Cutis Aplasia: An Unusual Presentation

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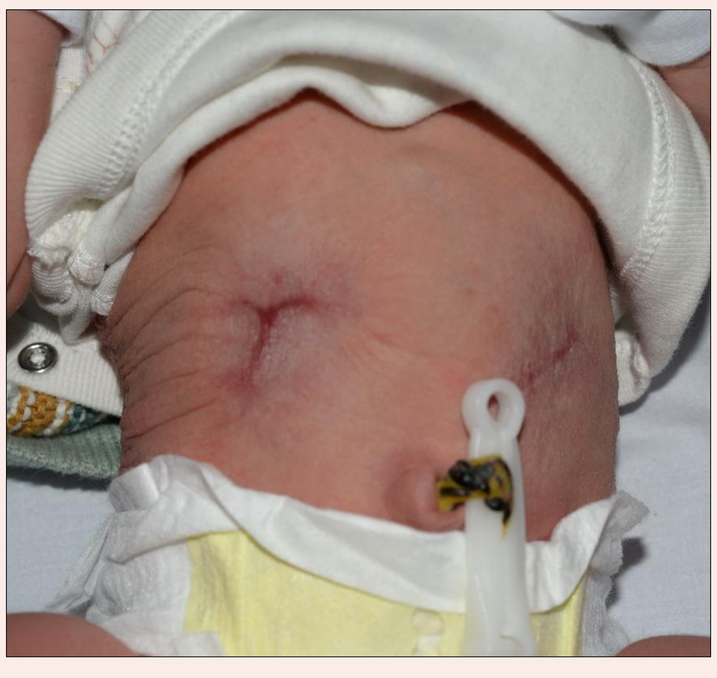
Case Description

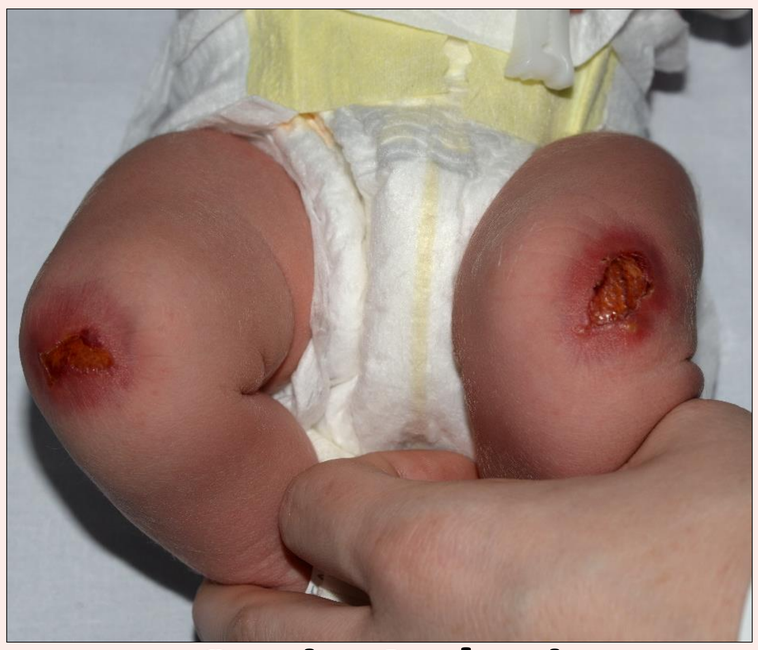
A term baby was noted to have extensive skin lesions following an uneventful vaginal delivery. The antenatal history had been significant for early demise of one twin in utero, with subsequent unremarkable development of the other twin.

At birth, the baby was noted to have symmetrical well-demarcated, flat, non-inflammatory lesions to the extensor surfaces of his legs and bilateral abdomen. There were no other skin lesions (including the scalp) and the baby was otherwise well (see pictures of lesions taken by medical photography on day 1 of life).

Diagnosis

The baby was seen by the dermatologist, who diagnosed non-syndromic cutis aplasia congenita. Scars were managed by the tissue viability team with daily moisturisation. The lesions were maturing at 6 month review and the baby is expected to be discharged from follow up by 18 months.





Cutis Aplasia

Cutis aplasia, or aplasia cutis congenita, is a rare congenital disorder affecting neonates, marked by partial or complete skin absence, commonly on the scalp. Etiology involves genetic and environmental factors. A classification system of 9 groups has been suggested. Management varies from conservative wound care to surgical interventions.[1]

Fetus Papyraceus

On reviewing the literature around cutis *aplasia/aplasia cutis congenita*, there have been several reports of *fetus Papyraceus* associated with *vanishing twin syndrome*[2]. Although rare, several cases have been reported[3]. Several case reports note foetal demise or iatrogenic foetal reduction in the first or early second trimester[4]. Case reports seem to describe a similar distribution of skin lesions where there had been confirmed twin pregnancy with early demise of one twin; the mechanism for this is unclear, although involvement of other systems should be considered[5].

Acknowledgements

All pictures shared with consent from parents.

References

(1) Graham, J.M. and Smith, D.W. (2007) 'Chapter 39 - Aplasia Cutis Congenita: Scalp Vertex Cutis Aplasia, Temporal Triangular Alopecia', in *Smith's recognizable patterns of human deformation*. Philadelphia, PA: Saunders, Elsevier.
(2) Bourque, S. and Preloger, E. (2015) 'Extensive aplasia cutis congenita with associated vanishing twin , *The Journal of Pediatrics*, 167(3). doi:10.1016/j.jpeds.2015.06.041.
(3) Pieretti, M.L. *et al.* (2015) 'Aplasia cutis congenita associated with fetus papyraceus', *Pediatric Dermatology*, 32(6), pp. 858–861. doi:10.1111/pde.12651.
(4) Bassi, A. *et al.* (2018) 'Aplasia cutis congenita and "vanishing twin" caused by iatrogenic fetal reduction', *Archives of Disease in Childhood - Fetal and Neonatal Edition*, 103(3).
(5) Louise, L. *et al.* (2013) 'Fetus papyraceus: Congenital pulmonary anomalies associated with congenital aplasia cutis on the surviving twin', *Pediatric Dermatology*, 30(6). doi:10.1111/pde.12096.
syndrome'