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TOTAL CIRCULAR CONGENITAL ABDOMINAL AMNIOTIC BAND

Paediatrics

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ABSTRACT

Introduction: Congenital amniotic band syndrome is a rare congenital abnormality causing multiple disfiguring and disabling manifestations. The most common affection occurs at the lower extremities. A total circular congenital amniotic band syndrome of the trunk is an even more rare condition. The incidence varied from 1 in 20000 live births. Here, we report a neonate with a total circular congenital amniotic band of the abdomen with another on the right leg.

Case: A female neonate, with an uneventful prenatal period, presented on first day of life with a constriction band around the lower abdomen with loss of intervening skin and subcutaneous tissue with a similar band over the mid-part of right leg with bilateral Congenital Talipes Equinovarus. X-ray images of abdomen and lower limbs showed soft tissue loss over the affected area without any affection of the muscle and bones.

KEYWORDS

Amniotic band, Abdominal amniotic band, Congenital amniotic band

INTRODUCTION

Congenital amniotic band syndrome is a rare congenital abnormality causing multiple disfiguring and disabling manifestations. The most common affection occurs at the lower extremities. A total circular congenital amniotic band syndrome of the trunk is an even more rare condition. The incidence varied from 1 in 20000 live births. Here, we report a neonate with a total circular congenital amniotic band of the abdomen with another on the right leg.

CASE REPORT

A female neonate, born to parents from the Dooars in West Bengal, was referred from post natal care services on her first day of life for a constriction band around the lower abdomen with loss of intervening skin. The prenatal follow-up as well as the delivery was uneventful.

The baby presented with a constricting ring over the abdominal wall in total circumference at the level of the anterior superior iliac spine (Figure 1) as well as over the mid-part of right leg (Figure 2). The baby also had bilateral Congenital Talipes Equinovarus (Figure 2).

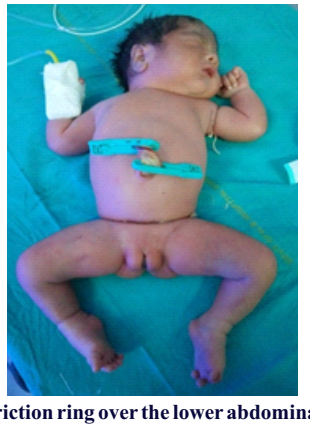

Figure 1: Constriction ring over the lower abdominal wall

Figure 2: Constriction ring over the lower part of the right leg

The neonate was full term born by normal vaginal delivery. There was no family history of congenital deformities.

There was loss of skin and subcutaneous fat with exposure of deep fascia over the lateral aspect of the waist (Figure 3) and back (Figure 4) with a thick fibrous intervening bridge of tissue centrally over the anterior aspect of the abdomen (Figure 5). The ring over the lower limb was thick and fibrous without any loss of skin continuity

Figure 3: Exposure of the deep fascia over the lateral aspect of the trunk

Figure 4: Exposure of the deep fascia over the back

Figure 5: Fibrous tissue over the anterior abdominal portion of the ring

X-ray images of abdomen and lower limbs showed soft tissue loss over the affected area without any involvement of the muscle and bones (Figure 6, Figure 7).

Figure 6: Defect in soft tissue showing the area of amniotic band

There was associated bilateral CTEV seen on the Xray image (Figure 7).

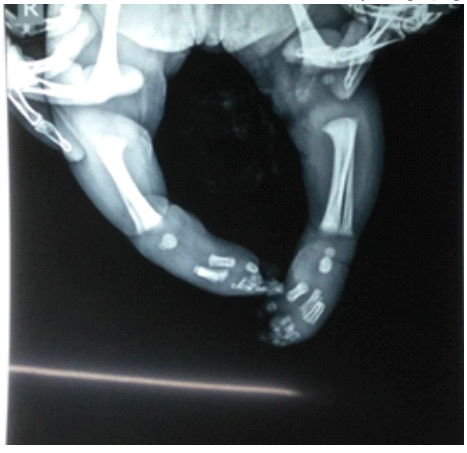

Figure 8: Bilateral CTEV and a soft tissue defect on Right leg indicating the amniotic band defect

DISCUSSION

A congenital constriction amniotic band is a disorder which presents in a newborn with constriction rings or bands. It causes tissue depression, digital encirclement, and involves extremities or limbs and rarely even the neck, thorax, and abdomen [1]. It may lead to severe malformations like anencephaly, misshapen head, truncal deformities and severe spinal as well as limb deformities [2]. The overall reported prevalence of amniotic bands in the UK and Western Europe is about 1 per 20,000 births [3]. There is neither any familial reoccurrence nor any genetic or gender predisposition.

The aetiopathogenesis is unknown. However there are two theories suggesting the aetiology [1, 4]. One theory states that these deformities are a result of defective intrinsic germ plasma within the embryo leading to the defect. According to the extrinsic theory, these lesions are caused due to strangulation by the mesodermic bands, following early rupture of the amnion.

These constrictions were first described by Patterson in 1961 [5]:

- Simple constriction rings without distal deformity
- Constriction rings with distal soft tissue deformities including or excluding lymphoedema
- Constriction rings with distal deformities including distal bony fusions from mild to severe acrosyndactyly
- Intrauterine amputations

Amniotic bands are either acellular or fibrous tissue containing fibroblasts, lined by squamous cells [6].

A circular constriction band over the abdomen is very rare. It was first reported by Brown et al. in 1957, and thus far, only about 20 cases have been reported in literature. Depth of the lesion varies from a superficial groove to a deep gutter and usually extends only up to the fascial layer. Histologically, it shows a thickened dermis with collagen bundles and elastic fibres with large amounts of hyalinized connective tissue in case the patient has intact skin with an indentation. Even in their rarity, these bands, when over the trunk, usually present above the pelvic rim with the mid abdominal variety being extremely rare [7]. However there have been extremely rare instances of patients having multiple bands over the body.

The treatment of the circular constriction band usually includes removal of the fibrotic tissue with 1 to 2 mm of intact skin including Z-plasties or W-plasties to avoid further constriction or re-contracture of the scar. Subcutaneous tissue deficiency under the constriction ring can be corrected with rectangular plasty techniques and by using flaps [8].
If there is digital or extremity constriction, urgent surgical treatment may be warranted to reverse vascular compromise. Otherwise, cosmetic staged correction is usually conducted to maintain the limb or digital vascularity.

CONCLUSION

A circular constriction amniotic band of the abdomen is an extremely rare congenital malformation. Limb or digital bands may cause vascular compromise or auto amputation and therefore should be intervened early but not all present as absolute emergencies. However abdominal bands usually have cosmetic implications. These are successfully treated by excision of the fibrotic band and the correction of the subcutaneous deficiency under the constriction.

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