Case report

Neonatal leg fracture and constriction ring syndrome: A case report and literature review

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ABSTRACT

Congenital constriction of the limbs is usually due to amniotic band syndrome, which often causes damage to the skin and soft tissues. We report an unusual case in which a neonate had recent fractures of both leg bones with an amniotic band encircling the limb. Non-operative treatment was successful. Challenges to the management of neonatal limb constriction include the absence of a consensus about the best treatment and the high frequency of damage to vessels and nerves.

1. Introduction

Constriction ring syndrome (CRS), also known as amniotic band syndrome, is characterised by various congenital defects due to constriction of one or more parts of the foetus by fibrous bands derived from the amniotic sac. Although the limbs are predominantly affected, the head and torso are not exempt [1,2].

Limb defects are the main features of CRS. Among them, the most common are band indentations, amputations, syndactyly, and clubfoot [1–6].

Here, we report an unusual case of CRS with recent fractures of both leg bones related to an amniotic band.

2. Case-report

A female newborn was delivered from a 30 year-old, gravida 3, para 2 woman, at the 33rd week of gestation, by Caesarean section for breech presentation and active-phase arrest of labour. The mother had not noticed any abnormal decrease in foetal movements during the pregnancy.

During the Caesarean section, the performing obstetrician found an amniotic band wrapped three times around the neonate’s left leg and released it gently. The neonate had severe, circumferential swelling of the left foot and ankle distal to the constriction, with three deep band indentations. There were no skin breaches or signs of acute lower-limb ischaemia. Capillary pulsations were felt at the toes and the capillary refill time was normal (Fig. 1).

The plain radiograph showed a fracture of the distal third of the tibia and fibula, at the level of the constriction. The size and density of the soft tissues of the distal half of the leg were considerably increased. Bone density was normal, with no evidence of osteodystrophy (Figs. 2 and 3).

The limb was immobilised in a long-leg posterior gutter splint and the limb was kept elevated. Monitoring consisted in a daily clinical examination and weekly radiograph. The outcome was favourable with no vascular, nervous, or cutaneous complications. The splint was removed after 8 weeks.

At last follow-up, the patient was 12 months of age and had been walking for 1 month. Fracture healing was complete (Figs. 4–6). The only abnormality by physical examination was a circumferential band indentation that was deepest at the posterior aspect of the leg (Figs. 7 and 8). A Z-plasty will be performed during the second year of life.

3. Discussion

CRS is not uncommon, and the diagnostic and management strategy is now well-standardised, despite continuing uncertainty about the underlying pathogenesis [1,2,5]. CRS may manifest as congenital defects or deformities of the limbs such as clubfoot, syndactyly, and lymphoedema [1–7]. Tadmor et al. reported a case of spontaneous limb amputation in utero due to a constriction ring encircling both lower limbs [8].
Apart from amputations, bone abnormalities are rare in CRS. Among them, the most common is congenital pseudarthrosis due to constriction by an amniotic band [9–13]. Table 1 recapitulates the published cases.

The case reported here is unique: we are not aware of previous reports describing a recent fracture due to an amniotic band, with no vasculo-nervous abnormalities or congenital defects of the other limbs. The radiographs showed no signs indicating an old fracture (e.g., sclerosis of the edges, incipient callus formation, or periosteal reaction) (Figs. 2 and 3). The recent nature of the fracture was further supported by the absence of a decrease in foetal movements during the pregnancy and by the development of signs of bone union at 3 weeks of postnatal age.

Another unusual feature of our case is the type of management. Surgery was performed in all five reported cases of congenital pseudarthrosis (four at the leg and one at the forearm) [9–13]. The indication for surgery was based on the high risk of non-healing with non-operative treatment and on the presence of complications affecting the skin, vessels, or nerves. However, in our case, given the recent nature of the fracture, we opted for non-operative treatment. The main challenges in this type of management were as follows:

- high risk of vascular complications due to the constriction, severe oedema, and small calibre of neonatal blood vessels;
- local changes in the limb, which contra-indicated both surgery and circumferential cast at the acute phase;
- fracture instability, due to the short oblique fracture lines located at the same level on the tibia and fibula;

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Table 1
Literature review on bone fractures due to amniotic band syndrome.

<table>
<thead>
<tr>
<th>Author</th>
<th>Type of bone lesion</th>
<th>Vessel and nerve injuries</th>
<th>Concomitant defects</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Zych et al. (1983)</td>
<td>Pseudarthrosis (leg)</td>
<td>Acute ischaemia/posterior tibial nerve entrapment</td>
<td>Constriction of the fingers of one hand</td>
<td>Fasciectomy/nerve release pin fixation</td>
</tr>
<tr>
<td>Martinot-Duquen-noy (1991)</td>
<td>Open pseudarthrosis (leg)</td>
<td>Acute ischaemia</td>
<td>Bilateral amputation of the hallux</td>
<td>Debridement/external fixation</td>
</tr>
<tr>
<td>Tanguy (1995)</td>
<td>Open pseudarthrosis (leg)</td>
<td>❌</td>
<td>Constriction of the other toes</td>
<td>Debridement/decortication/immobilisation</td>
</tr>
<tr>
<td>Agashe (2011)</td>
<td>Pseudarthrosis (leg)</td>
<td>❌</td>
<td>Bilateral amputation of the hallux</td>
<td>Amputation</td>
</tr>
<tr>
<td>Ho, Christine (2014)</td>
<td>Pseudarthrosis (forearm)</td>
<td>Acute ischaemia</td>
<td>Acro-syndactyly of both hands</td>
<td>Decortication/immobilisation</td>
</tr>
<tr>
<td>Our patient</td>
<td>Recent fracture (leg)</td>
<td>Acute ischaemia</td>
<td>Marked lymphoedema</td>
<td>Immobilisation</td>
</tr>
</tbody>
</table>

Fig. 5. Radiograph of the left leg during the 8th postnatal week: bone union.

Fig. 6. Radiographs of the left leg at last follow-up at 12 months of age.

Fig. 7. Clinical appearance at last follow-up at 12 months of age in the supine position.

Fig. 8. Clinical appearance at last follow-up at 12 months of age in the standing position.
high risk of fracture non-union related to the distal location of the fracture, poor blood supply, and limited bulk of the overlying muscles;
• risk of inducing vascular complications during external reduction of the fracture [11].

Emergent surgical treatment was considered but not performed, given the good toe colour with capillary pulsations, intact skin barrier, absence of neurological complications and compartment syndrome, and rapid decrease in swelling since the first week.

Among previously reported cases, only two required primary surgery to remove the constricting band [10,11]. Two other patients required delayed surgery (at 18 h and 4 days of postnatal age) because of clinical deterioration with cold cyanotic extremities, abolition of the capillary pulsations, development of a motor deficit, and worsening of the oedema [9,13].

In our patient, the persistence at 12 months of age of a deep and complex band indentation indicates that chronic constriction had occurred, probably due to an amniotic band. Band indentation and lymphoedema were probably the initial manifestations of limb constriction. We believe the fracture occurred only in late pregnancy, due to increasing mechanical stress on the bones related to foetal growth within the confines of the uterine cavity.

4. Conclusion

This unusual case of a recent leg fracture associated with CRS in a neonate highlights the challenges in managing this condition. Non-operative treatment may allow the fracture to heal but requires close monitoring of limb vascularisation. Surgical treatment should be considered only in case of complications such as acute ischaemia, compartment syndrome, or skin breach.

Disclosure of interest

The authors declare that they have no competing interest.

References