Picture Stories

Aplasia cutis congenita of the trunk

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Sri Lanka Journal of Child Health, 2017; **46**(2): 190-191
DOI: http://dx.doi.org/10.4038/sljch.v46i2.8282
(Key words: Aplasia cutis, fetus papyraceus)

Introduction
Aplasia cutis congenita (ACC) is a rare condition characterized by localized absence of skin and sometimes subcutaneous tissues[1]. ACC may occur anywhere in the body but in 84% the defect is found in the scalp, often as a solitary lesion in the midline vertex[2].

Case report
A term, baby girl was delivered by spontaneous vaginal delivery at Castle Street Hospital for Women with gestation appropriate growth parameters. She was the second child of non-consanguineous healthy parents after 8 years of secondary subfertility. Older sibling is a healthy 8 year old boy. Pregnancy was diagnosed to be monochorionic diamniotic twin pregnancy at 11 weeks. An ultrasound scan done at 21 weeks revealed that the other twin was a vanishing twin.

After she was born, she was noted to have a symmetrically distributed skin lesions on the trunk bilaterally, with areas consisting of transparent membranes with the absence of skin components. These lesions were extending from the axillary areas along the side of the trunk towards loin and buttock areas, on both sides of the trunk (Figure 1).

There were no skin defects causing herniation of the thoracic or abdominal contents and no associated bleeding or signs of inflammation at birth. Skin over the other body parts, the skull, upper and lower limbs were normal. She had normal hair distribution and normal nails. She was not dysmorphic and the rest of the systematic examination was normal. Dermatology referral confirmed the diagnosis of Aplasia cutis of the trunk secondary to fetus papyraceus.

She had a 2D Echo, an ultrasound scan of brain and an ultrasound scan of abdomen to exclude associated abnormalities. Echocardiogram showed a tiny haemodynamically insignificant patent ductus arteriosus while the other two scans were normal. Plastic surgical follow up was arranged and baby was put on atraumatic silver mesh for the oozing areas with further intervention planned in the future.

Discussion
ACC is reported to affect 1 per 10,000 live births[3]. Though numerous theories have been postulated to explain the occurrence of ACC, neither the pathogenesis nor the aetiology is clear[1]. Non-scalp lesions are usually bilateral and symmetrical mainly involving the trunk and/or extremities[3]. In our patient, there were bilateral symmetrically distributed skin lesions on the trunk. The management of non-scalp ACC is still controversial. Most lesions heal spontaneously with conservative dressing, but large lesions may necessitate surgical interference with skin grafts or local skin flaps[5]. Cultured epithelial autografts have been used together with acellular allogenic dermal grafts[6]. The non-scalp ACC may be associated with epidermolysis bullosa[7].

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(Received on 12 May 2016: Accepted after revision on 17 June 2016)
The authors declare that there are no conflicts of interest
Personal funding was used in formulating the article.

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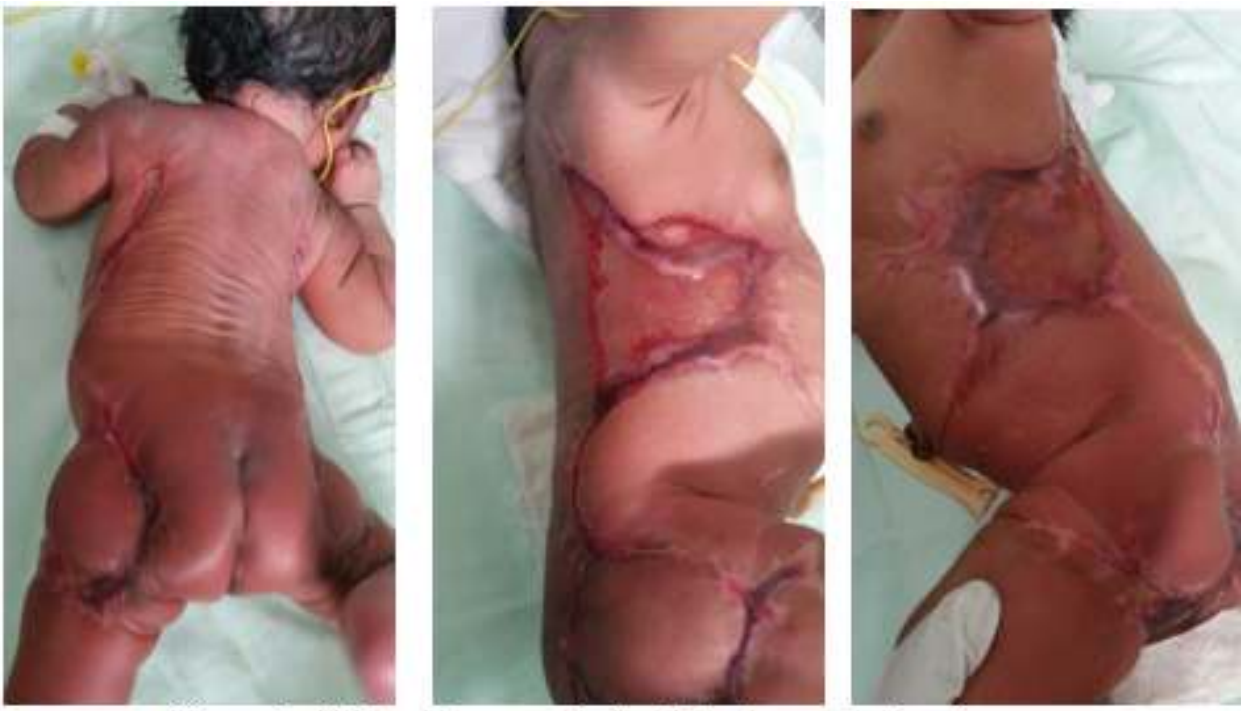

Figure 1: ***Bilateral symmetrical skin lesions noted on day one***