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Amniotic Band Syndrome: A Rare Congenital Anomaly.

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ABSTRACT

Amniotic band syndrome also called as ADAM sequence or ABC is a rare interesting congenital anomaly which has got multiple etiological factors so also variations in presentation.ABS is a term applied to a range of congenital anomalies most typically limb and digital constrictions leading to amputations associated with fibrous bands of amniotic membrane. It can also show other forms of anomalies such as craniofacial like cleft lip, cleft palate, micrognathia or visceral like omphalocoele and more complex syndromes like Patau Syndrome or Septal Optic Dysplasia. We came across such a case which is presented herewith due to its rarity and interesting nature.

Keywords: Amniotic band syndrome (ABS), Amniotic deformity adhesion and mutilation complex or sequence (ADAM), Amniotic band constriction (ABC),Intrauterine growth restriction(IUGR).

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INTRODUCTION

The earliest known case of ABS has been described in literature by Portal [1] in 1685. This term is applied to a wide range of congenital anomalies most typically limb and digital amputation and constriction rings associated with amniotic fibrous bands while in utero. They may be associated with visceral and cutaneous anomalies [2]. Its incidence varies from 1:11,200 live births while in cases of abortions and stillborns the incidence may rise to 1:1200 [3]. In Indian literature, the incidence has been given as 1:70 in abortions and 1:1300 to 2,000 births [4]. This anomaly is typically formed by wrapping of amniotic band around limbs or it can also wrap around the head or umbilical cord becoming life threatening for the fetus. Following theories have been postulated.

Amniotic disruption (Exogenous) theory

This amniotic band theory of Torpin [5] is based on exogenous nature of defects that result from rupture of amniotic sac like in cases of leaking or amniocentasis or during fetal therapy for TTS. According to this theory band entraps various parts of the fetus and disturbs normal development. Earlier occurrence is associated with severe malformations like craniofacial and visceral. Bhat et al in their case report have supported this theory [6].

Vascular disruption theory

It has been proposed by Van Allen [7] which suggests that vascular disruption is the cause of various malformations.

Embryonic dysplasia theory (endogenous)

This theory was presented by Streeter in 1930 which suggests that fetal disruptions result from imperfect histogenesis [8]. Considering multiple anomalies the possibility of disturbed organogenesis in embryonic development due to teratogenic agents should be thought of with positive history.

ABS is often difficult to detect before birth. However indirect findings on USG like constriction and swelling of limbs can be suggestive of ABS. 3D USG and MRI can be used for detailed study of detection of bands and resulting fetal damage. Fetal surgery may be considered in advanced centers for fetus in danger. However in developing countries treatment is usually given after birth in form of reconstructive and plastic surgery. The prognosis depends on the location and severity of the constricting bands. Every case is different and hence requires an individualized line of treatment.

CASE REPORT

22 yr Unregistered Primigravida was admitted with history of 8 months of amenorrhoea in active phase of labour. She was of low socioeconomic status and did not take ANC treatment neither got any routine investigations including USG. Menstrual history was not significant. She gave history of high fever in first trimester and was in habit of tobacco chewing, also her elder sister gave history of stillbirth with baby having some limb anomaly. Our USG findings showed SLIUG of 36wks with vertex presentation. PV findings suggested 8cms cervical dilatation and 80% effacement. She progressed normally and delivered male child of 2.3kg which cried immediately after birth. To our surprise baby showed following abnormalities suggestive of ABS as shown in figure 1.1,1.2,1.3.Undeveloped left upper limb with rudimentary bud, right ring and little finger showing constriction bands, both lower extremities showing constriction bands. In addition hypospadiasis was noted. Placenta and cord were found to be normal. Further screening of the neonate revealed no hematological or structural anomaly on USG and 2D Echo. Baby was handed over to paediatric surgeon for further management.

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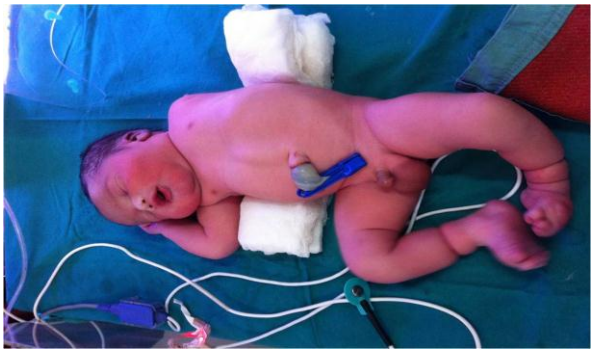

Figure 1.1

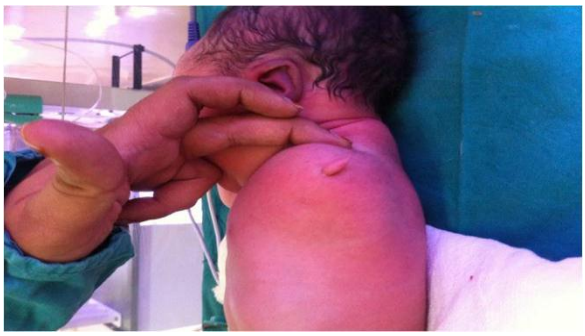

Figure 1.2

Figure 1.3

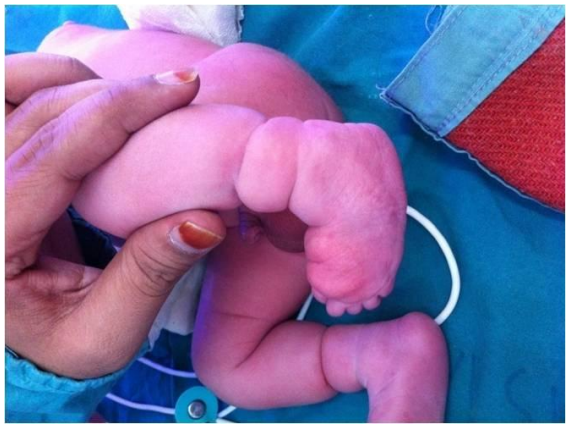

Figure 1.4

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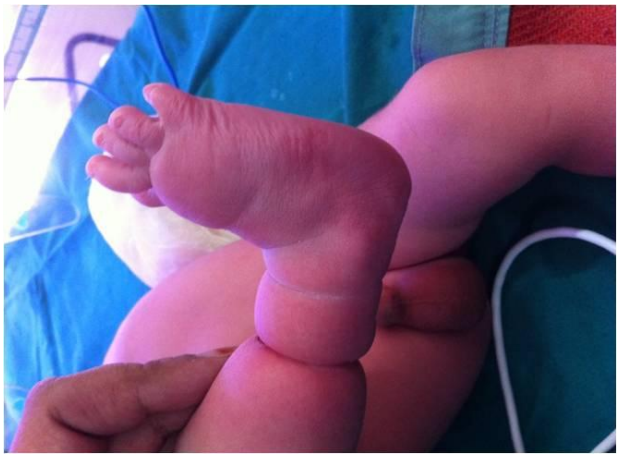

Figure 1.5

DISCUSSION

As she was an unregistered case, this type of multiple malformations were shocking and rather surprise to us. However it definitely signifies the importance of antenatal care and complete anomaly scan. The descriptions and photographs given, clearly showed that baby had ABS.

A very detailed review has been taken by Pietro Cignini et al in their article in Journal of prenatal medicine[9].Interestingly some risk factors described by them were associated in this patient which may have played some role in the formation of this anomaly. First and foremost is possibility of genetic predisposing factors because according to past history, first degree relative (elder sister) had delivered a stillborn baby with limb defect. As per Latin and American studies this syndrome shows familial occurrence and risk of ABS is 42.8 times higher than general population, so also IUGR was a common finding in their series like found in our case [9]. In the same review, data from study of Boston suggests that young Primigravida with low socioeconomic status are at increased risk which was also found in our case. Tobacco in any form is a known vasoconstrictive agent suggestive of vascular disruption theory and our patient was a chronic tobacco chewer. Another possibility is history of high grade fever due to viral infection in the first trimester because virus can be teratogenic and hypothermia is implicated as a vascular disruptor. It can be concluded that still many questions remain unanswered and more studies are required for this entity called ABS.

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