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A neonate with Amniotic Band Syndrome and osteomyelitis

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Abstract

Amniotic band syndrome (ABS) comprises of a broad term for a group of congenital anomalies involving the limbs more often, but any other anatomic region of fetus could be involved. The prevalence of amniotic band has been reported from 0.19 to 8.1 per 10000 live births. Different theories exist for explaining the etiology of amniotic bands, but it has been considered to have a multifactorial etiology. Fetoscopic release of amniotic bands affecting one or more extremities can prevent limb loss or preserve limb function when distal fellow is still identifiable on preoperative ultrasound scans. Congenital anomalies of amniotic band syndrome will happen approximately in 77% of fetuses. There has been reported equal involvement between male and female. In general, we do not have osteomyelitis in neonates, but it would be connected to a long-term hospital admission and possible long-term complications if not diagnosed on time. The long bones are the most frequently affected sites. Here we are introducing a neonate with left upper extremity necrosis due to ABS which needed amputation and it was associated with severe chronic osteomyelitis.

Keywords: *Neonates, Amniotic band syndrome, Osteomyelitis*

Introduction

Amniotic band syndrome (ABS) is a collection of congenital anomalities. Amniotic Band Sequence, Streeter's dysplasia and ABS are the other names used for this condition. These congenital malformations occur due to fibrous amniotic bands [1]. ABS with multifactorial etiology is a congenital anomaly of the fetus, in which the fetal body parts have deformities due to amniotic band wrapped around them, resulting in fetal structural abnormalities and dysfunctions [1]. Usually, the diagnosis is made by deformities that are consistent with amniotic bands (such as limb deformities) in the absence of visible bands [2]. Prenatal diagnosis of ABS mainly depends on fetal ultrasound (US) scans [1]. But it is difficult to identify the amniotic bands with standard prenatal imaging, including ultrasound and MRI [2]. It has been reported to have the prevalence of 0.19 to 8.1 per 10000 live births [3]. There are many theories about the etiology of ABS but none of them support the others, so it might have a multifactorial etiology [3]. As the fetus grow up, amniotic bands wrap the extremities and can cause deformity or amputation of the affected limbs, whereas more complex bands involving the abdominal wall can lead to large abdominal wall defects mimicking gastroschisis or omphalocele [2]. Amniotic band syndrome has shown to be associated with hydranencephaly, porencephaly, craniofacial abnormalities and spinal dysraphism [4]. Most cases are sporadic and would not affect the sibling with the same congenital anomalies [5]. Karyotyping is important in counseling the parents about risk of recurrence [3]. Affected neonates need individualized multidisciplinary approach [3]. In fact, amniotic bands isolated to the extremities remain the primary indication for fetoscopic band release. If the bands are interrupting other anatomic regions, they have

usually made irreversible defects at the time of diagnosis, and currently there does not appear to be any indication for fetal intervention in those cases [2]. In some cases, post-natal surgical repair is suggested [3]. Osteomyelitis in neonates will happen due to the wide spread of bacterial infections or less common etiologies, like the direct entrance of bacterial infection due to trauma or surgery [6]. The prominent sign and symptoms of osteomyelitis include bone pain, redness, swelling and disability to move (pseudoparalysis). The causative pathogens are different in different countries but *Staphylococcus aureus* is the more common pathogen which will be found in 70% of positive cultured cases [6], *Streptococcus* mainly group B and Gram-negative enteric bacteria (*Escherichia coli and Klebsiella pneumonia* and *Pseudomonas aeruginosa*) are other causes of osteomyelitis [7, 8]. If signs and symptoms of osteomyelitis be present for less than 14 days, we will call the acute osteomyelitis, and subacute or chronic are the one with signs or symptoms for more than 14 days [9]. We will use different ways for the diagnosis of osteomyelitis, Imaging (computed tomography (CT) scan, radiography, bone scan, US and/or MRI) will be used for understanding fluid collection, site of infection and aspiration needing. We either will use different kind of imaging to differentiate all sites of infection which is important for the treatment and to investigate complications of the infection such as joint involvement and etc [6, 10].

Case presentation

A male infant who was delivered at 35th week and six days of pregnancy, by normal vaginal delivery with birth weight of 3250 grams and height of 52 cm. He had normal Apgar score. His mother had gestational diabetes and she was treated by insulin (GDM). He had passed meconium intrauterine but did not need any resuscitation at birth. At birth, his left upper extremity was necrotized probably with amniotic band from proximal humerus to the distal of fingers and it had many wounds on dorsal hand and lateral of forearm(picture1). His hand was painful, cold, pulseless and without any movement. Other examinations were normal. His mother`s ultrasonographic imaging in 26th week of pregnancy was reported normal. He had normal vital signs. He was admitted to neonatal ward and after performing a sepsis work-up, antibiotic therapy was started with meropenem and vancomycin. An umbilical catheter was inserted and he was transferred to another hospital for consulting about amputation surgery. After hematologist consultant, Heparin was started. His hand was amputated distal to shoulder at 8th day of life in Hazrat Rasool hospital. After surgery he was slightly tachypneic and he received oxygen with hood which was discontinued after two days. The entire lab tests were normal (table 1). At the 10th day of life (two days after surgery), he had weakness and poor feeding and a consultation with pediatric infectious specialist was requested. It was recommended to change the antibiotics to ciprofloxacin, linezolid and metronidazole and an MRI image of amputation site be performed and smear and culture of wound was requested.

The MRI result showed evidence of bone marrow edema in distal of humerus and soft tissue of amputation stump with cortical thickening in distal of humerus in favor of *chronic osteomyelitis*. Brain and abdominal ultra-sonographic scans were normal.

Vital signs and general condition of our infant improved after changing the antibiotic therapy, and he was treated for 21 days. The heparin therapy was withdrawn at 14th days after surgery. He was discharged at 24th day of life with good condition and without any antibiotic and normal lab tests (table2).

After two weeks the neonate was visited again, he was well and there was no problem with the amputation site and the infectious.

Discussion

Amniotic band syndrome (ABS) is not a prevalent disease and it is a rare anomaly which caused by amniotic sac that separate and entangle different part of the fetus body such as digits and limbs. This condition can cause different anomalies of the fetus depending on the tightness of the amniotic band and the location of the band. It can cause different kinds of congenital malformations such as deformity, disruption, and other malformations of organs [11]. It is so difficult to understand the etiology of amniotic band sequence but different theories wanted to conceptualize the etiologies. It seems that there are multiple factors involving in the etiology of amniotic band syndrome due to none of the theories support the other one individually [3]. Predisposing factors such as prematurity, low birth weight, maternal illnesses, maternal drug usage and other feto-maternal condition will affect this situation [12], although the risk factors are poorly known till now. The approach to ABS is affected by identifying the pre- and post-natal anomalies. The amniotic bands which are isolated are released by fetoscopy and if it is associated with other anomalies, performing a post-natal surgery repair is recommended. For genetic investigations it will be worthful too [3]. There

are four categories for the clinical presentations including: limb defect, spinal defects, craniofacial defects and constrictive rings. The distribution of amniotic bands is also different, for example extremity involvement can account for more than 70% of cases, but any other anatomic region of the fetus can be involved (umbilical cord, abdomen, head, chest, limb-body-wall-complex) [2]. The amniotic bands may only involve the soft tissues or other superficial tissues such as skin or may go deeper into the tissue. The amputated part of the fetus body will be reabsorbed in the amniotic fluid and would not be visible if it amputated in uterus.

For fetal amniotic band syndromes there are not any evidence of standard diagnosis or treatment guidelines. Due to the severity of the amniotic band syndrome ultrasonography and the interventions with the device will be needed [13]. There is one way to reduce the progression of amniotic band sequence and that is intra uterine interventions which causes the lysis of amniotic band syndrome for blood flow restoring. Plastic surgery may be needed after delivery, despite the fetoscopic intervention. However, the efficacy of this intervention is unknown [14]. Prognosis depends on the extension and severity of the defects. Interprofessional team will be present for the parents counseling and helping them for making decision about the surgery, nursing cares and etc.

Osteomyelitis (OM) is a severe neonatal infection which usually has a predisposing factor and necrosis of soft tissue and limb surgeries could be a trigger for late infections. The most significant pathogens are those involved in neonatal sepsis with *Staphylococcus aureus*, which is positive in about 70-90% of cultured positive cases. Long lasting sequels could be observed if it is not appropriately treated and even with best treatments, shortening of the affected limbs could be observed. Osteomyelitis will have about 6-50% permanent complications such as joint disabilities and deformity, change in bone growth, arthritis, fractures, prospective length of neonate, and rarely complete destruction of joints. MRI is the gold standard device for the diagnosis of musculoskeletal infections and the positive rate of MRI in detecting osteomyelitis was 100% [15]. MRI has the potential for assessing joints, articular, muscular tissues and does not have a destructive radiation for neonates [6]. Marrow intensity increasing are the most suggestive signs of osteomyelitis. Radiography is the first step for osteomyelitis diagnosis, despite it would be negative for any signs in the first two weeks of osteomyelitis [15].

There were several differential diagnoses in our case presentation such as congenital malformation, protein c and protein s deficiency, DIC due to sever septicemia, amniotic band syndrome, diabetes of mother. According to the normal range of coagulation proteins, the diagnosis of protein c and s deficiency was omitted. We ruled out the septicemia due to the neonate's sign and symptoms and the lab data. Based on the mother's history and the lab tests during pregnancy the diagnosis of GDM was well defined. On the other hand, we surely attributed the diagnosis to the amniotic band syndrome since the amniotic band was whirled toward the humerus meanwhile the delivery.

Conclusion

Amniotic band syndrome mostly involves the limbs. It is important to diagnose it as soon as possible, because with the interprofessional team work approach, an optimal patient result with fetoscopic or surgery procedures could be achieved. Even after surgery, some complications such as osteomyelitis could prolong the recovery process and it might need long term occupational and physiotherapic interventions and limb replacement treatments.

Table No 1: Lab tests

WBC:25200 (70%segment,25%lymph)	**Platelete:189000**	**BS:80 Bun: 15 mg/dl Cr: 1.3 mg/dl**
Hb:14.6 g/L	*CRP: 12 rising to 26*	*Serum Na: 140 mEq/LSerum K: 5.1 mEq/L*
Hct :45.2%	*PT:14.7 s PTT:37s INR:1.07 Clotting time: normal*	*Serum Ca:8.2 mg/dl Serum Mg: 2.1 mg/dlSerum CPK:1623 Serum LDH: 1435*
Blood culture: negative Wound Culture: negative	*AST:75 ALT:32*	*Protein S: normal Protein C: normal*

WBC: White blood cell; Hb: Hemoglobin; Hct: Hematocrit; CRP: C-reactive protein; PT: Prothrombin time; PTT: Partial thromboplastin time; INR: International normalized ratio; AST: Aspartate

aminotransferase; ALT: Alanine aminotransferase; BS: Blood sugar; BUN: Blood urea nitrogen; Cr: Creatinine; CPK: Creatine kinase; LDH: Lactate dehydrogenase.

Table No 2: Lab tests after surgery

WBC:12800(1% band,42% segment,31% lymph)	Plt:608000	Serum Na: 138 mEq/L
Hb:12.2 g/L	*CRP less than 6*	*Serum K:4.7 mEq/L*
Hct:35.4%	*Bun:9 mg/dl*	
	Cr: 0.4 mg/dl	

Fig 1: before amputation

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