

Aplasia cutis congenita and amniotic band syndrome: an uncommon association

Joana Pimenta,¹ Patrícia Lapa,¹ Lina Ramos²

¹Department of Neonatology, Maternidade Dr Daniel de Matos—Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal
²Pediatric Hospital of Coimbra, Coimbra, Portugal

Correspondence to
Dr Joana Pimenta,
joanafmp@gmail.com

Accepted 14 January 2017

DESCRIPTION

Amniotic band syndrome is an unusual clinical entity with a wide variety of presentations that include constriction rings, syndactyly and limb or digital amputations.^{1 2} Aplasia cutis congenita is a rare congenital anomaly involving variant layers of the skin, most commonly affecting the scalp.³ Amniotic bands following rupture of the amniotic membrane in early pregnancy have been proposed as a cause of aplasia cutis congenita.³ Still, the presence of a fibrous amniotic band at the site of the constriction ring at birth is extremely unusual. This case further adds to the evidence of amniotic bands as aetiology for constriction rings and aplasia cutis.

A full-term female neonate was delivered by caesarean section as a result of non-progressive labour. The pregnancy had been uneventful and there was no remarkable family history. Physical examination at birth showed several constriction rings around both hands and feet, mostly affecting the digits and toes. All the distal phalanges of the right hand were involved (figure 1) and there was syndactyly of the 2nd and 3rd toes of the left foot (figure 2). There were no signs of limb perfusion abnormalities and there was no neurological deficit (motor and sensitive). A band of dark inelastic fibrous material was present within the constriction areas of the right hand (figure 1). Scalp examination revealed five well-demarcated, non-inflammatory, oval lesions, the largest with ~25 mm, involving the epidermis



Figure 2 Left foot with distal phalanges malformations and syndactyly between the 2nd and 3rd toes.

and the upper dermis (figure 3). The remainder of the physical examination was normal. Posterior investigation, namely transfontanelar ultrasound, revealed no other associated abnormalities.



Figure 1 Right hand showing constriction rings with persistent dark fibrous band around fingers digits, associated with distal partial autoamputation.



Figure 3 Areas of aplasia cutis congenita on the scalp.



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To cite: Pimenta J, Lapa P, Ramos L. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2016-218950

Learning points

- ▶ Amniotic band syndrome and aplasia cutis congenita are two unusual clinical entities with a variety of clinical presentations and an unclear underlying cause.
- ▶ Rupture of the amniotic membrane in an early pregnancy, forming amniotic bands, has been proposed as a cause of aplasia cutis congenita.
- ▶ Although aplasia cutis congenita often occurs as an isolated phenomenon, the presence of constriction rings seems to support the contribution of amniotic bands as a possible aetiology of aplasia cutis.

Contributors JP wrote the draft and conducted this publication. PL reviewed the manuscript. LR reviewed the manuscript and supervised the patient management. JP also was responsible for the patient orientation.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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