–2. Available from: http://dx.doi.org/10.4103/0256-4947.60530

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