



Amniotic Band Syndrome in a Preterm Infant: A Case Report

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Abstract

Amniotic band syndrome is a condition that begins with the premature rupture of the amniotic membrane and is characterized by an asymmetric destructive fetal process and is a broad spectrum that can cause limb amputation, craniofacial and visceral defects from a simple constriction ring. In this article, a 25-week-old preterm newborn case with amniotic band-related defects in the upper and lower extremities, born from a mother with a history of irregular use of lithium and biperiden hydrochloride with the diagnosis of bipolar disorder, is presented because there is no need for amputation with the early surgical method. The possible causes of this syndrome, whose etiology is not fully determined, are discussed.

Keywords: Amniotic band; Amputation; Intrauterine

Introduction

Amniotic Band Syndrome (ABS) is a disorder that may be limited to a simple constriction ring; or may cause limb, craniofacial and visceral defects, severe fetal structural anomalies, and fetal death [1]. Amniotic band syndrome constitutes 1% to 2% of physical malformations. Its etiology remains controversial [1]. Although its incidence is seen equally in both genders, there is no genetic predisposition. Diagnosis of neonatal amniotic band syndrome is often difficult and only 29% to 50% of cases can be diagnosed accurately. It is diagnosed at ultrasonography in the antenatal period and by examination, X-ray, and magnetic resonance in the postnatal period. Antenatal diagnosis and in utero intervention are critical [2,3]. Each case requires a multidisciplinary approach along with a specific approach. The presence of linear hyper echogenicity in obstetric ultrasonography, which is stimulating for the amniotic band, suggests not only the indication of the amniotic band, but also synechia, some uterine anomalies, and chorioamnionitis membrane separation. The antenatal course depends on the organ affected and the severity of the damage to the organ. Depending on the ultrasonographic findings, merely follow-up can be planned, and in some cases, even the decision to terminate the pregnancy may be taken due to the severity of the lesions. In terms of in utero treatment, the cases that will undergo fetal surgery should be selected well (due to the risk of maternal and fetal complications secondary to fetal surgery) [4-6]. In this article, a premature case born of a mother with a history of drug usage due to bipolar disorder, diagnosed with amniotic band syndrome with diffuse deformities in all extremities, and whose fibrous bands were surgically released on the postnatal third day, is reported.

Case Presentation

A male baby born with a bodyweight of 1,010 grams at 25 weeks of gestation from the first pregnancy of a 27-year-old mother, according to her Last Menstrual Period (LMP), was monitored in our neonatal intensive care unit after stabilization was ensured by intubation in the delivery room. In the antenatal history, Premature Rupture of Membranes (PROM) longer than 72 h and chorioamnionitis were detected. It was reported that she used levothyroxine due to the diagnosis of hypothyroidism and Biperiden hydrochloride due to the diagnosis of bipolar disorder, and she received lithium treatments whenever she admitted emergency department due to a bipolar attack. In the physical examination of the patient, with a birth weight of 1010 g (>90 p), a height of 30 cm (10-50 p), and head circumference of 23.5 cm (50 p), was found to be circularly compressed with a fibrous band starting from the distal left forearm, and diffuse edema and circulatory disorder were detected distal to the band. It was observed that the left lower limb had an extremely edematous appearance with more than one fibrous band starting from the distal of the cruris 1/2, circular band traces of the fibrous band were observed at the level of the proximal 1/3 of the proximal phalanx of 3 fingers to 4 fingers on the right upper limb, there were three toes on the right lower limb and it

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Figure 1: Fibrous band and excessively edematous appearance of the patient's upper and lower limbs.

was divided into two parts by a fibrous band distal to the middle toe. Other system examinations were considered naturally associated with prematurity (Figure 1).

Due to chorioamnionitis and EMR, blood culture was taken and empirical ampicillin and gentamicin treatment was started. A dose of surfactant was administered with the diagnosis of RDS. Arterial and venous Doppler Ultrasonography (USG) was performed for all extremities by elevating the extremities due to increased circulatory disorder and color change in the left hand and leg after birth. In Doppler examination, spontaneous flow and colored fillings of all veins were normal, vessel diameters were within normal limits, and no appearance compatible with thrombosis was detected in the lumens. Pentoxifylline was included in the treatment. Skin changes and edema due to band compressions in the extremities immediately after birth were followed up in the early period. Considering the general condition of the baby, surgical intervention was not considered since there was no significant circulatory disorder in the first two days. However, considering the increase in edema in the extremities and the onset of discoloration, the operation was performed by the plastic surgery team on the third postnatal day, and the band structures surrounding the left wrist of the patient were released by circular incision. Band compression was released with multiple Z plasties. In the same session, the superior and deeper band structures of the left ankle, were released by circular incision and multiple Z plasties (Figure 2). After the fibrotic band release procedures, the edematous appearance decreased significantly and extremity circulations were clinically normalized (Figure 3). There was no circulatory disturbance at discharge and active extremity movements were present. There was no circulatory disturbance at discharge and active extremity movements were present (Figure 4). Pentoxifylline treatment was discontinued. Cranial USG taken before, after surgery and during the follow-up of the patient was normal and no hemorrhage was detected.

In the echocardiographic assessment, the patient, who had no additional pathological findings other than Patent Ductus Arteriosus (PDA), was given two courses of medical PDA closure treatment. No Retinopathy of Prematurity (ROP) was observed in the examinations performed for ROP. He was monitored on invasive respiratory support for 18 days and noninvasive respiratory support for 35 days. Full enteral nutrition was started on postnatal 38th day. Oxygen support was discontinued on the 58th day, and the patient was fed orally on the postnatal 60th day. The patient was discharged on the postnatal 80th day in good general condition, with mobility in all four extremities, with normal neurological examination, on the condition

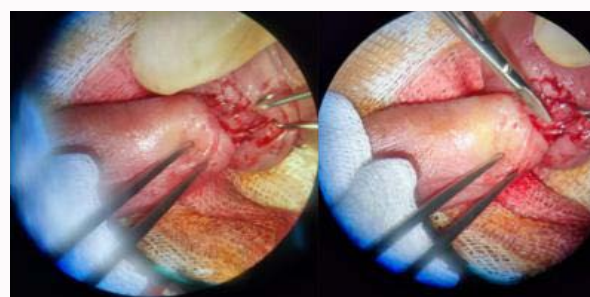


Figure 2: Release operation of fibrotic bands (postnatal 3rd day).



Figure 3: The appearance of postoperative extremities (postnatal 3rd day).

to continue his controls. The patient's parents were informed about the occurrence and course of the disease, and the increased risk of mortality and morbidity due to premature birth. Due to the mother's treatment for bipolar disorder, though it is known that it has no genetic transmission, in terms of the risk of recurrence due to drug use, it was recommended to take professional support considering the literature information; and to be closely monitored under detailed ultrasonography throughout pregnancy for the next pregnancy they would plan, taking the therapeutics used by the mother into consideration. The patient's parents were informed, and their written consent was obtained for the photo-sharing and publication of the case.

Discussion

In amniotic band syndrome, fetal malformations in the form of simple amputations, syndactyly, constriction rings surrounding the extremities without amputation, and reduction defects are common, especially in the extremities [7]. Although the detection of the amniotic band is not necessary for the final diagnosis, the detection of



Figure 4: The appearance of extremities before discharge.

the amniotic band alone is not sufficient for the diagnosis of amniotic band syndrome. There is no known clear etiology of amniotic band syndrome. In the etiology, pathologies such as infection, ischemia, blunt trauma especially in the placenta, interventional procedures such as amniocentesis, a history of antimetabolic drug use especially in the first 3 months of gestation, some connective tissue diseases such as Ehler Danlos, and uterine malformations are among the suspected reasons [8,9]. Furthermore, there are studies emphasizing that oligohydramnios, use of vasoconstrictive substances that decrease uteroplacental blood flow, prematurity and low birth weight for gestational age are among the risk factors [10]. Although there is no solid etiology, it has been reported in the literature that in cases where there is local deterioration in vascularity, for example, by increasing the contraction of the myometrium with the use of prostaglandin E, or by increasing the pressure on the uterus in large tumors located in the myometrium, it builds up the formation of a constrictive band in ischemia and venous congestion [11]. According to the UK National Teratology Information Service, lithium use increments all congenital malformations threefold and cardiac anomalies eightfold [12,13]. Although it is known that discontinuation of medication before or during gestation in half of the female bipolar patients leads to the relapse of the disorder, it is often recommended not to use lithium during gestation, especially in the first three months, and to discontinue it before gestation due to its teratogenic effect [14]. Although etiology is multifarious, in our case, the mother had not been under regular doctor follow-up during the antenatal period, however, lithium was administered in recurrent emergency service admissions due to bipolar attack and there had been an irregular intake of biperiden hydrochloride. She had a history of frequent urinary tract infections particularly as of the fourth month, premature rupture of membranes, and chorioamnionitis.

The most critical concern about ABS is the variety of findings [15]. ABS, which is observed at an equal rate in both genders, is considered to be sporadic, and although familial recurrences have not been reported, the risk of recurrence has been reported in some case reports, albeit low [16]. Mostly extremities are affected, and the face and, in some cases, various viscera may also be affected. The developmental disorders that occur can range from constriction rings, especially in the fingers, to limb amputations and craniofacial defects. Additionally, rare conditions such as cleft palate-lip, acrania, encephalocele, anophthalmia, and thoraco-gastroschisis may be among the detected findings [17]. Our case did not have any other anomaly except those in the extremities.

Prenatal diagnosis of amniotic band syndrome can be carried out by attentive ultrasonographic examinations. Today, intrauterine corrective and limb salvage surgical interventions are applied to cases with prenatal diagnosis [18]. In utero repair, besides its advantage

of providing scar-free tissue repair, decreases the development of secondary deformity by primary deformity repair, and it has advantages such as delivering a normal-appearing baby to the parents at birth [19]. Various hypotheses have been proposed to explain ABS. One of the hypotheses is Streeter's intrinsic theory, which also leads to the explanation of visceral malformations (it argues that defects in the germinal disc cause embryo developmental disorder). Torpin's extrinsic theory, on the other hand, argues that early amniotic rupture originates with mesodermal fibrous bands, and oligohydramnios plays a critical role in the development of constrictive bands [20-22]. According to Patterson, there are 4 types of congenital band constriction syndrome; Type-1: A simple constrictive band in the form of a groove on the skin, Type-2: Deformity independent of lymphedema in the distal constrictive band, Type-3: Fusion of the distal parts (Acro syndactyly) with the constrictive band, Type-4: Intrauterine amputation. In cases where in utero surgery is not performed, a surgery should be planned with a multidisciplinary approach as soon as possible following postnatal stabilization [23]. Surgically, there are approaches advocating a single-stage or multi-stage surgical interventions. While advocates of single-stage surgery emphasize that surgery-related complications are less common by reducing the patient's exposure to multiple anesthesia's, those who recommend multi-stage surgery argue that lymphedema and vascular complications are less common in the distal areas [24]. Aesthetic and functional results were generally found to be successful with the excision of the constriction bands and multiple Z-plasties [25,26]. In our case, Z-plasty was applied following one-stage surgery and band release, and a successful result was obtained in aesthetic and functional terms. There was no need for band reconstruction, repetitive surgery, and no complications occurred in the distal released band.

Conclusion

In conclusion, in utero surgery of selected cases with ABS, which can develop due to many reasons, or if this is not possible, surgery in the early postnatal period is critical to preserve limb functions. In case the mother has a history of using lithium or other suspected agents, close antenatal monitoring becomes crucial in cases where genetic changes in collagen structure occur.

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