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Fetal Study of a Rare Case of Amniotic Band Sequence with Craniofacial Involvement

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Abstract

Background: Amniotic Band Syndrome (ABS) is a rare congenital disorder involving fetal entrapment in strands of amniotic tissue resulting in a broad spectrum of anomalies ranging from minor constriction rings and lymphedema of the digits to complex craniofacial and body wall abnormalities. It is sporadic in occurrence with many theories postulated towards its pathogenesis.

Materials and methods: A 16 week-female fetus with congenital abnormalities was collected from Department of Obstetrics and Gynaecology, RIMS, Imphal after taking permission from concerned authorities and parents. The fetus was examined and observed externally and was dissected for further observation. Radiological investigations (X-Ray and MRI) and histological examinations were also done.

Results: On external examination of the fetus revealed right side anophthalmia, encephalocele on the left side (correlated with MRI), cleft lip with cleft palate, low set ears and acrosyndactyly constriction ring on left little finger and left leg with lymphedema of left foot, fused and malformed toes in right foot, club foot on the right side. On internal examination, asymmetry of cerebral hemispheres was found.

Conclusion: Recognition of amniotic bands is important, as many bands regardless of the etiology was found to be potentially harmful. This case is presented with an intention to understand the etiology and pathogenesis of this condition as well as to heighten the awareness of the amniotic band syndrome, which has implications for genetic counselling for early detection and diagnosis.

Keywords: Amniotic band syndrome, ADAM Complex, Streeter Dysplasia, Constriction rings, Encephalocele, Prenatal diagnosis

Introduction

Amniotic Band Syndrome (ABS) is an uncommon heterogenous group of congenital deformities caused by early amniotic rupture leading to the formation of fibrous bands which entangle or entrap fetal parts in-utero and thereby causes a single or multiple anomalies [1]. ABS also known as constriction band syndrome, Amniotic deformity, adhesions, mutilations (ADAM) complex, Streeter's dysplasia, is a sporadic, non-recurrent condition, with the incidence of 1:1,200 to 1:15,000 live births and no sexual predilection [2].

Fetopathologic studies reveal three types of lesions in ABS: (a) constrictive tissue bands, caused by primary amnion rupture with entanglement of fetal parts (b) amniotic adhesions, between disrupted fetal parts with intact amniotic membrane (c) more complex limb-body wall and craniofacial defects. ABS produces a range of anomalies including craniofacial defects, thoracic or abdominoschisis, amputations, ring constrictions [3].

Risk factors identified are: high altitude, primiparous, young maternal age, obesity, maternal drug abuse, uterine malformations, collagen vascular disease,

history of uterine surgery, complication of amniocentesis sampling [4].

Case Report

In the present study, an aborted female fetus of 16 weeks of gestation with Crown Rump Length (CRL) 16.5cm and weight 200g presenting features of ABS was examined (Figure 1A). Gross examination and radiological investigations (X-Ray and MRI) were done. The fetus had microcephaly and low-set ears on both the sides (Figure 1B).

External examination of the fetus revealed presence of malformed, fused fingers of both hands (Acrosyndactyly) (Figure 2A and 2B). Thumb was spared on both the hands. Left little finger had a constriction ring (Figure 2A). Right foot toes were also fused and malformed. On all the limbs, thin amniotic bands were attached. The fetus had club foot on the right side and rocker-bottom foot on the left side (Figure 2C and 2D). Further examination of the limbs revealed constriction band on the left leg just above the ankle with distal lymphedema (Figure 2C).

Facial features were observed which revealed absence of left eye and bilateral cleft lip along with right oblique facial cleft (Figure 3A). On the left side, there was left unilateral complete cleft palate (Figure 3B). There was also presence of dysplastic nose. A layer of amniotic membrane was found entering the nasal cavity (Figure 3A).

Left frontal encephalocele with attached amniotic membrane was present. After resection of the cerebrum, gross cerebral asymmetry was observed (Figure 3C & 3D). The ventricular system was found to be distorted (Figure 3E). On inspecting the cranial fossa, the right temporal surface of middle cranial fossa was larger than the left with distorted posterior cranial fossa. There was also absence of orbital part of frontal bone and left orbit.

No abnormality was observed in the viscera, spinal column and external genitalia.

Histological Study

The amniotic bands attached were subjected to H&E staining and consisted of fibro-connective tissue and flattened epithelial cells (mesenchymal) along with degenerated squamous cells (Figure 4A).

Histological correlation of the digits with the attached amniotic bands and the constriction band on the left leg was done and discontinuation of the epithelium in the invaginated part was observed (Figure 4B, 4C & 4D).

Both the gross and histological observation of umbilical cord was normal. Microscopically, cerebral cortex and cerebellum were also found to be normal.

Figure 1: A) 16-week old fetus showing features of Amniotic band syndrome. B) fetus with low-set ear (right) and microcephaly.

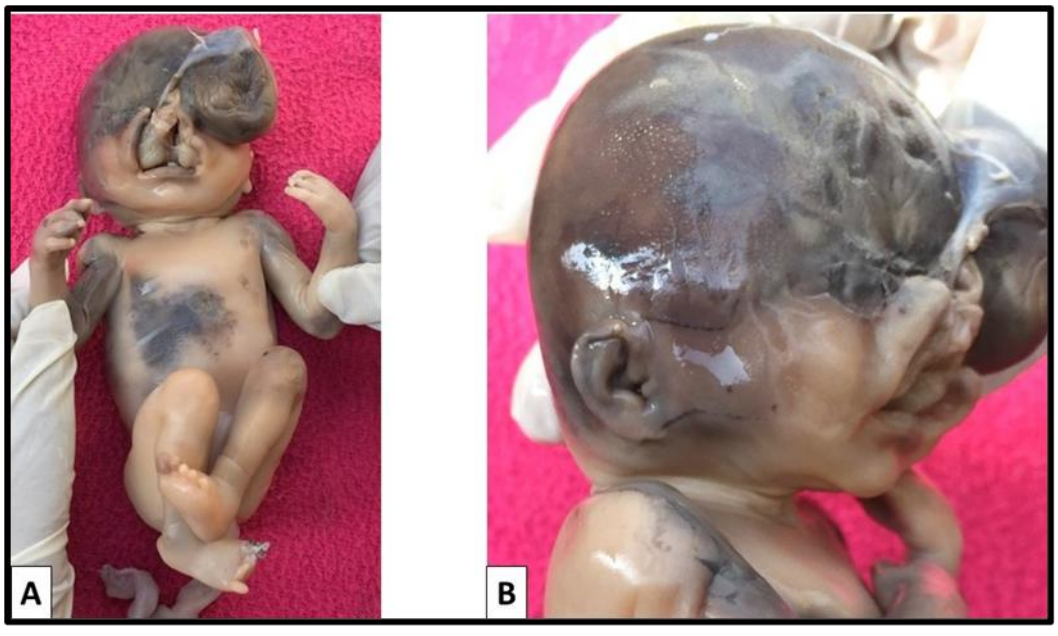

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Figure 2: A) Left hand – acrosyndactyly & constriction ring on little finger (red arrow). B) Right hand – acrosyndactyly. C) Right club foot & left foot constriction ring (black arrow) with distal lymphedema. D) Limb features correlated with the X-Ray.

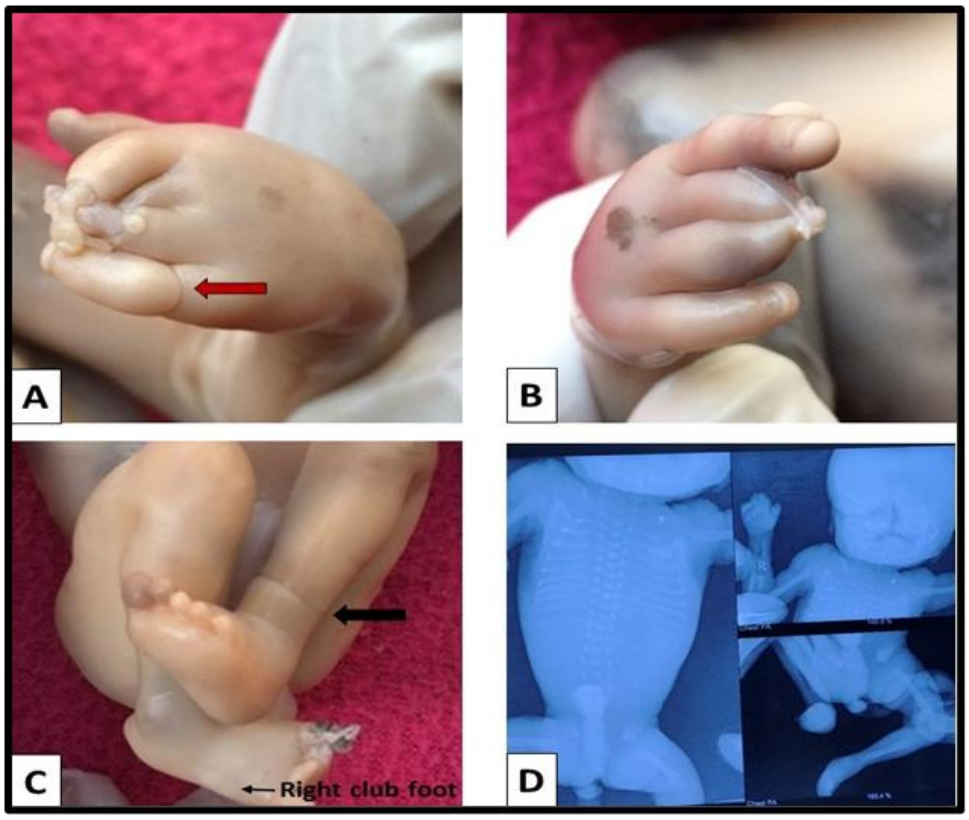

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Figure 3: A) Right encephalocele with cleft lip and palate; amniotic membrane entering the nasal cavity (red arrow). B) Cleft lip and palate on further dissection (black arrow); Rathke's pouch (red arrow). C) Cerebral asymmetry with distorted fissure. D) MRI Axial view – demonstrating cerebral asymmetry. E) MRI Sagittal view – demonstrating distorted ventricles and fissures.

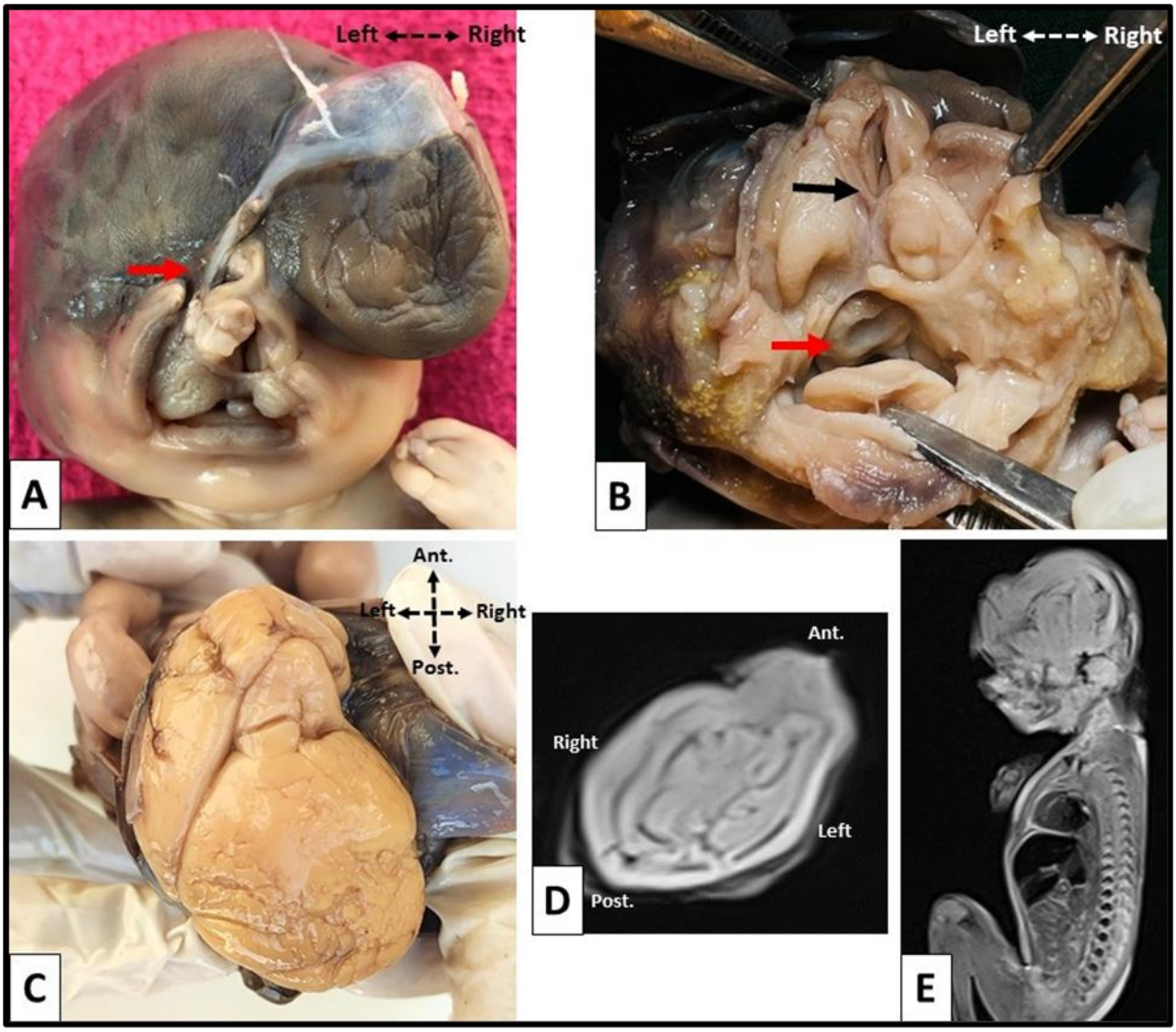

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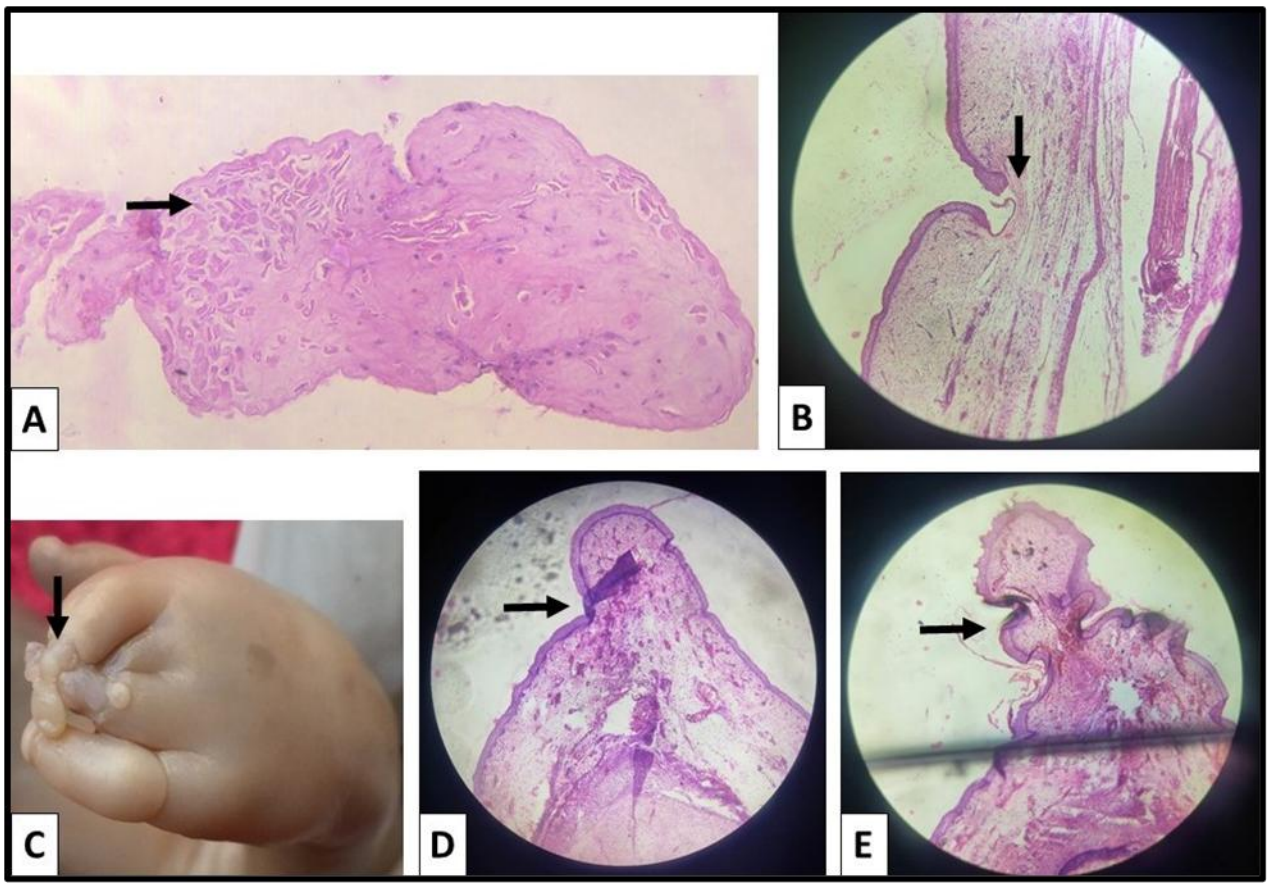
Figure 4: A) Amniotic band with mesenchymal and degenerated squamous cells (H&E, X100). B) Left leg at the site of constriction ring – invaginated part with discontinued epithelium (H&E, X100). C), D) & E) Malformed and fused digits (acrosyndactyly) (H&E, X100).



Discussion

Several theories have been proposed regarding the etiopathogenesis. According to the Intrinsic theory by Streeter [5], anomalies and fibrous band have a common origin because of the improper development of germinal disc in the early embryo. The extrinsic model by Torpin [6] elucidated events involving amniotic rupture followed by loss of amniotic fluid and extravasations of fetal parts into the chorionic cavity.

Lee SH et al revealed through intracranial ultrasound and MRI, bilateral dilatation of lateral ventricles, as the constriction band affected the development of brain and skull [7]. In case of the present study, similar distortion of ventricular system was observed.

Similar to the present study, Yang SS microscopically observed degenerating vernix squamous cells in the fibrous stroma of attached amniotic bands. In this series, pseudosyndactyly, amputation of limbs, midline facial cleft was seen. In contrast to the present study, anencephaly was the severest manifestation of ADAM sequence in this series [8].

There is a predilection for the hand as the site of involvement [9,10]. The central digits are more often involved than the thumb and little fingers due to their separate blood supply and fetal hand positioning [11]. These findings by Kawamura K et al, went hand in hand with the limb involvement in the present study.

Ferris NJ and Tien RD observed abnormal orientation of intracranial structures relative to the cranial bones with deformed and asymmetric cerebral hemispheres [12]. This observation was found to be similar to the present study. It was suggested that the cerebral structures may have been anchored by an amniotic adhesion, when the remainder of the embryo rotated

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90º about its long axis resulting in the malalignment of the cranial contents relative to the craniofacial skeleton [12].

The variability and severity of the defects depends on the timing of the amniotic rupture [13]. Early rupture within 45 days of gestation, leads to the most severe defects such as central nervous system and skull defects, facial clefts, cleft lip and palate and limb anomalies. If rupture occurs after 12 weeks of gestation, constriction of isolated limb parts occurs more frequently [14]. Hence, in the present study the amniotic rupture has probably occurred within 45 days of gestation.

Muraskas JK revealed that the craniofacial lesions such as cleft lip and palate, microphthalmia and encephalocele in ABS are frequently asymmetrical and do not conform to the anatomy of the normal facial clefts [15].

Conclusion

In conclusion, ABS is a rare congenital abnormality resulting in a range of abnormalities from limb to complex body wall defects. Early diagnosis and recognition is essential for counselling parents regarding the etiopathogenesis of the child's birth defects and also provides flexible therapeutic options to reduce complications. Future studies should shed more light on the importance of investigative tools in diagnosing ABS as early as possible.

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