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Case Report

Amniotic band syndrome: a rare entity to be encountered

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ABSTRACT

Amniotic band syndrome(ABS) is a disruption sequence with a broad spectrum of clinical manifestations ranging from partial amputations to major craniofacial and limb body wall defects. The commonest abnormalities usually involve the limbs and can range from simple constriction rings to complete amputation occurring at various levels. This case report is about a new born female baby who was normally delivered and she presented with complete absence of metacarpals and phalanges of the left hand. All the other limbs were normal. This case report throws a light on the different diagnostic modalities which can be used to diagnose amniotic band syndrome. Therapy of ABS is mostly surgical, with an individual approach to every single case. Interdisciplinary consulting and work is very often needed.

Keywords: Amniotic band syndrome, Constriction rings, Septo-optic dysplasia

INTRODUCTION

Amniotic band syndrome (ABS) is a rare congenital disorder that is associated with a wide range of physical abnormalities in the new born infant, some of which are significantly disabling and disfiguring in nature.[1]

The commonest abnormalities usually involve the limbs and can range from simple constriction rings to complete amputation occurring at various levels.

Abdominal wall defects and abnormalities of the cranio-facial region such as cleft lip and cleft palate are also associated with ABS while in the more complex cases, visceral defects such as renal agenesis 3 and rarely septo-optic dysplasia are also known to occur.[2-4]

Various studies estimate the incidence of ABS to be between 1 in 1300 to 1 in 15000 though the real figure is likely closer to the latter mark.[4,5]

CASE REPORT

A 33-year-old female, primi presented to our hospital at 39 weeks POG with c/o leaking per vaginum for 1 hr. Patient was thoroughly examined including general physical examination and local examination.

Patient's vitals were stable and her P/A corresponded to term size uterus, irritable with a regular FHS. Patient has no associated comorbidities such as Hypertension, Diabetes etc.

Patient has been explained regarding premature rupture of membranes and hence consent was taken for induction of labor. Patient was induced with prostaglandins and was given injectable antibiotics. After a labor duration of approximately 10-11 hrs patient delivered a single alive newborn female baby with birth weight 3.25 kg with APGAR score 9,9. On gross examination, the baby had absent fingers on the left hand, while right hand was

normal. Rest of the findings were normal. The baby was thoroughly evaluated. Anthropometric measurements were appropriate for gestational age. An infantogram, ultrasonography, X-rays B/L upper and lower limbs was done. On X-ray left hand phalanges and metacarpals were absent. Right hand was normal.

According to patient her pregnancy had been uneventful. Patient was asked to bring her previous USG reports with films. There was only one USG available with the patient at 12 weeks which revealed an amnion detachment of 1.27 cm, no follow up scans were done by patient since then.

Figure 1: Absent phalanges and metacarpals of left hands.

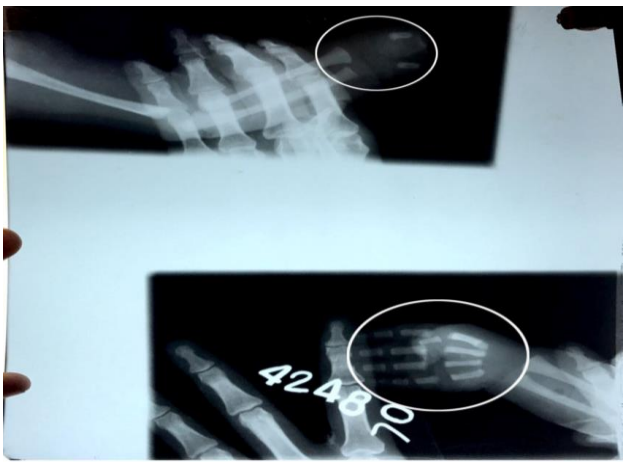

Figure 2: Absent phalanges and metacarpals of left hand (Upper),and normal right hand (Lower).

DISCUSSION

Amniotic band syndrome is a rare disorder. It often results in congenital physical defects in the infant, which are disabling and disfiguring. ABS etiopathogenesis is still unknown, but there are two main theories.[6-9] The widely accepted extrinsic model proposed by Torpin and Faulkner in 1966 explains defects genesis by rupture of the amnion in early pregnancy, with forming of amniotic bands and amniotic liquid loss, followed by extrusion of all or parts of the fetus into the chorionic cavity. Bands entrap the parts of the growing fetus and fetal limbs and other body parts become entangled and are subjected to compression. This compromise fetal circulation and also growth and development with consecutive disturbances of functions and anatomy.

The intrinsic model was proposed by Streeter in 1930 and suggests that the anomalies and the fibrous bands have a common origin, caused by a perturbation of developing germinal disc of the early embryo. ABS can be diagnosed prenatally by ultrasound, which can sometimes show amniotic bands but more often, malformation consistent with ABS as well as oligoamnion and reduction of fetal movements.[10]

The most important ultrasound diagnostic criteria are visible amniotic bands, constriction rings on extremities and irregular amputations of fingers and /or toes with a terminal syndactyly. Mild defects, however are less likely to be diagnosed prenatally, in which case defects are seen after birth.[11]

Physical examination is the main stay of postnatal diagnosis of ABS. However, uses of additional investigations like ultrasound, echocardiography, X-rays are important in order to establish potential malformations of different organs and body parts. Therapy of ABS is mostly surgical, with an individual approach to every single case. Interdisciplinary consulting and work is very often needed, (plastic surgeon, orthopaedic surgeon, orthodontist, ophthalmologist, neuro surgeon).[12]

Lately there have been some attempts of prenatal ABS treatment - fetoscopic laser cutting of amniotic bands, before their compression on the fetus makes malformations.[13]

CONCLUSION

ABS are not seen very often, but should be considered in every newborn with congenital anomalies, especially defects of extremities and /or body walls. The basis for postnatal diagnosis is physical examination of the newborn, with additional investigations to rule out other potential internal organ malformations. Because of ABS complexity, the treatment and follow-up of these children requires a team of specialists according to special needs of every patient.

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