Single stage surgical treatment of amniotic band syndrome – Case report

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SUMMARY

Introduction Amniotic band syndrome is a rare congenital disorder with clinical presentation of constricting bands in different parts of extremities or whole extremities. Conservative or surgical treatment is provided depending on the type and severity of the anomaly.

Case Outline The paper presents the case of a neonate patient with constriction bands localized on the left leg. During the second week of life, a surgery was indicated, and a single-stage multiple Z-plasty was performed to correct the anomalies on the left lower leg. Postoperative edema in the distal part of the lower leg was easily managed by incisions and drainage. Two months later, the correction of the stricture of the left thigh was managed using the same procedure. The postoperative course was uneventful and the outcome was satisfactory after a two-year follow-up.

Conclusion Evaluation of a patient with amniotic band syndrome, as well as diagnosis, monitoring, treatment and postoperative care, should always be multidisciplinary. A single-stage correction approach provided satisfactory both functional and aesthetic results. Given many morphological variations of the syndrome, a decision on the strategy of treatment should be made individually for each patient.

Keywords: amniotic band syndrome; congenital anomaly; pediatric surgery; treatment; outcome

INTRODUCTION

Amniotic band syndrome (ABS), also known as Streeter’s dysplasia, amniotic deformity, adhesion and mutilations, ADAM complex, represents a heterogeneous group of congenital anomalies with the incidence of 1:1,200 to 1:15,000 live births with equal frequency in both sexes [1, 2]. As many as 34 different names corresponding to this entity can be found in the literature [3].

The syndrome refers to a broad spectrum of anomalies including simple or multiple strictures, affecting only a portion or the whole circumference of limbs, oligodactyly, acrosyndactyly, pes equinovarus, cleft lip and palate, hemangioma, anencephaly or intrauterine fetal death due to umbilical cord strangulation [4]. Deformities may involve any portion of the fetus depending on the intrauterine localization. The extremities are involved most commonly, especially distal parts, while neck, chest and abdomen are rarely affected [5, 6].

Being formed inside the uterus, bands exert variable pressure on the tissue. As a result, changes that occur may vary from shallow wrinkles to complete amputation. In most cases, constriction bands extend to the first layer of fascia [6].

Peterson classifies clinical ABS into the following four types: type 1 – the presence of simple constriction ring; type 2 – the emergence of the ring combined with changes of distal parts, with or without lymphedema; type 3 – the presence of the strictures in combination with the fusion of the soft tissues in the distal parts; and type 4 – intrauterine amputation at any level of limb or finger. Deformities are usually caused by merger of soft tissue or lymphedema, but there are also angulations, flexion contractures, and stiff joints [7].

A routine prenatal ultrasound examination could reveal the presence of bands in utero, restricted fetal movements, oligohydramnios or characteristic deformities of the fetus. Besides ultrasound, in prenatal diagnosis, magnetic resonance imaging could give more detailed information [8]. Unfortunately, since prenatal diagnosis of ABS is not always possible, clinical examination after the birth is critical for diagnosis [9, 10].

Surgical treatment is not necessary in cases where vascularization is not compromised and there is no occurrence of lymphedema. However, it is often carried out for aesthetic reasons. If there are signs of lymphedema and functional disorder, urgent surgical treatment is necessary [1, 4].

CASE REPORT

A twin female newborn was sent from the regional hospital to our hospital after detecting anomalies corresponding to the ABS, during the first day of life. Pregnancy began as in vitro fertilization. It was uncomplicated and terminated with cesarean section at term (BM 3,045 g, 48 cm BL, AS 10/10).
A physical examination revealed the annular strictures in the left leg, and lymphedema with interruption of continuity of the skin, subcutaneous tissue and fascia in the both upper and lower leg (Figure 1). Ultrasound of head and abdomen, ECG and X-ray (Figure 2) were performed to detect if there were any other anomalies. Pediatrician and geneticist were consulted as well.

Since no other disorders were detected, an ultrasound and CT scan of lower extremity were conducted. Lymphedema of the entire circumference from the middle third of the left thigh to the lower leg and foot with hypoplasia of bone structures were observed, while muscular structures of the middle third of the lower leg and thigh were not well visualized. Deep veins were elastic, compressible, with satisfactory flow. They could be traced up to the second groove of edematous segment of the left leg. Numerous superficial venous blood vessels along with hyperechogenic muscles were present in edematous segment, while preserved architectonics of a muscle layer was visualized in the proximal non-edematous part of the leg.

The newborn was admitted to the intensive care unit where preoperative preparations were conducted. The surgical treatment of the anomalies was performed on the thirteenth day. The surgical procedure started with excision of the fibrous tissue and skin of the entire circumference around the stricture, followed with the reconstruction of the defect using multiple “Z” plasty, forming local triangular lobes. Multiple “Z” plasty was used instead of direct closure to prevent future contracture of the scar or functional impairment which can be expected during the growth due to uneven formation of “normal” scar. Postoperatively, an edema was observed distally from the incision line. Edema resolved after incision and drainage of the area, and on the fourth postoperative day, the child was transferred to the Neonatal surgical ward where compressive bandaging was conducted on daily basis.

Two months later, the correction of the constriction band in the left thigh was performed applying the same technique (Figures 3 and 4). Postoperatively, there was no formation of lymphedema, and the child was discharged.
The child was referred to physical therapy and had periodical check-ups during the recovery. Two years after the treatment, the child had no functional impairment, walking scheme was well preserved, and postoperative scars had satisfactory characteristics (Figures 5 and 6). The plan was to continue regular check-ups in order to monitor the possible changes during the growth and development of the child, as well as assessing the need for any corrective interventions.

DISCUSSION

The cause of the stricture development is not fully understood yet. There are two theories of the ABS onset described in the literature. In 1930, Streeter [11] claimed that before the formation of the embryo primary defect arises from the subcutaneous tissue in the early embryonic period (intrinsic theory). In 1965, Torpin [12], however, suggested that the formation of mesenchymal, amniotic bands is a consequence of premature rupture of membranes, oligohydramnios, protrusion of fetal parts into the chorionic cavity, and vascular compression of parts or entire limbs (extrinsic theory).

This theory could not explain the simultaneous occurrence of conditions such as anal atresia, polydactyly, cleft lip with or without cleft palate. A number of papers indicate that vascular compromise is obvious in the development of craniofacial and abdominal wall defects [13]. Some studies, which have proven the existence of blood vessels anomalies (bifurcation or trifurcation of artery, absence or segmental atresia of large blood vessels), compared to the contralateral unaffected limb, point out that the same pathogenic mechanism is also involved in developing ABS [14, 15].

The association of malformations of the limbs with malformations of other organs in ABS supports the “intrinsic” theory of vascular accident during the early stages of embryogenesis. Greater incidence of the syndrome in closest relatives supports the idea of the existence of predisposing genetic factors for ABS.

On the other hand, additional factors such as smoking, use of drugs, alcohol, and psychoactive substances, some diseases that affect the vascular system (diabetes), and iatrogenic lesions (injuries to the amniotic membrane during amniocentesis), seem to be equally significant, and all in favor of “extrinsic” theory [16].

Both theories give a good explanation for the onset of ABS, which is most likely caused by the combination of genetic and environmental factors.

Numerous cases of ABS are described in the literature. However, the same two cases of this syndrome have never been described. This emphasizes the importance of an individual approach to the treatment of each patient [16, 17].

Today, several approaches are used in correction of the strictures. The main aim of all surgical techniques is preservation of the function of the affected region, followed
by an improvement of the aesthetic appearance. The most commonly used techniques in solving the strictures are Z-plasties, W-plasties, Mutaf technique, the sine plasty, and direct closure of the defect after the excision of the band in one or two stages [4].

Stevensons, and many other authors indicate that the treatment in two acts reduces the likelihood of vascular complications in distal parts and lymphedema, as well as that closure of the defect without mobilization of subcutaneous adipose tissue could cause the formation of the scar tissue that will have a constrictive effect after surgery (the hourglass phenomenon) [1, 18–22]. In the literature, there are several papers that favor resolving congenital constrictive bands in the single stage, with very satisfactory results [23, 24].

In our case, the strictures were removed in the single-stage procedure, and reconstruction of the defect was performed using Z-plasty. As already mentioned, the goal of multiple Z-plasty was to prevent expected contracture of the scar or functional impairment. Although there was development of lymphedema after removal of the first stricture in the lower leg, after incision and bandaging, lymphedema withdrew on the fourth postoperative day. After eliminating other stricture in the thigh, there were no other complications in the further course of the treatment.

The operating method of removing an amniotic band in one stage have given quite satisfactory functional and aesthetic results. Also, by using the single-stage procedure, multiple surgeries and general anesthesia were avoided. Given the many morphological variations of the syndrome, a decision on the treatment strategy should be made specifically for each individual patient.

REFERENCES

1. Choulakian MY, Williams HB. Surgical correction of congenital constriction band syndrome in children: Replacing Z-plasty with direct closure. Can J Plast Surg. 2008; 16(4):221–3. [PMID: 19949501]
2. Shetty P, Menezes LT, Tauro LF, Diddigi KA. Amniotic Band Syndrome. Indian J Surg. 2013; 75(3):401–2. [DOI: 10.1007/s12262-012-0468-6] [PMID: 24426485]
3. Rayan GM. Amniotic constriction band. J Hand Surg (Am). 2002; 27:1110–1. [DOI: 10.1053/jhsu.2002.36517] [PMID: 12457366]
4. Nguyen Ngoc Hung. Congenital constriction ring in children: sine plasty combined with removal of fibrous groove and fasciectomy. J Child Orthop. 2012; 6(3):189–97. [DOI: 10.1007/s11832-012-0240-4] [PMID: 23814619]
5. Kiehn M, Leshem D, Zuker R. Constriction rings: the missing link. Eplasty. 2007; 8:4. [PMID: 18224248]
6. Ciloglu NS, Gumus N. A Rare Form of Congenital Amniotic Band Syndrome: Total Circular Abdominal Constriction Band. Arch Plast Surg. 2014; 41(3):290–1. [DOI: 10.5999/aps.2014.41.3.290] [PMID: 24883282]
7. Patterson T. Congenital ring constrictions. Br J Plast Surg. 1961; 14:1–31. [DOI: 10.1016/S0007-1226(61)80002-7] [PMID: 13733379]
8. Novakov-Mikić A, Koprivšek K, Lučić M, Stojić S. Prenatalna ultrazvučna i magnetnorezonantna dijagnostika tumora glave i vrata fetusa. Srp Arh Celok Lek. 2012; 140(1-2):97–9. [DOI: 10.1007/s11832-012-0240-4] [PMID: 23814619]
9. Matic A, Komazec J. Amniotic band syndrome. Acta Medica Medianae. 2009; 46(3):244–8.
10. Ruijwetpongstorn J, Tongsong T. Amniotic band syndrome following septostomy in management of twin – twin transfusion syndrome: a case report. Journal of Perinatology. 2008; 28:697–700. [DOI: 10.1016/j.jpeds.2007.09.005] [PMID: 18464662]
11. Streeter GL. Fetal deficiencies in fetal tissues and their relation to intrauterine amputations. Contrib Embryol Carnegie Inst. 1930; 22:1–44.
12. Torpin R. Anmiochorionic mesoblastic fibrous rings and amniotic bands: associated constricting fetal malformations or fetal death. Am J Obstet Gynecol. 1965; 91:65–73.
13. Hunter AGW, Carpenter BF. Implication of malformations not due to amniotic bands in the amniotic band sequence. Am J Med Genet. 1986; 24:691–700. [DOI: 10.1002/ajmg.1320240414]
14. Van Allen MI, Curry C, Gallager L. Limb body wall complex I, pathogenesis. Am J Med Genet. 1987; 28:529–48. [DOI: 10.1002/ajmg.1320280302]
15. Kuburović V, Vukomanović V, Košutić J, Rakić S, Gazikalović S. Adams-Oliverov sindrom – prikaz bolesnika. Srp Arh Celok Lek. 2011; 139(3-4):221–4. [DOI: 10.2298/SARH1104221K] [PMID: 21626769]
16. Guzmán-Santos IY, Domínguez-Moreno R, Muñoz-Delgado M, Martínez-Quintana JD. Amniotic band syndrome: report of a case. Ginecol Obstet Mex. 2013; 81(6):353–6. [PMID: 23837302]
17. Cignini P, Giorlandino C, Padula F, Dugo N, Cafà EV, Spata A. Epidemiology and risk factors of amniotic band syndrome, or ADAM sequence. J Prenat Med. 2012; 6(4):59–63. [PMID: 23272276]
18. Baker C, Rudolf AJ. Congenital ring constriction and intrauterine amputations. Am J Dis Child. 1971; 121:393–400. [DOI: 10.1001/archpedi.1971.02100160083006] [PMID: 4326366]
19. Greg A, Erol G. Congenital constriction band syndromes. J Pediatr Orthop. 1988; 8:461–6. [DOI: 10.1097/00124398-198807000-00016] [PMID: 2298/8/SARCH1202097N] [PMID: 22462336]
20. Anmolgrasundaram TK. Congenital anomalies. In: Kulkarni GS. Textbook of orthopaedics and trauma. Vol 4. Chennai: Jaypee Brothers Medical Publishers; 1999. p. 3439–947.
21. Herring IA, Tachdjian MO. Paediatric Orthopaedics. 3rd ed. Philadelphia: W. B Saunders; 2002. p. 461–6.
22. Upton J, Tan C. Correction of constriction rings. J Hand Surg. 1991; 16A:947–53. [DOI: 10.1016/0303-6303(91)01066-2] [PMID: 21626769]
23. Greene WB, Hill C. One stage release of congenital circumferential constriction bands. J Bone Joint Surg Am. 1993; 75:650–5. [PMID: 8501079]
24. Visuthikosol V, Hompuem T. Constriction band syndrome. Ann Plast Surg. 1988; 21:489–95. [DOI: 10.1097/00000637-198811000-00017] [PMID: 32329460]
Хируршко лечење синдрома амнионских брида у једном акту – приказ болесника

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КРАТАК САДРЖАЈ
Увод Синдром амнионских брида (ABS) ретка је конгенитална аномалија коју карактерише појава стриктура најчешће на деловима или целим екстремитетима. У зависности од степена захађањености и типа аномалије лечење је конзервативно или оперативно.

Приказ болесника У раду је приказан случај рочног новорођенчета са ABS локализованим на левој нози. У другој недељи живота се оперативно коригује аномалија на левој потколеници. Постоперативно долази до појаве едема у дисталном делу оперисане потколенице, што се лако решава инцизијом и дренажом. Два месеца касније се оперативно коригује стриктура леве натколенице у једном акту, методом мултипле „Z” пластике. Постоперативни ток протиче уредно, стање на контролном прегледу након две године је задовољавајуће.

Закључак Приступ пацијентима са овим синдромом требало би да буде мултидисциплинар, посебно у погледу дијагностике, праћења и лечења. Оперативни метод уклањања брида у једном акту је дао сасвим задовољавајући функционални и естетски резултат. С обзиром на бројне морфолошке варијације синдрома, одлука о плану лечења треба да буде донета индивидуално за сваког пацијента.

Кључне речи: синдром амнионских брида; конгениталне аномалије; дечја хирургија; лечење; исход

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