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Severe acrosyndactyly and digital amputation following amniotic band syndrome in a Nigerian neonate, case report and review of literature

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Abstract

Aim: To highlight an uncommon presentation of a complex case of severe acrosyndactyly and digital amputation in a neonate following amniotic band syndrome. Introduction: Congenital amniotic band syndrome as a characterized by strands of amnion entrapping portions of the developing fetus. It can result in a wide range of congenital abnormalities, amputations and even death of the fetus. It affects commonly the extremities of the body. The presentation varies depending on the affected part of the body and its severity. Case report: The patient presented at 5days of life with a swelling of the distal portion of the left foot distal to a ring of hyperpigmentation. The portion proximal to this ring was normal. This was noticed at birth. There was also digital amputation of the index finger of the right hand noticed also at birth. Discussion: The etiology of amniotic band syndrome is believed to be from rupture of the amnion inutero and this condition is known by several other names. The management of these patients is usually multi-disciplinary in approach. Cosmesis was a major reason for intervention in this patient.

Keywords: *Acrosyndactyly, amnion, band, constriction*

Introduction

Congenital amniotic band syndrome is a condition involving fetal entrapment in strands of amniotic tissue causing an array of malformations. Band syndrome most frequently affects the distal segment including the hand[1]. It is an uncommon congenital pathology that may lead to malfunction and fetal – infant death[2]. Two main pathogenic mechanisms are proposed; early amnion rupture (exogenous theory) leading to fibrous bands, which entrap the fetal body and the endogenous theory with vascular compromise and mesoblastic strings[3].

Patients with this syndrome will usually present early to the surgeon because of the obvious cosmetic abnormality. A multidisciplinary approach is usually employed in managing these patients. Contributions from the pediatric, plastic, orthopedic and neurosurgeons are usually required. The outcome of the disease depends on the gravity of the malformation[3].

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In patients with distal edema and acrosyndactyly early repair portends better prognosis. Improvement in prenatal diagnosis and fetoscopic surgical technique may eventually allow treatment of amniotic band syndrome in-utero[1].

This article discusses a female neonate with involvement of both upper and lower extremities following amniontic bands.

Case Report

The patient is a 5day old male neonate delivered to a 33year old woman. The child is the third in monogamous setting of three children. The other siblings, a male and a female do not have any known congenital issue.

She presented to our pediatric surgical clinic with complaints of a swelling on the distal aspect of the left foot subsuming all the digits of the foot. There was also a complaint of shortened index finger of the right hand. These were all noticed at birth.

The mother had a materno-fetal ultrasound scan at 17 weeks gestation and these malformations not noted. The period of pregnancy was uneventful, and delivery was via spontaneous vertex delivery at a government hospital. There were no other complaints traceable to any other system.

At presentation there was a distal swelling involving the whole of the forefoot of the right foot taking up all the toes with it. Sparingly obvious were the pulp of the big toe and two other toes. Toenails were not present. The swelling was round, smooth surfaced with firm consistency, not tender and measured about 6cm in diameter.

There was a darkish circumferential ring of hyperpigmentation proximal to the commencement of the swelling. The patient had a complex syndactyly (rosebud foot) with forefoot swelling and edema.

The index finger of the left hand was observed to be shortened with a deformed nails bed and finger. There appears to be loss of the distal phalanx.

Examination of the other part of the musculoskeletal system was essentially normal. No other obvious congenital abnormalities were noticed on complete systemic examination of the neonate.

Figure 1 and 2: Pictures showing the distal swelling
The toenails were absent with only the distal pulp of some of the toenails visible. The contralateral limb appeared normal.

Figure 3: Picture showing the circumferential ring of hyperpigmentation proximal to the swelling
Figure 3 depicts the area of the amniotic band.

Figure 4: Picture showing the roundish swelling.

Figure 5 and 6: Pictures showing digit amputation with shortened index finger of the right hand.

Discussions

Congenital amniotic band syndromes are problems seen in the neonatal age-group accounting for some of the early causes of neonatal surgical consultations. It can affect virtually any system of the body with attendant significant morbidity and mortality depending on the severity.

The incidence of amniotic band syndrome varies from 1/1200 and 1/15000 live births[4]. The male to female ratio is about 1:1[5].

Amniotic band syndrome is also known by several names which include constriction ring syndrome, mesoblastic fibrous string and constriction band syndrome among many others. Its etiology is poorly known; it affects various fetal parts causing a wide range of conditions including amputation and death. Various abnormalities can be seen distal to the point of constriction, however the portion proximal to this is usually normal [6, 7,8.] This is clearly seen in the patient being discussed in this paper (Figures 1, 2, 3, and 4) who had a normal portion of the foot proximal to the constriction and the abnormality distal to it.

Amniotic band syndrome is not genetic and not inheritable. The commonly acceptable theory of etiology is the rupture of the inner membrane (amnion) without injury to the outer membrane (chorion) exposing the fetus to sticky tissue forming bands around body structures.

Diagnosis of amniotic band syndrome can be made in the intrauterine life by a fetal ultrasound scan. The index patient had a fetal ultrasound scan at about 17week gestation, but the abnormality was not noticed. The time of rupture of the amnion in amniotic band syndrome is not exactly known and could be variable. However, it is believed to occur after the period of organogenesis.

In this patient there was marked edema (commonly lymphedema) of the distal portion of the affected foot with complete absence of all the digits without interdigital web spaces. The only visible is the pulp of some of the toenails (Figure1) signifying the complex form of acrosyndactyly. The swelling was soft, not tender with a smooth surface.

The patient also had amputation of the distal phalanx of the index finger of the right hand (Figure 5, 6) making it shorter compared to the contralateral side.

Surgical correction is recommended in amniotic band syndrome with or without lymphedema[9]. Complex syndactyly and acrosyndactyly may require surgery early usually 6 months before simple syndactyly may wait until the infant is 18 months[10]. The main essence of surgery in this case will be to reduce the size of the swelling and achieve a cosmetically acceptable look. This is to also enable the foot to fit into sandals and shoes. Thus, the surgeries are usually individualized as no two cases of acrosyndactyly are the same[11].

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