AMNIOTIC BAND SYNDROME – A HOSPITAL BASED FETAL STUDY

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1. Introduction

Amniotic band syndrome (ABS) is a group of sporadic congenital anomalies of varying severity involving limbs, craniofacial region or trunk [1]. Congenital bands that occur as a result of rupture of amniotic sac usually before 12 weeks of gestation are called amniotic bands (2). The estimated incidence of ABS is 1 in 1200 to 15,000 live births [3 & 4]. The malformations range from mild deformities to severe anomalies that are incompatible with postnatal life. Although a wide range of malformations can occur in this syndrome, characteristic features can often suggest the diagnosis [2]. Amniotic band leads to secondary effects on the fetus producing malformation due to interruption of normal morphogenesis, deformation due to distortion of established structures and disruption of structures already formed [3].

MATERIALS AND METHODOLOGY

The present study was conducted on 1000 humans fetuses (since Jan 2008 - October 2015) in the Department of Anatomy, Government Medical College & Hospital, Chandigarh. Fetuses were sent by the Department of Obstetrics & Gynaecology of the same institute for routine autopsy. In our present study the incidence of amniotic band syndrome was 0.8% (out of 1000 fetuses ABS was noted in 8 fetuses). Associated anomalies were noted in each case. Prenatal diagnosis of ABS is important to detect and avoid further complications in life.

Observation

Out of 1000 autopsies, amniotic bands were noted in 8 fetuses (0.8%)

| TABLE -1 |
| Incidence of amniotic band syndrome |
| Male | Female |
| 7(87.5%) | 1(12.5%) |

| TABLE -2 |
| MODE OF FETUS OBTAINED |
| Mode of fetus obtained | No of cases(%) |
| Spontaneous abortion | 4 (50%) |
| Induced /therapeutic abortion | 3(7.5%) |
| IUD | 1 (12.5%) |

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### Table 3

<table>
<thead>
<tr>
<th>Associated anomalies</th>
<th>No. of cases(%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Musculoskeletal defects</td>
<td>7 (87.5%)</td>
</tr>
<tr>
<td>Neural tube defects</td>
<td>4 (50%)</td>
</tr>
<tr>
<td>Cardiovascular defects</td>
<td>1 (12.5%)</td>
</tr>
<tr>
<td>Respiratory defects</td>
<td>0</td>
</tr>
<tr>
<td>Urogenital defects</td>
<td>0</td>
</tr>
</tbody>
</table>

### Figure 1a

- AB seen at tip of fingers with constriction in distal phalanges

### Figure 1b

- Common band joining right hand & left leg

### Figure 3

- Amputation of great toe of right foot with other toes fused.
- AB are seen at tip of toes.

### Figure 4

- Figure showing split mandible, encephalocele, and cleft in left upper eyelid

### Figure 5

- Bilateral club foot
AMNIOTIC BAND SYNDROME: A REVIEW

Discussion

Amniotic bands, also called constriction bands, congenital rings, Streeter dysplasia and annular defects are anomalous bands that occur due to rupture of amniotic sac. These strands encircle the fetus or parts of fetus either partially or completely. Montgomery in 1832 was the first one to describe the amniotic constriction band.

ABS is an uncommon congenital pathological condition that leads to malformation and fetal death. Amniotic bands occur in 1 of every 1,200-15,000 births and are demonstrable in 1-2% of malformed infants. According to Kulkarni et al the triad of ABS includes amnion-denuded placenta; foetal attachment or entanglement by amniotic remnants; foetal deformation, malformation/or disruption.

Studies had found out connection between ABS and mothers age, especially primiparas under the age of 25 are affected more. In our findings 7 cases were under the age of 25 & only one case was the age 29.

Previous studies had mentioned that the most frequent organs involved in ABS are the fingers and toes with or without association with deformed lip and palate. In our present study in almost all cases amniotic bands was seen associated with fingers and toes. One of the cases there was amputation of index finger. In another case there was amputation of great toe and amputation of 3rd and 4th toe.

Literatures had mentioned that ABS does not have genetic origin, and there is no recurrence of siblings or children’s of affected adults. In our present study also we never came across the recurrence of ABS in affected adults.

Lockwood et al mentioned the role of a teratogenic insult as one of the causative factors of ABS. Where as Ossipoff and Cigninni suggested that different ethiological factors including maternal drug abuse, cigarette smoking, unplanned pregnancy, anorexia, hyperthermia, young maternal/paternal age, dietary glycemic index, abdominal trauma, infections, amniocentesis etc. as causation of ABS. In the index cases, there was no history of maternal exposure to teratogens before or during pregnancy. Experiments on mice had proved that the amniocentesis can be a major cause of ABS. Walker and Fraser (1956) were able to produce facial cleft in mice by removing amniotic fluid on 13th day of gestation (mice). Poswillo supported them by his experiments on rats, he reported a incidence of 100% of cleft lip and cleft palate on rats undergoing an amniocentesis at 15½ day of gestation. He was also able to demonstrate the occurrence of micrognatia, ring constriction, talipes, syndactyly, amputation, phocomelia etc in such cases. However, Vrettos et al mentioned that very small quantity of amnion removed does not cause any hazard unless the infant is actually impaled by the amniocentesis needle.

There are many theories proposed to explain the occurrence of this syndrome. Streeter’s (1930) intrinsic hypothesis attributes that the development of amniotic band syndrome to impaired blood flow to specific parts of the developing fetus (vascular disruption or compromise). Even though he could not explain the exact reason for the impaired blood flow, he suggested that in areas where the blood flow is poor, injury occurs to walls of the blood vessels of the fetus. This leads to bleeding (hemorrhaging) and tissue loss in the affected areas, which in turn results in the varied symptoms associated with the disorder & he proposed that presence of constriction bands as a secondary effect of this impaired blood flow and subsequent damage to the fetus.

Whereas Torpin’s (1965) extrinsic hypothesis, which is the most accepted theory suggested that ABS occurs due to rupture of inner layer (amnion) of amniotic sac. As a result of this rupture the fetus get exposed to strands of fibrous tissue that float in the amniotic fluid or remain partially attached to the amniotic sac. These bands could disrupt the normal development of an embryo or fetus. They can wrap around or entangle (constrict) the fingers, toes, arms, legs and other parts of the developing fetus, leading to minor constricting rings, lymphoedema and sometimes even amputation of digits. If the amniotic bands are still partially attached to the amniotic sac, they may wrap around a fetal body part and tether (anchor) that body part to the amniotic sac. This can restrict movement and proper development of an embryo or fetus. According to Moerman et al (1992) ABS is a collection of three distinct lesions-constrictive tissue bands, amniotic adhesions and the more complex limb-body wall complex. He suggested that adhesive amniotic bands are different from constrictive bands morphologically and pathogenetically as they caused severe defects like encephalocele and facial clefts, the latter are linked to limb anomalies. In our present study, we had one case with encephalocele and another case with oblique facial cleft.

Ho DM & Huang CC classified the disorders of ABS based on timing of disruption and the anomalies.

Group I- comprised anomalies such as anencephaly, craniofacial clefts and limb body wall complex; the timing of disruption being in the early embryonic period - <4 weeks post conception (pc) or 3-6 weeks pc.

Group II- comprised cleft palate, cleft lip, CHD, limb reduction, the insult occurring in the mid embryonic period (4-7 weeks pc).

Group III- comprised of oligohydramnios and Pierre Robin sequence (late embryonic to early foetal period - 7-12 weeks pc).
According to our present investigation we had
- 3 cases coming under Group 1 which included anencephaly, encephalocoele, cleft in the mandible and cleft in upper eyelid
- 1 case under Group II comprising oblique cleft lip
- 1 case under Group III including oligohydraminos
- 8 cases under Group IV with constriction bands in all cases and amputation of digits in 3 cases

Goldfarb et al mentioned that the most common problems that were associated with amniotic band syndrome are cleft lip/palate and clubbed foot. In our present study we came across 2 cases of bilateral club foot and a case of oblique cleft lip.

According to Patterson’s diagnostic criteria for ABS, should at least fulfill one of the following criteria: 1) simple ring constrictions, 2) ring constrictions with distal deformity plus or minus lymph oedema, 3) ring constrictions accompanied by syndactyly or acrosyndactyly, 4) amputation. Our present cases fulfilled the above criteria with simple constriction rings in almost all cases, amputation of digits in 3 cases, and syndactyly in 3 cases.

Allington et al. had mentioned that anomalies of internal organs in infants with ABS are very rare. We in the index cases also never came across any internal organ anomaly.

Higginbottom et al. suggested that early rupture of amniotic sac (early insult) before 45 days gestation produced severe facial clefts and severe defects of the brain and calvarium, while late rupture (after 45 days gestation) resulted in limb involvement with no facial clefts or CNS involvement. In our present study, out of 8 cases one case showed encephalocoele, cleft in left upper eyelid, split mandible, bilateral club foot (figure 4 & 5) and another case with placenta & umbilical cord attached to head, brain matter & degenerative tissue covered with skin, Oblique facial cleft on right side. Both these cases can be included under early insult since they correlate with the above mentioned features, where as rest of the cases can be included under late insult as it included only limb defects.

Musculoskeletal disorders commonly associated with ABS includes club foot, syndactyly or acrosyndactyly, hypoplastic nails or fingers, pseudoarthrosis of underlying bones, peripheral nerve defects, distal lymph oedema, amputation of digits, deformed lip and palate and umbilical hernia. In our present investigation, we also came across syndactyly, acrosyndactyly, club foot, lymphoedema cleft lip etc.

Ashutosh Halder in an article described about a case of a male fetus with multiple congenital malformations. He observed that the placenta was attached with head and face resulting in severe disruptive effects of the face (clefting) and head (anencephaly) along with thoracoabdominoschisis. The left thigh had a constriction ring, thoracic and abdominal organs showed several anomalies including generalized hypoplasia32. In our study on ABS, we came across a case of amniotic band syndrome in which placenta and umbilical cord was attached to head, brain tissue along with degenerative tissue was covered with skin and was seen continuing with placental membrane. We also observed a swelling measuring 4.5×5 cm extending from upper part of face and head on the right side. Rt. eye was displaced laterally, oblique facial cleft was seen on right side. AB was seen at fingers of right hand, little finger was swollen, and ring finger was flexed (figure 6a, 6b).

McKenzie stated that since ectodermal placodes were involved in the formation of many organs and structures including the neural tube, nose, branchial arches, ventral body wall and limbs. Maldevelopment of these ectodermal placodes could result in various anomalies for example malfunction of limb bud placodes may result in the limb reduction anomalies and constriction. Likewise the involvement of rostral placodes results in neural tube defects and facial cleft 33. Harris et al. described acalvaria as a postneurulation defect, calvarial bones, duramater and associated muscles are formed as a result of migration of the membranous portion of the neurocranium under the calvarial ectoderm after the closure of the anterior neuropore. In acalvaria migration of the membranous neurocranium apparently does not occur. This could explain the reason for encephalocoele and anencephaly 34. We in our present study on ABS had one case of encephalocoele and one case of anencephaly.

Researchers have speculated that trauma to the abdominal area during pregnancy may play a role in some cases. Blunt trauma to the placenta has been shown to cause amniotic band syndrome in some cases 35.

Prenatal diagnosis of ABS is difficult. ABS should be excluded when gross fetal abnormalities are detected by ultrasound or radiography. Diagnosis of cranial bone defects can be established by ultrasonography early in the second trimester of pregnancy after mineralisation of the skull bones has been completed 12. However cases of acrania can be diagnosed even in the first trimester of pregnancy, with extensive use of transvaginal sonography. Anencephaly and exencephaly, both of which are accompanied by acrania, are the immediate differential diagnosis as facial structures and base of the brain are intact in both. As a result of slow degeneration of the unprotected brain secondary to mechanical and chemical trauma on exposure to amniotic fluid, most cases of acrania can eventually progress to anencephaly 36. In acalvaria, the usually intact over-lying skin protects the brain against this process 34.

Amniotic band syndrome can be diagnosed prenatally by ultrasound which can show amniotic bands, congenital malformations, oligohydramnios. The most important diagnostic criteria are visible amniotic bands, constriction rings and irregular amputations of toes or fingers. Three dimensional and four dimensional ultrasound and MRI contribute to more sensitive prenatal diagnostics of amniotic band syndrome 37. ABS can be treated in utero by fetoscopic laser surgery before the bands can compress fetal parts 38.

REFERENCES: