Case report

Amniotic band syndrome- A rare case report

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ABSTRACT

Amniotic band syndrome (ABS) is a rare condition in which there is entrapment of the growing foetus or its organs by the band of tissue formed from the amniotic membrane. We received 20 weeks' gestation-old female foetus for pathological examination. On external gross examination foetus was detected with only the thumb in the right hand with the absence of the remaining four fingers. On thorough examination there was membranous structure attached to the right hand running towards the right leg and partly attached to the toes. This on microscopy showed fibrosis. ABS is a rare syndrome with varied presentations which sometimes can endanger the life of the foetus. Though exact aetiology is unknown till date various risk factors are attributed to the condition.

Keywords: Amniotic band syndrome; constriction ring; autoamputation; fetoscopy.

INTRODUCTION

Amniotic band syndrome (ABS) is a rare condition in which there is entrapment of the growing foetus or its organs by the band of tissue formed from amniotic membrane. That's why it is also known as constriction band syndrome. The other names like congenital annular constrictions, congenital ring constrictions, and intrauterine amputation are also used synonymously in the literature. Sometimes this can cause ischemic damage to the growing foetal parts resulting in auto amputation (1).

The reported incidence of ABS varies from 1 in1, 200 to 1 in 15,000 live births (1, 2). There is no gender preference seen. Some sort of abnormal antenatal history is observed in more than half of cases (3). Here we report a case of ABS which was undetected antenatally and diagnosed on post mortem examination. Written and informed consent for the publication of the case report obtained from the mother of the foetus.

Case report

We received a twenty-week gestation old female foetus with placenta for pathological examination. The mother was primigravida, 34 years old and had conceived this baby by *in vitro* fertilization. Her routine antenatal blood investigations were within normal limits. Ultrasound NT (Nuchal Translucency) scan done at 13 weeks was also showing normal development of the baby. Quadruple screening done at the second trimester had also shown very low risk/negative results for chromosomal anomalies.

In the placenta large subchorionic clot was present extending to about 40% of the placental disc area (Fig. 1A). There were no areas of infarction visible grossly. Microscopically the subchorionic haemorrhage was confirmed with focal areas showing intra villous fibrin deposition and necrosis. Neutrophilic inflammation was also seen at foci (Fig. 1B).

On external gross examination of the foetus, we detected only the thumb in the right hand of the foetus with the absence of the remaining four fingers. Other extremities were normally developed. All orifices were patented. Internal organs were also grossly unremarkable. On thorough examination there was some membranous structure attached to the right hand running towards the right leg and partly attached to the toes also (Fig. 1C). Microscopic examination of that membrane showed only fibrosis without any viable cellular structures (Fig. 1 D, E).

DISCUSSION

For the first time ABS was reported by Portal in 1685. The aetiopathogenesis for this syndrome was proposed by Montgomery in 1832 (4). Orioli *et al*., in her studies observed that the extremities are more affected by this disease (5). Till date the aetiology of ABS is not understood clearly. Various theories have been proposed by various authors trying to unfold the complicated mechanisms resulting in ABS. In 1930 intrinsic theory was proposed by Streeter which describes that ABS results from intrinsic defects in embryogenesis (6). Contrary to this Torpin in 1965 proposed the extrinsic theory which stated adherent bands formed due to early amnion rupture which detach from chorion and form fibrous bands that can be entangled on limbs (7). This was further supported later by many authors including Kino *et al*., (8) Higginbottom *et al*., (9).

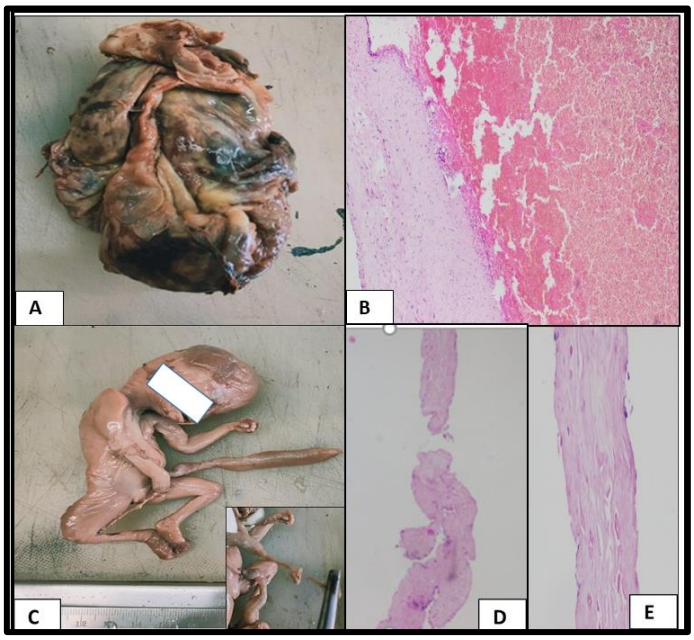

Fig.1: A: Placenta showing subchorionic hematoma. B. Histopathology showing subchorionic haemorrhage (5x, Hematoxylin & Eosin). C: Amniotic band attached to right hand extending to right leg. Inset-Closer view showing the band attachment to the right hand. D: Histopathology of fibrotic band showing fibrosis without any cellular components (5x, Hematoxylin & Eosin). E: High power view of the fibrotic band showing fibrosis (20x, Hematoxylin & Eosin).

Foulkes *et al*. observed that the time of amniotic rupture will decide the spectrum of deformity. Rupture during early weeks of gestation (before 7 weeks) mostly produces limb segmentation defects such as polysyndactyly, syndactyly or limb reduction. Rupture in later weeks causes mechanical obstruction by amniotic bands resulting in clubfoot, hypoplasia, lymphedema, and amputations (10). In our case there was sub chorionic haemorrhage evident on gross examination of the placenta but amnion rupture was not seen.

Patterson explained in 1961 that bands develop due to improper development of subcutaneous tissue. Vascular theory was supported by Van Allen *et al*., (10) Daya *et al*., (11) studied vascular abnormalities in ABS patients and concluded that there is a definite association of ABS with vascular abnormalities which depends on the thickness and depth of the band. They also concluded that though this vascular anomaly appears as an effect of band, possibility of this being the initial causative factor in ABS however cannot be totally excluded. Lockwood *et al*. in their study reviewed 14 cases of ABS in twins and concluded that occurrence in monozygotic twins suggests teratogenic cause (12).

Though the syndrome can present with various clinical features some findings are relatively common in most of the cases which include distal ring constrictions, amputated digit or extremity within the womb, and acrosyndactyly (3, 13).

Studies have shown various risk factors which can enhance the development of this syndrome. This can be antenatal like maternal drug intake, illness, trauma including attempts for abortion in first trimester prenatal factors like low birth weight (<2,500 g), and preterm delivery (less than 37 weeks). There is no solid evidence to suggest autosomal inheritance for this syndrome so far (1). Apart from limb defects other organ involvement is also reported in literature. Cleft lip, orbital cleft, nasal bony deformity, eye defects like ectropion, hypertelorism, corneal opacities are reported. All cranial and neural tube defects like microcephaly, asymmetric encephalocele, hydro-cephalus, and anencephaly are also rarely seen with this syndrome. Internal organs like the heart, urinary system or genital organs can be involved rarely (14). In our case there were no such external defects or internal organ anomalies as noted on gross examination and in antenatal ultrasound scan.

Guzman-Huerta *et al.,* proposed a new classification of the amniotic band sequence phenotypes depending on the anatomic region which are involved. These include a. Craniofacial defect + limb defect, b. Craniofacial defect + limb defect + abdominal wall, spinal column, and/or thoracic defects, c. Limb defect + abdominal wall, spinal column, and/or thoracic defects; and d. Isolated defect (craniofacial, limb, or thoraco-abdominal wall; 15) According to this classification our case falls under the category of isolated defect (only limb involvement).

Devi *et al*., reported a case of amniotic band sequence which had facial clefts, gastroschisis, spinal defects, congenital talipes equinovarus and also abnormalities in the internal organs such as short oesophagus, cysts in the liver and retroperitoneum. Thus, this case had combined features of two or more phenotypes described by Guzman-Huerta *et al.*, which is very rare (16).

ABS should be suspected when classical asymmetric fetal anomalies are detected on ultrasonography

though the presence of fibrous membrane or constriction ring is not so evident (17). Gray *et al*., in their study observed that the prenatal ultrasound scan is less sensitive to detect upper extremity abnormality. Fetoscopy can detect the anomaly with more accuracy (18).

CONCLUSION

ABS is a rare congenital abnormality with varied presentations where in more than half of cases will survive and can be treated with surgical interventions. But a few cases can endanger fetal life though it might be the direct cause or because of associated abnormalities. Our case highlights the role of fetal autopsy in detecting such an entity and documenting the anomalies.

CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

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