

Amniotic band syndrome associated with malformation of extremities

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Abstract

Amniotic band syndrome (ABS) is a rare congenital condition, occurring as an outcome of entanglement of fetal parts with amniotic bands during intrauterine life resulting in anatomical abnormalities. ABS should be considered in every newborn with congenital malformations. In this report, a case of a preterm female baby delivered to a diagnosed (Preterm pre-labour rupture of membranes) PPRM mother, with abnormalities of the extremities due to ABS is discussed.

Key words: amniotic band syndrome, abnormalities, extremities, PPRM, preterm

Introduction

Amniotic band syndrome (ABS) is a rare congenital condition which has an incidence ranging from 1:1200 to 1:15000 in live births,^{1,2,7} 1:70 among still births and as high as 1:56 in abortuses.^{2,6,11} It occurs in both genders equally as an outcome of entanglement of fetal parts with amniotic bands during intrauterine life of the fetus.^{1,2,6,7} ABS commonly affects extremities and abnormalities range from simple ring constrictions to amputations of limb at various levels.⁴ Also this

syndrome causes cranial-fascial malformations such as cleft lip, cleft palate⁸ and encephalocele to acrania,¹ abdominal wall defects and visceral defects for example renal agenesis, and less commonly septo-optic dysplasia.⁴

The diagnosis of the ABS is made with the two-dimensional (2D) ultrasound scan at the end of 1st trimester or beginning of second trimester. The fetal malformations are clearly visualized by the three-

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Dimensional (3D) ultrasound scan (USS).^{4,7} The mainstay of the treatment of ABS is plastic and reconstructive surgery after the birth. Nowadays, there is increase in tendency towards in-utero surgeries for the ABS with advances in imaging techniques.^{4,10}

Case report

A 27 year, blood group B negative primi-gravida, diagnosed with thalassaemic trait was admitted to the hospital at period of gestation (POG) 22 weeks, for the further continuation of conservative management of preterm pre-labour rupture of membranes (PPROM) since 22 weeks of pregnancy. On admission the patient was having a low haemoglobin count of 8g/dl and she was administered several periodic blood transfusions throughout the pregnancy. Although there were no features of chorioamnionitis, patient was given prophylactic antibiotics.

Following admission regular fetal monitoring done. Serial 2D USS revealed reduced liquor, single live fetus

with cephalic presentation and low lying posterior placenta reaching internal os. Her pregnancy was complicated with right side ureteric obstruction due to calculi and rigid cystoscopy with ureteric stenting was done at POG 27. During hospital stay at POG 27+5, patient complained on and off abdominal pain which suggest pre term labour and was sent to labour room where the patient expelled a small chocolate coloured tissue part per vaginally. The tissue part was not sent for histology and abdominal pain settled. Next day patient developed clinical features of placenta abruptio and emergency hysterotomy was planned and delivered a baby girl weighing 965g. During hysterotomy, fine threads like bands were noticed within uterine cavity by the operating team.

At birth APGAR score was 1⁶5⁷10⁷ and on examination congenital abnormalities of limbs; bilateral syndactyly of middle and ring finger (Figure 1 A and B), absent foot in left lower limb (Figure 2), malformed right lower limb and big toe with wound on medial aspect of right leg (Figure 3) were noted.



A



B

Figure 1. A and B syndactyly of middle and ring finger.



Figure 2. Malformed right lower limb and big toe with wound on medial aspect of right leg.



Figure 3. Absent foot on left lower limb.

After birth of the baby , following problems were identified; extreme prematurity (27+5), low birth weight (965g), infant respiratory distress syndrome (IRDS), neonatal jaundice, anaemia of prematurity, umbilical sepsis and bilateral conjunctivitis. Regularly full blood count, c-reactive protein (CRP), serum bilirubin (total and direct), renal function tests (RFT) done. 2D echo which was done on day 3 of birth revealed small patent

foramen ovale (PFO)/ atrial septal defect (ASD). Ultrasound scan on brain was normal. No abnormalities were detected during ultrasound scan of the abdomen. The baby was given prompt and proper care at the hospital.

The baby is now 9 months old while she has undergone skin grafting and doing well with adequate development appropriate for her age.



Figure 4. Nine months old baby after the skin grafting.

Discussion

ABS responsible for 1-2% of the congenital malformations within the general population. ¹ Most of the cases are sporadic and no known genetic origin or recurrence among the siblings or children of the affected adults^{1,2}.

But there is evidence that, the syndrome is found among the families with the connective tissue disorders, namely osteogenesis imperfecta and collagen disorders; Ehlers-Danlos syndrome⁶. The other possible etiological factors for the syndrome are primiparous mother of under 25 years of age ², unplanned pregnancy³, direct abdominal trauma ^{1,2}, induced¹ or unsuccessful abortion,² intrauterine contraception ², amniocentesis^{1,2}, malformations of the uterus ², cerclage⁶ maternal exposure to drugs such as ergotamine, misoprostol² and non-steroidal anti-inflammatory drugs³, bacterial infection of the amniotic membranes, oligohydramnios³ hyperthermia³ and prematurity⁶. According to literature, considerable amount of cases were related with PPROM^{12,13}.

Though the exact cause for the ABS is unknown, there are two main theories which widely used to describe the syndrome^{2,9}. In 1930, Streeter has explained the

intrinsic model for ABS which suggests that amniotic bands and the anomalies have common origin from subcutaneous germplasm^{2,6,9}. From this model, cranio-facial and abnormalities of internal organs can be explained. In 1965, Torpin has introduced the extrinsic model which conveys that the defects are formed due to rupture of amnion at early pregnancy, suggestive loss of amniotic fluid and extrusion of the fetal parts into the chorionic cavity which leads to forming of amniotic bands^{2,4,6,9}. These amniotic bands can cause limb abnormalities in the fetus for example, entanglement of the limbs by amniotic bands causing constrictions⁹.

According to the timing of the amniotic rupture, the possible malformations of the fetus vary. If the amniotic rupture occurs at early stages, within 45 days, it can lead to severe cranio-facial and visceral malformations of the fetus⁶. Cranio facial defects can differ from asymmetric facial clefts, orbital defects (anophthalmos, microphthalmos, enophthalmos), corneal abnormalities, central nervous system malformations (anencephaly, asymmetric meningocoele) and calvaria effect^{2,6}. ABS affects extremities, mostly to the upper limbs. The malformations for example; constriction rings, digit amputations, amputation of the limbs syndactyly, foot

deformities, hypoplasia of digits and peripheral nerve palsies^{6,8}. ABS can also lead to abdominal wall defects with organ extrophy and heart extrophy due to chest wall defects⁶.

Amniotic band syndrome can be detected at prenatal ultrasonography by visualizing the amniotic bands attached to fetus^{3,5}, constriction rings on extremities, amputations of the fingers or toes and terminal syndactyly⁶. Nevertheless, due to severe oligohydramnios makes prenatal detection difficult. The diagnosis of amniotic band syndrome postnatally is based on physical examination of the baby⁶. But additional radiological investigations; x-ray, ultrasound scan are needed for the visualization of malformation of some potential organs⁶.

The mainstay of the treatment for the ABS is plastic and reconstructive surgery after the birth of the baby⁴. Foeto-scopic laser cutting is a novel developing method for the treatment of amniotic band syndrome prenatally minimizing the harmful effects due to constrictions from amniotic bands⁶.

On a negative note regarding the relevant case, during the preterm labour at POA of 27, the patient has expelled a chocolate coloured tissue part per vaginally. But it was not sent for histology, which may have been an evidence for amputated foot due to amniotic band syndrome. In our case, the patient was diagnosed with PPRM for a month duration and due to oligohydramnios the visualization of the amniotic bands was difficult. Hence, antenatal diagnosis of the amniotic band syndrome was not made.

In a positive way in regard of the case, the amniotic band syndrome was diagnosed with the visible physical findings such as bilateral syndactyly of middle and ring finger (Figure 1A and B), absent foot in left lower limb (Figure 2), malformed right lower limb and big toe with wound on medial aspect of right leg (Figure 3). Along with the supportive care, for this baby an eye referral was made because of retinopathy of prematurity. For the future treatment of the baby immediate referral to plastic surgery made and planned for referral to physiotherapy.

Conclusion

In a summary, Amniotic Band Syndrome is a rare condition but should be included as a differential

diagnosis in most of congenital malformations of newborn. The post natal physical examinations with other supportive investigations are of diagnostic value in amniotic band syndrome.

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