Findings that shed new light on the possible pathogenesis of a disease or an adverse effect

Isolated cranial distortion mimicking caput succedenum from amniotic band disruption without any neurological abnormality

Urmila Pillai,¹ Kausik Ray,² Anil Garg¹

¹ Department of Paediatrics, Worthing Hospital, Worthing, UK

² Department of General Surgery, Worthing Hospital, Worthing, UK

Correspondence to Kausik Ray, ray_kausik2003@yahoo.co.in

Summary

This report describes a term newborn with isolated distortion in the left parietal bone without any other visible congenital anomaly, due to amniotic band disruption. A skull x-ray, ultrasound scan and subsequent MRI scan of the brain did not show any apparent distortion apart from depression and concavity in the left parietal bone. The purpose of this case report is to raise awareness of this possible, mild outcome of this little-known entity, which may mimic caput succedaneum (moulding of the presenting part in the birth canal during natural delivery), and to provide a historical and embryological background.

BACKGROUND

Amniotic band disruption syndrome is a rare entity which occurs in 1 in 1200 to 1 in 15 000 live births.¹ It may cause a myriad of deformities of fetal body parts from mild defects in limbs to severe craniofacial defects incompatible with life. The spectrum of defects includes disruption, deformation and malformation of different body parts due to interference from the amniotic bands at different stages of organogenesis.² The syndrome is also described as ADAM complex (for 'amniotic deformities, adhesions, mutilation') or amniotic band disruption complex (ABDC) for its nature of presentation. Since Tropin described the condition in 1968,³ many questions regarding this syndrome remain unanswered because of its rarity and complex mechanism. This case is unique in its presentation as it mimics caput succedaneum without any neurological defect. This type of

presentation has not been described before in the medical literature.

CASE PRESENTATION

A girl, born at 38 weeks of gestation by vaginal delivery of normal duration of labour, presented with a large caput over the left parietal region (figure 1A,B). The mother was a 32-year-old primigravida without any significant medical illness. There was no family history of consanguinity or chronic illness. The pregnancy was uneventful and antenatal scans were reported as normal. The mother had a history of premature rupture of the membrane of more than 24 h and had prophylactic antibiotics. The baby did not cry immediately after birth. The Apgar score at 1 min was 5, and this increased to 9 after five inflation breaths. Her blood sugar was 6.1 mmol/litre. Her birth weight was 1960 g (9th

Figure 1 (A) The newborn with scalp defect. (B) Profile view.

Figure 2 x-Ray of the skull (anterioposterior view).

to 25th centile) and head circumference was 29 cm (9th centile). She was pink, handling well and moving all her limbs spontaneously. Her oxygen saturation on air was 99%. She did not have any respiratory distress, visible congenital anomaly or audible cardiac murmur. There was no frontal bossing, ear abnormality or hypertelorism. Musculoskeletal and neurological examination of the baby was normal. The craniofacial contours of the parents were unremarkable.

The circumference of the asymmetric swelling of the scalp was 26 cm. A clear wide demarcation line was present between the swelling and the vessels of the scalp. The skin over the swelling appeared darker than the rest of the scalp. Cranial sutures including the sagittal suture could not be felt due to the swelling. The newborn was given nasogastric feeding and intravenous cefotaxime.

INVESTIGATIONS

Her blood investigations were all within normal limits. A skull x-ray showed moulding of the skull bone over the left parietotemporal region (figures 2 and 3). No overriding or fracture was found except soft tissue swelling overlying the vertex. An ultrasound scan of the brain showed symmetrical lateral ventricles in both hemispheres with falx at the midline. A collection of fluid of 0.7–1 cm in diameter was found over the calvaria, which was suggestive of caput succedaneum.

A subsequent MRI showed the left hemicranium was distorted with relative preservation on the right side (figure 4). There was some concavity of the right parietal bone at the edge of caput succedaneum and similar inbowing of the lateral cranium on the left side, which was suggestive of modelling by an amniotic band.

DIFFERENTIAL DIAGNOSIS

The initial impression for the abnormal shaped skull was either a caput or isolated presentation of ABDC.

Figure 3 x-Ray of the skull (lateral view).

TREATMENT

The baby was treated conservatively. Paediatric neurosurgeons suggested masterly inactivity.

OUTCOME AND FOLLOW-UP

The baby has been doing well without any neurodevelopmental delay at 6-month and 9-month follow-ups.

DISCUSSION

Though the exact aetiology of ABDC is unknown, two main pathogenetic mechanisms are proposed. Exogenous theory suggests early amnion rupture leading to a fibrous band that can entrap fetal body part (figure 5). The endogenous theory suggests vascular compromise.⁴ Genetic factors might operate in some cases.⁵

Moerman *et al* described three types of lesions in this complex: (A) constrictive tissue bands (shrivelled amniotic strands), (B) amniotic adhesions (fusion between disrupted body part and intact amniotic membrane) and (C) limb–body wall complex.⁶ According to their observations, most of the craniofacial defects occur as a result of a vascular disruption sequence with or without cephaloamniotic adhesions, unlike the case described here that had the clear mark of a constriction band without significant vascular compromise.

An accurate diagnosis may be achieved by looking for the major features of amniotic band syndrome, and a routine chromosome study and placental examination in cases with multiple congenital anomalies.^{2,7}

Figure 4 MRI of the head.

Though encephalocele, clefts, distortion and dislocation of craniofacial structures have been described as presentations of craniofacial defect related to this syndrome in the medical literature,⁵ distorted hemicranium (mimicking caput succedaneum) without any neurological defect has never been described previously. The outcome of the syndrome depends on the gravity of malformation. Termination of pregnancy is considered in cases of severe craniofacial or visceral abnormality. Successful limb salvage by fetoscopic release of an amniotic band has been reported.⁸ Most of the minor craniofacial defects require multiple surgical procedures to restore function, with significant compromise to satisfactory cosmesis.

Learning points

- ▶ Isolated calvarial deformity mimicking caput succedaneum from amniotic band disruption is a possibility.
- ▶ Careful neurological assessment and MRI are useful to exclude any neurological abnormality.
- ▶ Masterly inactivity with regular follow-up is appropriate management in such cases.

Competing interests None.

Patient consent Obtained.

Figure 5 Diagrammatic representation of amniotic bands.

REFERENCES

1. **Sentilhes L**, Verspyck E, Eurin D, *et al*. Favourable outcome of a tight constriction band secondary to amniotic band syndrome. *Prenat Diagn* 2004;**24**:198–201.
2. **Huang CC**, Eng HL, Chen WJ. Amniotic band syndrome: report of two autopsy cases. *Changgeng Yi Xue Za Zhi* 1995;**18**:371–7.
3. **Tropin R**. *Fetal malformations caused by amnion rupture during gestation*. Springfield, Illinois, USA: Charles C Thomas, 1968, 1–76.
4. **Sentilhes L**, Verspyck E, Patrier S, *et al*. Amniotic band syndrome: pathogenesis, prenatal diagnosis and neonatal management [in French]. *J Gynecol Obstet Biol Reprod (Paris)* 2003;**32**:693–704.
5. **Keller H**, Neuhauser G, Durkin-Stamm MV, *et al*. “ADAM complex” (amniotic deformity, adhesions, mutilations) – a pattern of craniofacial and limb defects. *Am J Med Genet* 1978;**2**:81–98.
6. **Moerman P**, Fryns JP, Vandenbergh K, *et al*. Constrictive amniotic bands, amniotic adhesions, and limb-body wall complex: discrete disruption sequences with pathogenetic overlap. *Am J Med Genet* 1992;**42**:470–9.
7. **Wang JK**, Changchien CC. The amniotic band syndrome—report of two cases [in Chinese]. *Changgeng Yi Xue Za Zhi* 1990;**13**:343–9.
8. **Keswani SG**, Johnson MP, Adzick NS, *et al*. In utero limb salvage: fetoscopic release of amniotic bands for threatened limb amputation. *J Pediatr Surg* 2003;**38**:848–51.

This pdf has been created automatically from the final edited text and images.

Copyright 2010 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.

BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Pillai U, Ray K, Garg A. Isolated cranial distortion mimicking caput succedenum from amniotic band disruption without any neurological abnormality. *BMJ Case Reports* 2010;10.1136/bcr.12.2009.2549, date of publication

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow