Case report

Access this article online
Website: www.cjisa.org

Anaesthetic management of amniotic band syndrome: A case report

Address for correspondence:
Dr. Shobha. V
Assistant Professor,
Department of Anaesthesiology
and Critical Care,
Himalayan Institute of
Medical Sciences,Dehradun,
Uttarakhand, India
E-mail: drshobha.v.v@gmail.com

Shobha V[1], Veena Asthana[2], Amit Kumar Chauhan[3]

[1]Assistant Professor, Department of Anaesthesiology and Critical Care, Himalayan Institute of Medical Sciences, Dehradun, Uttarakhand, India

[2]Professor, Department of Anaesthesiology and Critical Care, Himalayan Institute of Medical Sciences, Dehradun, Uttarakhand, India

[3]Senior Resident, Department of Anaesthesiology and Critical Care, Himalayan Institute of Medical Sciences, Dehradun, Uttarakhand, India

ABSTRACT

Amniotic Band Syndrome (ABS) is a complex congenital condition caused by strands of amniotic sac associated with a broad spectrum of anomalies. It is also known as amniotic band sequence, Amniotic Deformity, Adhesions, Mutilations (ADAM) complex, congenital constriction rings, constriction band syndrome, limb body wall complex, Streeter anomaly, Streeter dysplasia, Annular grooves, Congenital amputation, Pseudoainhulm[1]. The abnormalities vary from isolated to multiple bizarre disfiguring complications involving craniofacial region, limbs and visceral organs. General anaesthesia is the preferred choice for surgical correction of anomalies.

Key words: Amniotic band syndrome, High arched palate, Syndactyly

INTRODUCTION

Amniotic Band Syndrome is a rare congenital deformity with a broad spectrum of structural abnormalities ranging from minor constricting rings and lymphedema of digits to major craniofacial and limb body wall defects, *thoraco*-abdominoschisisthat have been attributed to amniotic bands that entangle and amputate fetal parts. The estimated prevalence is 1 in 1200 to 15,000 live births[2–4], 178 in 10,000 miscarriages[5] affecting both sexes equally. There is no family predisposition or risk of recurrence. There is a significant predilection for upper extremities particularly distal limbs. We report this case with the aim of highlighting its existence in this region and of academic interest.

CASE REPORT

A 2 months old male baby weighing 4 kgs was scheduled for elective surgery for release of constriction ring affecting limbs with Z-plasty under general anaesthesia. He was a full term baby delivered vaginally in hospital at 38 weeks of gestation with no maternal complications. Mother admits no ultrasound scan was performed during the antenatal period. Clinical examination revealed a constriction rings 2 cms above right ankle and 2 cms proximal to left wrist in total circumference associated with syndactyly and high arched palate. Figure 1-3.

Systemic examination was within normal limits. Routine laboratory investigations: Complete hemogram, electrolytes and urine analysis were normal. Chest x-ray (PA view), ECG, Ultrasound abdomen and pelvis and 2D echocardiography were normal. X-ray of limbs showed soft tissue constriction without the involvement of the bones. Colour Doppler study of the limbs revealed normal blood flow in all the vessels and at constriction site.

Airway assessment revealed adequate mouth opening with the high arched palate. The baby was nil by mouth for 4 hrs. Insideoperation theatre, standard monitors like 5 lead ECG, NIBP cuff, pulse oximetry were attached, and vital parameters were recorded which were as follows: pulse rate -148/min, NIBP-92/62mmhg, SpO_2 -100% on room air. An intravenous access was made with the 24G cannula and Isolyte-P started using paediatric burette. Preoxygenation was done with 100% oxygen for 3 mins. Premedicated with Inj. Glycopyrrolate 0.01mg/kg, Inj. Midazolam 0.02mg/kg, Inj. Fentanyl

How to cite this article: Shobha V, Asthana V, Chauhan AK. Anaesthetic management of amniotic band syndrome: A case report Central Journal of ISA 2018;2(1):25-28.

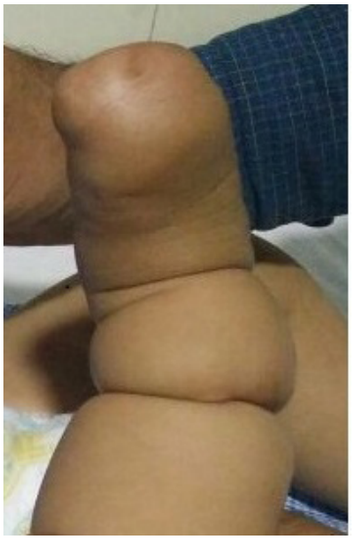

Figure 1. A circumferential band above right ankle.

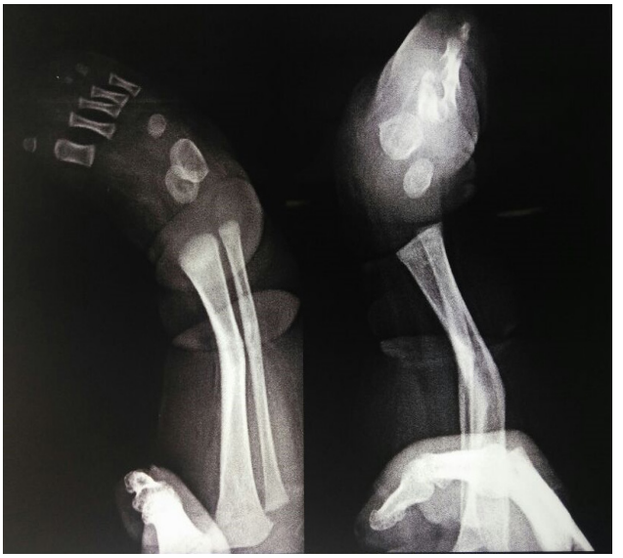

Figure 2. X-ray lateral and AP view of right leg with foot.

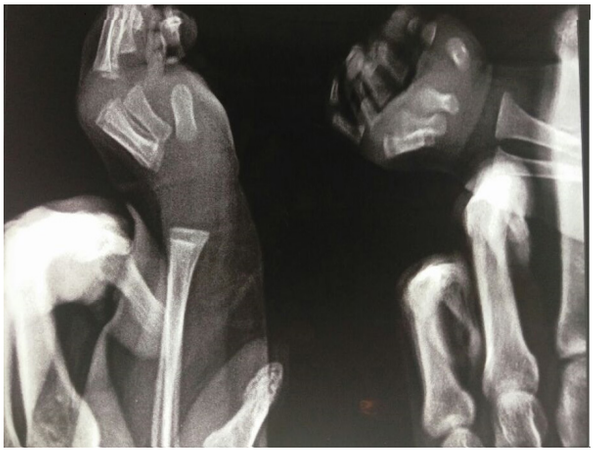

Figure 3. X-ray lateral and AP view of left upper limb with constriction band above wrist.

2mcg/kg followed by induction with Inj. Propofol 2mg/kg and Inj. Atracurium 0.5mg/kg after check ventilation. Direct laryngoscopy was done and airway secured with 3.0 mm ID uncuffed endotracheal tube (Cormack Lehane Grade 2,type b) and was confirmed by bilateral air entry and $EtCO_2$, the tubewas fixed at 9 cms. Anaesthesia maintained with 50% oxygen + 50% nitrous oxide, Sevoflurane, Inj. Atracurium 0.1mg/kg and positive pressure ventilation was done. Heat conservation was made with warming blanket, warm IV fluids and maintaining ambient room temperature. Antibiotic was given prior to skin incision. Surgery continued for one and half hour. Inj. Paracetamol 10mg/kg was given slow ivi. The patient was reversed with Inj. Neostigmine 0.05mg/kg and Inj. Glycopyrrolate 0.01mg/kg. Extubation was done after clearing oral secretions. Intraoperative and post-operative periods were uneventful. The baby was discharged after 3 days.

DISCUSSION

Amniotic constriction bands are fibrous strands of the amniotic sac that entangle fetal parts and disfigure arms, legs, fingers, or toes. Though the etiopathogenesis is unknown, two theories have been proposed "extrinsic model" and the "intrinsic model". In 1930 the intrinsic model was proposed by Streeter[6] and suggests that the fibrous bands and the anomalies have a common origin, caused by a perturbation of developing germinal disc of the early embryo. In 1965, the Torpin's model of "extrinsic theory"[7], suggested that the birth defects are caused by the action of the fibrous amniotic bands with the sequence rupture of the amnion, followed by loss of amniotic fluid and extrusion of all or parts of the fetus into the chorionic cavitywhich comes in contact with 'sticky' mesoderm on the chorionic surface of the amnion leading to entanglement of the fetal parts and skin abrasions. Entanglement of the fetal parts causes constriction rings and amputations, whereas skin abrasions can lead to disruption defects, such as cephaloceles. The fetus limbs, while trapped there are subjected to vascular compression and then necrosis[8–11].

Beside ABS etiopathogenesis, risk factors are poorly known. However, there are some reports of amniotic band syndrome associated with Ehler-Danlossyndrome[12], osteogenesisimperfecta[13], and congenital epidermolysis bullosa[14,15]. Few studies showed association between ABS and mother's age (especially primiparas of age less than 25 years)[10,15], prematurity[16], abdominal trauma[15,18], abortion[17], intrauterine contraception[18], amniocentesis[17-21], malformations of the uterus[15], some drugs like ergotamine[22], acetaminophen[10] with no firm evidence.

Amniotic band syndrome has very complex clinical findings. Early amniotic rupture, during the first 45 days, leads to the

most severe craniofacial, limb body wall defect and visceral malformations[4]. Every part of the fetal body may be damaged, but most often upper extremities are affected where minor defects such as constriction rings or digit amputations[23] are seen. Other abnormalities like constriction rings of the soft tissue accompanied by distal edema, shortening of the limb or intrauterine limb amputation, amputation of the digits (most often II, III and IV finger) and toes, syndactyly, polydactyly, hypoplasia of the digits, clubfoot, pseudoarthrosis, peripheral nerve palsy[2,22] and oligohydramnios causes deformities such as metatarsovarus, scoliosis[4] or hip dislocation[2]. Iffibrous strands compress the fetal head or face, different cranio-facial disturbances can appear- asymmetric face clefts, orbital defects, corneal abnormalities, central nervous system malformations, calvaria defect. Other anomalies like chest wall defect with heart extrophy[24], gastroschisis, small intestinal atresia, terminal transverse limb reduction defects, microtia, clubfoot[25] and Septo-optic dysplasia[26].

Another important association with ABS is the presence of cleft lip with or without palate (CLP) in a few patients. Mutations of genes involved in cleft lip and cleft palate are known to be directly related to genes involved in the amniotic band syndrome. It is important to note that recently identified cleft lip and palate genes have oral or facial fibrous bands as single component finding which includes Van der Woude and popliteal pterygia syndrome, caused by mutations in IRF6[27,28] and Hay-Wells Syndrome, caused by mutations in p63[29] which is associated with limb anomalies[30]. There are reports of patients with Amniotic band syndrome and Cleft lip and palate, who have additional anomalies, such us supernumerary left nipple, polydactyly, vertebral segmentation defects, imperforate anus, renal agenesis, and a skin papilla. Study on mice by Donnai and Winter (1989) concluded that these cases represented the human homolog of a gene whose mutation is due to similar disorders (Ds). It is a gain-of-function mutation explained through a two-hitmechanism[31]. In our case, we encountered high arched palate in association with ABS and syndactyly.

Amniotic band syndrome is typically diagnosed at or soon after birth based upon characteristic physical findings. In some cases, the condition may be suspected prenatally, Latest ultrasound techniques- three dimensional and four-dimensional ultrasound contribute to more sensitive prenatal diagnostics of ABS, and in complicated cases, fetal MRI can be helpful[3].

Management includes surgical correction either one-stage or two-stage procedure depending upon the extent of the associated anomalies. In a study conducted by Patterson[32] where 52 patients of congenital constriction rings had reported only 2 below knee amputations in addition to other musculoskeletal defects. In a study conducted by Greene[33] one-stage release for circumferential congenital constriction bands was performed in four extremities. In 1983 Zych[34] et al., reported a case of ABS with pseudarthrosis and impending gangrene of leg, which was salvaged with multiple Z-plasty. Mistry[35] et al., in 2015 reported a case of congenital bands in upper and lower limbs associated with cleft lip and palate which was salvaged with multiple Z-plasty and staged cleft lip and palate repair. In our case, the baby presented to us at the age of 2 months with the high arched palate and there were no threatened complications like gangrenous changes, bony deformity or loss of motor or sensory function and it was treated in a single stage by multiple Z-plasty. The high arched palate did not pose any airway risk to the patient during the perioperative or postoperative period.

It is wise to consider ABS patient as difficult airway due to the abnormalities in addition to normal paediatric airway anatomy and physiology. In our case, it was a difficult intubation as it required external manipulation.

CONCLUSION

Every case of ABS is unique with a constellation of anomalies and needs to be assessed individually. No two cases have an identical presentation. Because of its complexity, the treatment and follow-up of these neonates requires a team of specialists depending upon the special needs of each patient. A well planned surgical treatment can produce excellent results for complete restoration of the form and function of the patient.

REFERENCES

1. Gonçalves LF, Jeanty P. Amniotic band syndrome; 1999-09-26-18
2. Poeuf B, Samson P, Magalon G. Amniotic band syndrome. Chir Main. 2008 Dec; 27(Suppl 1):S136–47. https://doi.org/10.1016/j.main.2008.07.016 PMid:18948051
3. Merrimen JL, McNeely PD, Bendor-Samuel RL, Schmidt MH, Fraser RB. Congenital placental–cerebral adhesion: an unusual case of amniotic band sequence. J Neurosurg. 2006;104(5 Suppl):352–5. PMid:16848094
4. Jabor MA, Cronin ED. Bilateral cleft lip and palate and limb deformities: a presentation of amniotic band sequence? J Craniofac Surg. 2000; 11(4):388–93. https://doi.org/10.1097/00001665-200011040-00021
5. Pretorius DH, Mahony BS. The role of obstetrical ultrasound. Nyberg DA, Mahony BS, Pretorius DH, editors. Diagnostic ultrasound of fetalanomalies. Text and Atlas, Mosby, St. Louis; 1990. p. 1–20.
6. Streeter GL. Focal deficiencies in fetal tissues and their relation to intrauterine amputations. Contrib Embryol Carnegie Inst. 1930; 22:1–44.
7. Torpin R. Amniochorionicmesoblastic fibrous rings and amniotic bands: Associated constricting fetal malformations or fetal death. Am J Obstet Gynecol. 1965; 91:65–75. https://doi.org/10.1016/0002-9378(65)90588-0
8. Robin NH, Franklin J, Prucka S, Ryan AB, Grant JH. Clefting, amniotic bands and polydactyly: A distinct phenotype that supports an intrinsic mechanism for amniotic band sequence. Am J Med Genet A. 2005; 137A(3):298–301. https://doi.org/10.1002/ajmg.a.30885 PMid:16088913
9. Lateo SA, Taylor AE, Meggitt SJ. Raised limb bands developing in infancy. Br J Dermatol. 2006; 154(4):791–2. https://doi.org/10.1111/j.1365-2133.2006.07160.x
10. Werler MM, Louik C, Mitchell AA. Epidemiologic analysis of maternal factors and amniotic band defects. Birth Defects Res A Clin Mol Teratol. 2003; 67(1):68–72. https://doi.org/10.1002/bdra.10001 PMid:12749386

11. Fathallah ZF. Unusual Presentation of Amniotic Band Syndrome. Bas J Surg. 2007; 11:77–9
12. Young ID, Lindenbaum RH, Thompson EM, Pembrey ME. Amniotic band syndrome in connective tissue disorders. Arch Dis Child. 1985; 60:1061–3. https://doi.org/10.1136/adc.60.11.1061 PMid:3840971 PMCid:PMC1777608
13. Barsh GS, Byers PH. Reduced secretion of structurally abnormal type I procollagen in a form of osteogenesis imperfecta. Proc Natl Acad Sci USA. 1981; 78:5142–6. https://doi.org/10.1073/pnas.78.8.5142
14. Kroes HY, Pals G, van Essen AJ. Ehlers–Danlos syndrome type IV: unusual congenital anomalies in a mother and son with a COL3A1 mutation and a normal collagen III protein profile. Clin Genet. 2003; 63(3):224–7. https://doi.org/10.1034/j.1399-0004.2003.00047.x PMid:12694234
15. Dyer JA, Chamlin S. Acquired Raised Bands of Infancy: Association with Amniotic Bands. Pediatric Dermatology. 2005; 22(4):346–9. https://doi.org/10.1111/j.1525-1470.2005.22414.x PMid:16060874
16. Clarke JC, Becker EJ, Salonen DC. Amniotic band syndrome. Can Assoc Radiol J. 2000; 51(2):134–6. PMid:10786923
17. Ferns S, Sharma R, Narayanan P, Bhat BV. Limb amputation defects in Utero-A case report. J Anat Soc India. 2001; 50(2):156–8.
18. Burk CJ, Aber C, Connelly EA. Ehlers-Danlos syndrome type IV: Keloidal plaques of the lower extremities, amniotic band limb deformity, and a new mutation. J Am Acad Dermatol. 2007; 56(2 Suppl):S53–4. https://doi.org/10.1016/j.jaad.2006.11.008 PMid:17224388
19. Kanit H, Aslan D, Dicle N, Ugurel G. Amniotic band syndrome after cervical cerclage. Perinatoloji Dergisi. 2001; 9:1–3.
20. Bower C, Norwood F, Knowles S. Amniotic band syndrome: A population-based study in two Australian states. Paediatric and Perinatal Epidemiology. 1993; 7(4):395–403. https://doi.org/10.1111/j.1365-3016.1993.tb00421.x PMid:8290379
21. Baltaci V, Akyol D, Gul D, Sayli BS. Digital amputations caused by amniotic bands – A case report. Tr J of Medical Scienses. 1998; 28:701–3.
22. McGuirk CK, Westgate MN, Holmes LB. Limb deficiencies in newborn infants. Pediatrics. 2001; 108(4):e64–e70. https://doi.org/10.1542/peds.108.4.e64 PMid:11581472
23. Evans C, Marton T, Rutter S, Anumba DO, Whitby EH, Cohen MC. Cranial vault defects: The description of three cases that illustrate a spectrum of anomalies. Pediatr Dev Pathol. 2009; 12(2):96–102. https://doi.org/10.2350/08-02-0415.1 PMid:18442303
24. KutluDilek TU, Yazici G, Gulhan S, Polat A, Dilek B, Dilek S. Amniotic band syndrome associated with cranial defects and Ectopia cordis: A report of two cases. J Turkish German Gynecol Assoc. 2005; 6(4):308–10.
25. Van Allen MI, Curry C, Walden CE, Gallager L, Patten RM. Limb body wall complex: II, limb and spine defects. Am J Med Genet. 1987b; 28:549–65. https://doi.org/10.1002/ajmg.1320280303 PMid:2962494
26. Stevens CA, Dobyns WB. Septo-optic dysplasia and amniotic bands: Further evidence for a vascular pathogenesis. American Journal of Medical Genetics. 2004; 125A:12–6. https://doi.org/10.1002/ajmg.a.20417 PMid:14755460
27. Leck GD, Aird JC. An incomplete form of the popliteal pterygium syndrome? Br Dent J. 1984; 157:318–9. https://doi.org/10.1038/sj.bdj.4805480 PMid:6595011
28. Kondo S, Schutte BC, Richardson RJ, Bjork BC, Knight AS, Watanabe Y, Howard E, de Lima RL, Daack-Hirsch S, Sander A, McDonald-McGinn DM, Zackai EH, Lammer EJ, Aylsworth AS, Ardinger HH, Lidral AC, Pober BR, Moreno L, Arcos-Burgos M, Valencia C, Houdayer C, Bahuau M, Moretti-Ferreira D, Richieri-Costa A, Dixon MJ, Murray JC. Mutations in IRF6 cause Van der Woude and popliteal pterygium syndromes. Nat Genet. 2002; 32:285–9. https://doi.org/10.1038/ng985 PMid:12219090 PMCid:PMC3169431
29. McGrath JA, Duijf PH, Doetsch V, Irvine AD, de Waal R, Vanmolkot KR, Wessagowit V, Kelly A, Atherton DJ, Griffiths WA, Orlow SJ, van Haeringen A, Ausems MG, Yang A, McKeon F, Bamshad MA, Brunner HG, Hamel BC, van Bokhoven H. Hay-Wells syndrome is caused by heterozygous missense mutations in the SAM domain of p63. Hum Mol Genet. 2001; 10:221–9. https://doi.org/10.1093/hmg/10.3.221 PMid:11159940
30. Barrow LL, van Bokhoven H, Daack-Hirsch S, Andersen T, van Beersum SE, Gorlin R, Murray JC. Analysis of the p63 gene in classical EEC syndrome, related syndromes, and non-syndromic orofacial clefts. J Med Genet. 2002; 39:559–66. https://doi.org/10.1136/jmg.39.8.559 PMid:12161593 PMCid:PMC1735218
31. Crosby JL, Varnum DS, Nadeau JH. Two-hit model for sporadic congenital anomalies in mice with the disorganization mutation. Am J Hum Genet. 1993; 52:866–74. PMid:8488837 PMCid:PMC1682043
32. Patterson TJ. Congenital ring-constrictions. Br J Plast Surg. 1961; 14:1–31. https://doi.org/10.1016/S0007-1226(61)80002-7
33. Greene WB. One stage release of congenital constriction bands. J Bone Joint Surg Am. 1993; 75:650–65. https://doi.org/10.2106/00004623-199305000-00003 PMid:8501079
34. Zych GA, Ballard A. Constriction band causing pseudarthrosis and impending gangrene of the leg. A case report with successful treatment. J Bone Joint Surg. 1983; 65A:410–2. https://doi.org/10.2106/00004623-198365030-00024
35. Mistry T, Mathur R, Saini N, Rathore P. Perioperative management of amniotic band syndrome: a case report and literature review. Anaesth Pain and Intensive Care. 2015; 19(4):505–9.