A RARE PRESENTATION OF CONGENITAL HEMOLYTIC ANAEMIA IN AN INFANT WITH AMNIOTIC BAND AND MENINGOCELE

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Abstract

Amniotic band syndrome (ABS) is a rare condition, potentially associated with a variety of different birth defects. It is also known as ADAM complex (amniotic deformities, adhesion, mutilation), amniotic band sequence, amniotic disruption complex, annular grooves, congenital amputation, congenital constricting bands, Streeter bands, Streeter anomaly, transverse terminal defects of limb, aberrant tissue bands, amniochorionic mesoblastic fibrous strings, and amniotic bands. The severity of amniotic band syndrome can range from a single, isolated finding to multiple, disfiguring complications. The arms and legs are most often affected. Up to 50% of cases have other congenital anomalies including cleft lip, cleft palate, and clubfoot deformity. The head and face and, in some cases, various internal organs can also be affected.

We present a case of an infant born with a closed meningocele along with an amniotic band. Although there have been several case reports of encephalocele, anencephaly, meningocele and a case of tethered cord associated with amniotic band syndrome (ABS), there has yet to be a report of a meningocele associated with ABS having congenital hemolytic anaemia.
constricting the left hand and a lumbar meningocele (fig 1). On examination the child was having amputation of second toe of left leg, syndactyly, hypoplasia of the digits (fig 2, 3 and 4), severe pallor along with liver enlarged 5 cm and spleen enlarged 3 cm. There was a history of blood transfusion 6 weeks back, reticulocyte count was high along with peripheral smear suggestive of hemolytic anaemia. There was no history of fever. With this a provisional diagnosis of congenital hemolytic anaemia was suspected. Sickling test was negative and HPLC showed high levels of fetal hemoglobin. Thus confirming the diagnosis as Congenital hemolytic anaemia most probably thalassemia major in an amniotic band syndrome with meningocele.

The baby was given another blood transfusion as the Hemoglobin was less than 7 and given folic acid for long term use. The parents were advised to repeat HPLC after 3 months as this time the report was done within 6 weeks of blood transfusion. The baby was planned for surgical correction of meningocele after the age of 1 year.

**Discussion**

Amniotic constriction bands are strands of the amniotic sac that surrounds a baby in the womb. They may cause a congenital deformity of the face, arms, legs, fingers, or toes. Although there are many unanswered queries regarding congenital constriction band syndrome, studies regarding its cause have come a long way. There are two main theories. Widely accepted “extrinsic model” theory, proposed by Torpin et al. in 1968 explains, the rupture of the amnion without the rupture of the chorion leads to transient oligohydramnios due to loss of amniotic fluid through the initially permeable chorion. The fetus passes from the amniotic to the extra embryonic coelom through the defect and comes in contact with ‘sticky’ mesoderm on the chorionic surface of the amnion. This leads to entanglement of
the fetal parts and skin abrasions. Entanglement of the fetal parts causes constriction rings and amputations, whereas skin abrasions can lead to disruption defects, such as cephaloceles. Further, swallowing of the bands will cause asymmetric clefts on the face. 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.

Amniotic band syndrome has very polymorphic clinical findings. Early amniotic rupture, during first 45 days, leads to the most severe cranio-facial and visceral malformations.[11] The most common finding in the amniotic band syndrome is the constriction rings of the fingers and toes.[12] It occurs in 77 percent cases with multiple anomalies.[13] In our case, there were constriction rings of the soft tissues accompanied by distal edema of the right hand. Other associated features of ABS are shortening of the limbs or intrauterine limb amputation, amputation of the digits (most often 2nd, 3rd and 4th fingers) and toes, similar to our case where there was amputation of second toe of left leg, syndactyly, hypoplasia of the digits, foot deformities, pseudarthrosis, peripheral nerve palsy. If bands compress the fetal head or face, different cranio-facial disturbances appear like asymmetric face clefts, orbital defects (anophthalmos, microphthalmos, enophthalmos), corneal abnormalities, central nervous system malformations (anencephaly, encephalocele, asymmetric meningocele), like in our case having lumbar meningocele. Amniotic bands can also cause abdominal wall defect and abdominal organs extrophy, chest wall defect with heart extrophy, umbilical cord strangulation with often lethal outcome. Amniotic rupture and consecutive oligoamnios can cause deformities such as metatarsal clubfoot, scoliosis, hip dislocation by mechanical pressure on the fetus. Because of such a wide spectrum of possible anomalies and many combinations of their simultaneous appearance, there are no two identical cases of ABS. Beside all previously mentioned malformations caused by amniotic bands itself, a subset of cases manifest additional findings that are not consistent with that mechanism, such as congenital heart defects, renal anomalies, hemangiomas, imperforate anus, polydactyly, septo-optic dysplasia, typical cleft lip and palate. Like in our case there is congenital hemolytic anaemia.

There is no proven causality association between amniotic band meningocele and congenital hemolytic anaemia. Thalassemia being a genetic defect, it presents more in association with amniotic band alone.

In severe cases experts from Orthopedics, Occupational Therapy, Orthotics and Prosthetics, Plastic Surgery, Neurosurgery, Craniofacial and other areas of healthcare are needed for better functioning of child. Plans should be made for careful delivery and management of the problem after birth. Our child presented with amniotic band sequence with very rare association of lumbar meningocele with no neurological defect and congenital hemolytic anaemia.

**Conclusion**

Amniotic band syndrome is an accidental event with no genetic relationship. There is no chance of recurrence in subsequent pregnancy. If we are getting any case of ABS first of all we have to search any associated finding. If constrictions are not so deep, no surgical treatment needed. Baby may need urgent surgery right after birth if bands press deeply on blood vessels or nerves, or if they cause serious swelling (lymphedema) or other problems. If not, we can wait until the baby is stabilized.

In our case as there was no neurological deficit, meningocele can be repaired on elective surgery basis. However thalassemia will need life long blood transfusion, folic acid supplementation and after 3 years iron chelating agents.

**Conflict Of Interest**

There was no conflict of interest and no funds received.

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References