

Case Report

A NEONATE OF AMNIOTIC BAND SYNDROME WITH PROGRESSIVE LYPHEDEMA AND RECENT FRACTURES: A RARE CASE REPORT

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ABSTRACT

Amniotic band syndrome (ABS) is an uncommon congenital anomaly causing entrapment of fetal parts (usually limbs, digits) in fibrous amniotic bands in utero. It may affect the bone, muscle, nerves, and vascular bundles according to the depth of the constriction band. Every case of this syndrome is unique and so the management is different for each case. We report one such interesting case of amniotic band syndrome who presented at day three of life with tight constriction at right lower third leg with concomitant fracture both bone leg in association with acrosyndactyly right hand and autoamputated left thumb. The limb was salvaged in spite of late presentation as compartment syndrome with satisfactory functional outcome.

Keywords: *Amniotic Band Syndrome, Entrapment, Acrosyndactyly*

INTRODUCTION

Amniotic band syndrome (ABS) consists of a wide spectrum of congenital malformations depending on the affected body parts. There is no genetic cause for ABS so no recurrences in siblings. Maternal trauma, teratogenic insult, oophorectomy during pregnancy, intrauterine contraceptive device, amniocentesis and familial incidence of connective tissue disorders (Ehler-Danlos syndrome) are some of the implicated etiopathological factors (Mistry *et al.*, 2015)

It affects both sexes equally with an incidence of 1 in 1,200 to 15,000 live births (Stevenson *et al.*, 2006) and 1 in 70 stillbirths (Kalousek *and* Bamforth 1988). Due to possibility of different combinations of anomalies, there are no two identical cases of ABS. According to Paterson (Patterson, 1961), the diagnosis the ABS should include at least two of the following diagnostic criteria: Simple constriction ring; ring of constriction with distal deformity, with lymphedema or without it; fusion of distal parts; congenital digital amputations.

Treatment is individualised and depends on severity of deformity at the time of presentation. The prognosis depends on the severity of the abnormalities. Orthopedic treatment is based on release of constriction band with tissue decompression to improve function and appearance of the limbs. However, few authors prefer staged procedure in form of W or Z-plasty without removal of soft tissue for fear of vascular insufficiency. In literature there is few information regarding management of progressive lymphedema in the cases of ABS with associated fracture at the site of band. We present one such rare case of complicated amniotic band syndrome who underwent staged procedure with satisfactory outcome.

CASE

We report an unusual case in which a three-day neonate presented with right foot gross swelling, distal pulses feeble and delayed capillary time with tight amniotic band encircling lower third of leg (Fig 1). She also had fracture both leg bones at the same site as the band. Associated anomalies included acrosyndactyly of right hand and left foot with autoamputated left thumb (Fig 2). She was evaluated with colour doppler right foot showing compartment syndrome and roentgenogram suggestive of fracture both tibia and fibula at the site of constriction ring. Emergency fasciotomy with release of amniotic band was done for compartment syndrome to salvage limb. Fractures were managed non surgically by immobilisation in plaster slab. Post-procedure colour of limb improved and edema reduced. After seven

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days she underwent debulking of dorsal tissue and closure of defect. Post-operative recovery was uneventful with good functional result obvious within 15 days of surgery (Fig 3). Fractures healed with three weeks of POP slab (Fig 4). Acrosyndactyly release was done at the age of 3 months along with creation of adequate web spaces with the help of free full thickness skin graft (Fig 5). The functional outcome of both lower limb and hand were satisfactory.



Figure 1: Preop picture



Figure 2: Acrosyndactyly



Figure 3: Immediate post op picture



Figure 4: X ray at presentation and after 3 weeks



Figure 5: Acrosyndactyly released

DISCUSSION

Management strategy of ABS depends upon the extent of the associated anomalies. Treatment is mostly surgical though few cases of non-operative management have also been described. Most references recommend the use of Z-plasty or W-plasty after the excision of the constriction band, in one- or two-stage approach. For severe craniofacial and visceral abnormalities termination of pregnancy is proposed, whereas minor limb defects are repaired postnatally. Lately, prenatal ABS treatment in the form of fetoscopic laser cutting of amniotic bands has been attempted for cases which are at risk of severe deformity or limb loss if not managed antenatally (Quintero, 1997). Patterson in his study of 52 patients of congenital constriction rings had reported only two cases of below knee amputations in addition to other musculoskeletal defects (Patterson, 1961) Zych and Ballard, in 1983 reported a case of involvement of congenital bands, pseudarthrosis and impending gangrene of leg, which was salvaged with multiple Z-plasty (Zych and Ballard, 1983) Greene (1993) had advised a one-stage release for circumferential

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congenital constriction bands which was performed in all four extremities (Greene,1993). In 2006, Samra *et al.*, reported a case of severe constricting amniotic band with a threatened lower extremity in a neonate, which was salvaged with multiple Z-plasties over a 6-year functional follow up. Samra *et al.*, (2006) Recently, Choulakian and Williams (2008) has described a two-staged approach of direct closure after excision of the constriction band. Aleman and Russo (2017) have described a case of progressive lymphedema due to amniotic constriction band leading to vascular compromise which was managed by decompressive fasciotomy followed by radical excision of overgrown tissue and primary anastomosis (Aleman *et al.*, 2017) K Masmoudi et al described non-operative management in a neonate who had recent fractures of both leg bones with an amniotic band encircling the limb (Masmoudi *et al.*, 2016). Our case was unique because of its presentation in the form of progressive lymphedema causing compromised vascularity along with recent fractures both leg bones. It was managed with satisfactory functional outcome as a staged procedure for the constriction band and removal of hypertrophied tissue with non operative management of fractures per se.

CONCLUSION

Every case of amniotic band syndrome is unique and so the management is individualised ranging from non-operative intervention to staged procedure. Overall outcome depends on the severity of deformity caused by the bands.

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