

AMNIOTIC BAND SEQUENCE: A RARE CASE SEEN IN BIRAT MEDICAL COLLEGE

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ABSTRACT

A 21-years-old primigravida gave birth to a Single/Live/Term/Female baby via Lower Section Caesarean Section at our hospital. The mother was supervised, immunized, had uneventful antenatal periods, with no significant medical history. At birth, the baby had no signs of asphyxia with a normal Appearance, Pulse, Grimace, Activity and Respiration score. Physical examination revealed acrosyndactyly in the right hand, left hand at the stage of impending auto amputation with ring-constriction accompanied by deformity of the distal part with lymphoedema, bilateral hyperextension of the knee joints, and simple ring-constriction in lower limbs, rest were within a normal limit. Clinically diagnosed as a case of amniotic band sequence.

KEYWORDS

Amnion; Amputation; Constriction band; Syndactyly



INTRODUCTION

Amniotic band sequence is rare (incidence 1:1200 to 1:15000 live births) and sporadic condition associated with three general types of anomalies: disruptions, deformations, and malformations as a complication of insult to the amnion.^{1,2} Clinical presentation and its severity dependent upon the time of amniotic insult during intrauterine life leading to amniotic constriction band deformities with or without other anomalies.^{1,2} Asymmetric distribution of defects is the hallmark of syndrome.

CASE REPORT

A single live, term, female weighing 3.2kg delivered by 22 years primigravida at 40 weeks of gestation via Lower Section Caesarean Section for breech presentation with non-progress of labor. The mother was immunized, with irregular antenatal care visits, irregular folic acid, and iron supplements, otherwise, her antenatal period was uneventful with no history of trauma, teratogenic medications, diabetes, hypertension, thyroid disorders, recent infection, and substance abuse. Prenatal ultrasound scanning done in the first and third trimesters did not reveal any abnormality. At birth, the baby had no signs of asphyxia with an Appearance, Pulse, Grimace, Activity and Respiration score of 7/10 and 9/10 at 1 minute and 5 minutes respectively. Grossly umbilical cord and placenta seemed normal. Physical examination revealed acrosyndactyly in the right hand, left hand at a stage of impending auto amputation with ring constriction accompanied by deformity of the distal part with lymphoedema, bilateral hyperextension of the knee joints, and bilateral constriction ring band in lower limbs without neurovascular compromise. Otherwise systemic examinations were within the normal limit. The child was neurologically intact with an appropriate response to stimulation and normal feeding and sleep wake cycle. Routine screening investigations were normal. Amniotic band sequence was diagnosed clinically based on constriction ring deformities in a random non embryonic distribution. The baby was admitted at neonatal intensive care unit for two days, but the patient's party refused further systemic investigations and went on against medical advice. On follow up at next day, they said that the baby expired at the age of 3 days of life.



Figure 1: Baby with acrosyndactyly, impending amputation, and constriction bands



Figure 2: Left hand with constriction band with impending auto amputation.



Figure 3: Right hand with acrosyndactyly



Figure 4: Constriction bands in bilateral lower limbs

DISCUSSION

Embryologically, the amniotic cavity is the small cavity developed in the epiblast layer of embryoblast, which is lined by amnion developed from amnioblast derived from epiblast adjacent to the cytotrophoblast.³ So the amnion is the innermost fetal membrane and is essentially fetal epidermis (skin) extending continuously out over the umbilical cord and from there it lines the chorionic cavity forming a closed sac containing the fetus, cord, and amniotic fluid. It is an integral part of the embryo which maintains the structural integrity of the gestational sac by its mechanical strength.

Amniotic Band Sequence may be multifactorial in origin but exact etiology is an ongoing question since the 1930s. However several theories have been given, most known theories are:

- i) Intrinsic theory: defective germ plasm theory given by streeter in 1930.¹
- ii) Extrinsic theory is given by torpin in 1965.²
- iii) Vascular disruption theory is given by van allen in 1981.²

According to torpin's extrinsic theory (the most accepted theory), amniotic band syndrome is the result of fetal entanglement with mesodermic bands from the chorionic surface of amnion. Rupture of amnion (due to idiopathic, maternal disorders like epidermolysis bullosa and connective tissue disorder, or iatrogenic: curettage, fetoscopy, septostomy) results in inappropriate growth of amniotic sac and gradual separation from the chorion.⁴ Amniotic fluid with the fetus exits the amnion and comes to lie within the chorion that may lead to transient oligohydramnios (cause of deformation of extremities like clubbing of feet, angulation of spine). Mesodermic bands form from the chorionic surface of amnion (rarely from chorion) entangle and entrap limbs, digits, cranium, or maybe bowel leading to various deformities depending upon the period of gestation. Pathology in the early gestational period (i.e 25 -45days) leads to Craniofacial Defects, Visceral Defects, Abortion or Prematurity, and in the late phase (i.e 45 days -18 weeks) leads to Limb Constrictions, Digital Constrictions, or Amputations. Early the period of gestation more is the severity and diversity of presentation.¹

Clinical presentations may be disruptive (such as limb defects with constrictions band as a most common feature that may be complicated to lymphedema to autoamputation) or malformative (such as craniofacial defects like encephalocele, anencephaly, cleft lip, and non midline body wall and Visceral defects like Gastroschisis, Omphalocele, Bladder exstrophy, Lung hypoplasia) or deformative (such as spinal defects like scoliosis, joint contractures).² Based on the pattern of deformity distal to the constriction band, Patterson(1961) classification is:

1. Simple ring constrictions.
2. Ring constrictions are accompanied by deformity of the distal part with or without lymphoedema.
3. Ring-constrictions accompanied by fusion of distal parts ranging from fenestrated or terminal syndactyly to "exogenous" syndactyly
4. Intrauterine amputations.⁵

Diagnosis is usually clinical based on constriction ring deformities in a random nonembryonic and asymmetrical distribution of defects. Prenatal diagnosis (as early as in late first trimester) can be made by ultrasound with findings of "constriction rings with or without edema distal portion or amputated limbs", "Thin wispy undulating strands of amnion are seen crossing the gestational sac"-if not seen, we can look for restriction of movement because of its adherence (i.e change of maternal position may float fetus away from uterine wall revealing short band/amniotic band).^{6,7} Perfusion status of distal extremity can be assessed by using color Doppler and measuring pulsatility index. Other measures are Prenatal MRI (T₂ weighted: amniotic bands can be seen as thin wispy hypotense strands) and Mother alpha-fetoprotein may be increased.^{6,7}

Treatment of amniotic band syndrome requires a multidisciplinary approach. The lesions are both static and irreversible, and direct surgical relief is the only approach for limb constrictions. But the timing of surgical intervention and modality of intervention depends upon severity and type of presentation. Very mild case without any functional impairment does not require surgical intervention while acute vascular compromise, severe lymphedema, and distal nerve compression are definite indications for surgical intervention. Staging a surgical intervention is required like excision of constriction band at first stage surgery followed by nerve and distal soft-tissue reconstruction in second stage surgery followed by decompression fasciotomy if needed. But in some cases, one-stage surgery can be done with radical excision of constriction band with soft tissue reconstruction.⁹ With complete circumferential constriction bands, it is recommended that a two-stage correction approach be used. At the first operation, one-half of the circumference is excised and the other one-half can be excised after three to six months.⁹

If diagnosed in prenatal period then fetoscopic surgery and release of constriction band (In color Doppler pulsatility index abnormal but blood flow distal to the constricted area may identify cases suitable for fetal surgery).⁶ Prognosis depends upon the severity of its presentation, from good prognosis with normal life expectancy for the mildly affected infants with only minor digital or limb to death of a baby for severely affected infants with craniofacial involvement.¹

CONCLUSION

Amniotic band sequence is the congenital condition of unknown etiology, related most likely to the single insult i.e. amniotic band disruption leading the unpredictable natural course. Severity depends upon the time of insult, leading to functional complications to mild cosmetic complications only. Which may need a multidisciplinary approach for a better outcome.

CONFLICT OF INTEREST

No conflict of interest

FINANCIAL DISCLOSURE

None



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