

Amniotic band syndrome with tethering of the spinal cord: a case-based update

Benedetta Pettorini · Naem Abbas ·
Shailendra Magdum

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Abstract

Background Amniotic band syndrome consists of a wide spectrum of clinical manifestations attributed to entanglement and disruption of different developing parts of the embryo. Multiple asymmetric encephalocele and anencephaly have previously been reported with amniotic band syndrome. Tethering of the spinal cord secondary to amniotic band constriction is exceedingly rare, and this is the second reported case in the literature.

Case report We present a case of amniotic band resulting in tethering of spinal cord. It is a rare entity, and it is the second reported case of amniotic band causing tethering of the spinal cord. Standard operative approach was used to untether the cord. The child made good post-op recovery without any neurological deterioration. A review of the literature and causative theories is discussed.

Conclusions Neural tube defects involving head and spine are thought to result from adhesions between craniofacial structures and chorionic wall or compression forces by amniotic bands. Tethering of the thoracic spinal cord with amniotic band is an exceedingly rare occurrence. It is a rare entity, but it can be treated with a conventional approach with a favourable outcome.

Keywords Amniotic band syndrome · Neural tube defect · Tethered cord

Background

Introduction

Amniotic band syndrome (ABS) consists of a wide spectrum of clinical manifestations attributed to entanglement and disruption of different developing parts of the embryo [1, 9]. These congenital malformations range from minor constriction rings and lymphoedema of the digits to complex craniofacial and central nervous system abnormalities [1]. It can result in cutaneous scars, erosions and ulcerations, digital constricting bands, amputations, club feet, cleft lip and craniofacial and visceral anomalies. The prevalence for live births is 1:875 to 1:15,000; for spontaneous abortions, it can be as high as 7–14% [1, 9]. Males and females are equally affected. Central nervous system defects include encephalocele, anencephaly and scoliosis. Multiple asymmetric encephalocele and anencephaly have previously been reported with ABS [2]. Tethering of the spinal cord secondary to amniotic band constriction is exceedingly rare, and this is the second reported case in the literature.

Pathogenesis and epidemiology

A process of cavitation begins between inner cell mass and cytotrophoblasts, which gradually enlarges; this results in the formation of amniotic cavity by the eighth day of gestation, and it is lined internally by ectodermal cells and externally by extraembryonic somatic mesoderm [9]. The developing embryo sits within the amniotic cavity that is

B. Pettorini (✉) · N. Abbas
Department of Paediatric Neurosurgery,
Birmingham Children Hospital,
Steelhouse Lane,
Birmingham B4 6NH, UK
e-mail: benedetta.pettorini@bch.nhs.uk

B. Pettorini
e-mail: benedettaludovica@virgilio.it

S. Magdum
Department of Neurosurgery, Oxford Children's Hospital, John
Radcliffe Hospital,
Oxford, UK

contained within the chorionic cavity; these two membranes are separated by extraembryonic coelom [9]. Gradual expansion of the amniotic cavity entails the almost complete fusion of the amniotic and chorionic membrane. This leads to complete obliteration of extraembryonic coelom by the 12th week of gestation.

Pathogenesis of this intriguing malformation of the foetus has been studied since the time of Hippocrates and Aristoteles. Many speculations were presumed to be the cause of this anomaly, but the exogenous theory by Torpin gained much popularity. This theory was first proposed by Montgomery in 1832 [1] and strengthened by the extensive study by Torpin in 1968 [11]. Torpin examined 10,000 placentas and 400 cases of amniogenic foetal anomalies. Poor fusion of amniotic and chorionic membranes may result in insufficient support for the cavity, making it vulnerable to rupture easily when subjected to mechanical event, vascular compromise or infectious processes [11]. This theory suggests that rupture of the amnion in early pregnancy leads to entrapment of foetal structures by “sticky” mesodermic bands that emanate from the chorionic side of the amnion, followed by disruption [11]. Fibrous strand encircle the extremities and cause asymmetric constriction of developing parts. Compression deformities such as club feet, superficial abrasion, equinovarus or skull deformity can be explained by oligohydramnios resulting from leakage of amniotic fluid through permeable chorionic membrane [11]. Swallowing of loose mesodermal tissue by the foetus is believed to contribute in the formation of cleft palate and cleft lip. This theory has been recently contested based on clinical and experimental data [11].

For many years, the defects associated with amniotic band syndrome were thought to result from focal developmental errors in the formation of limb connective tissue. Lockwood contests the exogenous theory based on clinical and experimental evidence [5]. Exogenous theory cannot explain the high prevalence of internal visceral anomalies and asymmetric locations not predicted by constriction effects of fibrous bands, normal histology and intact amnion in certain cases of amniotic band syndrome and lack of consistent association with maternal trauma or toxins [5]. The pathogenesis would involve vascular compromise, leading to damage of the mesenchymal and endothelial cells of the superficial vessels of the embryo and the amnion, with disruption of epiblastic cells and secondary limb amputation, constriction bands, cephaloceles, syndactyly, club foot and clubbed hands [5]. Amniotic band formation would be a late and secondary event, analogous to peritoneal adhesion formation following an inflammatory or surgical trauma insult.

Moreman introduced the idea that both theories complement each other in explaining the varied clinical picture of ABS [7].

Clinical presentation and diagnosis

Tethering of the spinal cord represents a condition in which the cord is adherent to an immovable structure such as skin, bone, dura or fat. Stretching of the cord happens when the cord cannot accommodate the growth of vertebral column. This will result in vascular, neuronal and axonal derangements manifested as pain, motor, sensory or sphincter disturbances.

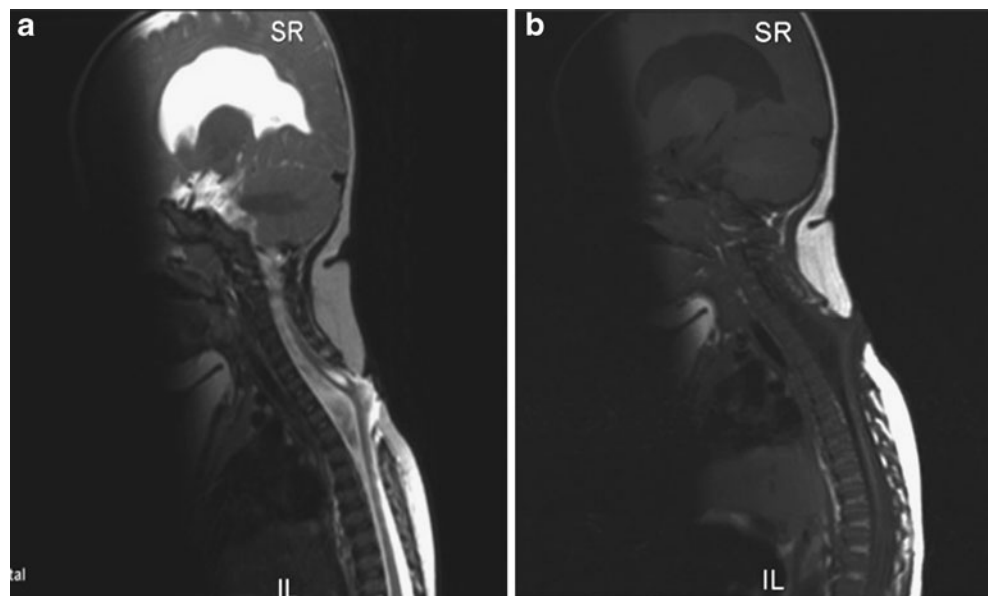
The theory behind the tethering in cases of ABS was elegantly postulated by Prabhu and Tomita in 1999 [9]. Compression from the encircling band prevented normal mesodermal migration between the ectoderms, leading to dysraphic lesion and attending tethering. This would have happened after the primary neurulation after days 23–24. On reviewing the literature on ABS and involvement of the spinal cord, there have been three case reports, with one tethering in an infant who was operated at 4 months of age [9]. The second report is of open thoracic meningocele with ABS who was treated at 7 days of age [3], and the third one is an adult diagnosed at 27 years of age and treated conservatively [6]. Interestingly, all cases with a spinal cord involvement presented with a thoracic spinal cord lesion at the level T1 to T5 associated with prominent central canal or syringomyelia just above the lesion. This is the second case report of spinal dysraphism with dorsal spinal cord tethering associated with ABS in children who had similar anatomic features.

Our patient demonstrated early signs of motor weakness that gradually improved over the next few months.

Management and prognosis

The ABS refers to a heterogeneous group of congenital anomalies characterised by amputation, constriction bands and different multiple craniofacial, visceral and body wall defects. Some of these cases are not compatible with life [2]. A wide spectrum of clinical deformities has been reported so far, ranging from a simple ring constriction band to major visceral defects [4, 12]. Management of amniotic band is usually tailored to the individual case and is focused on improving function and development. Lower limb malformations are the most commonly related and associated to better prognosis [12]. Extremity amniotic band is now commonly released fetoscopically [10]. Surgical excision of the deformity and the band can be performed within the first week of life or according to the general conditions [8]. Considering the two reported cases on ABS and involvement of the spinal cord, one tethering in an infant was performed at 4 months of age with a favourable outcome [9]; the child with an open thoracic meningocele with ABS and tethered cord was treated at 7 days of age, remaining neurologically intact after the repair and untethering [3].

Fig. 1 T2-weighted (a) and T1-weighted (b) sagittal images demonstrating the tethered cord with syrinx above the tethering



Illustrative case

This 16-month-old baby girl was born at 26 weeks of gestation. She was the first child of an Asian female by normal delivery. The antenatal course was uneventful, with a normal antenatal ultrasound and with no history of maternal trauma or infection. The weight at birth was 864 g, and ventilator support was required for 11 days because of surfactant-deficient lung disease. The examination revealed a flat facial profile and abnormal sternum and nipples. A circumferential scar around the mid-thoracic region, which widened in the midline, was noticed; the skin

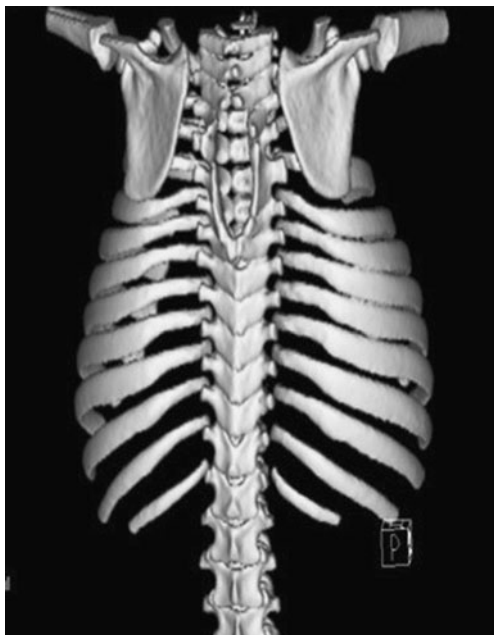


Fig. 2 3D reformatted CT showing the bony defect with neural arch defect

over the scar looked normal. The karyotype study revealed a normal 46 XX genotype. She did not have any neurological deficit at birth. On day 22, the patient developed a necrotising enterocolitis managed conservatively. The child needed intensive care management for about 3 months. The patient was referred to the neurosurgical team at 15 months of age. The neurological assessment at that time revealed a normal head circumference without any signs of raised intracranial pressure. She was found to have a short, thick neck and a palpable bony defect in the spine from T1 to T5 with atretic skin with no hairy anomalies but a band of scar tissue running circumferentially. Symmetrical and good power with normal sensation was demonstrated in upper extremities. She was able to stand with support but had a tendency to fall, suggestive of early motor or posterior column involvement. Her preferred position was lying flat rather than sitting. Ear, nose, throat and ophthalmology assessment was normal. No other features of amniotic band syndrome was noted on physical examination. There was no developmental delay. A magnetic resonance imaging of the craniospinal axis revealed

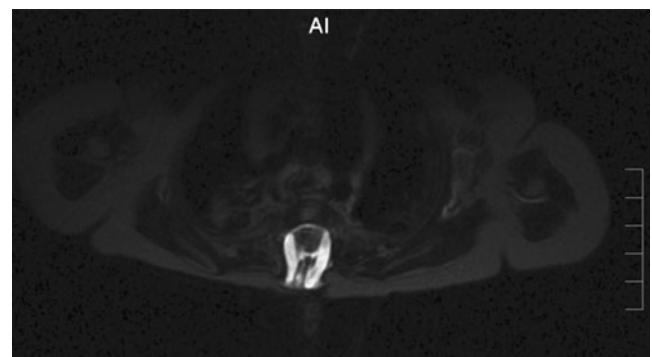


Fig. 3 T2 axial at the level of tethering showing fibrous bands tethered to skin

dilated third and lateral ventricles, absent septum pellucidum and delayed myelination (Fig. 1a, b). There was a posterior fusion defect from T1 to T5 (Fig. 2) with dorsal cord tethering with associated syringomyelia from C5 to T1 and extension of the meninges and cerebrospinal fluid through the defect to the subcutaneous tissue (Fig. 3). There was no radiological evidence of tonsillar descent. At 16 months of age, she underwent untethering of thoracic spinal cord through a standard de-tethering procedure. Intraoperatively, there was an abnormal, mostly fibrous tissue merging imperceptibly with the thoracic cord, causing the tethering to the skin. She made an uneventful recovery, without any neurological sequelae. The follow-up at 24 months from surgery revealed a normal development with good control of her lower limb function.

Conclusion

Only two case reports of amniotic band causing tethering of the spinal cord have been reported so far. An additional variant case of ABS with open meningocele but with similar characteristics has also been reported. Neural tube defects involving head and spine are thought to result from adhesions between craniofacial structures and chorionic wall or compressions forces by amniotic bands. Tethering of the thoracic spinal cord with amniotic band is an exceedingly rare occurrence. It is a rare entity but can be treated with a conventional approach with a favourable outcome.

References

1. Bourne MH, Klassen RA (1987) Congenital annular constricting bands: review of literature and a case report. *J Ped Orthop* 7:218–221
2. Cincore V, Niniios AP, Pavlik J, Hsu CD (2003) Prenatal diagnosis of acrania associated with amniotic band syndrome. *Obstet Gynecol* 102:1176–1178
3. Jackson T (2001) Open thoracic meningocele associated with amniotic band syndrome. *Pediatr Neurosurg* 34:252–254
4. Kawamura K, Chung KC (2009) Constriction band syndrome. *Hand Clin* 25:257–264
5. Lockwood C, Ghidini A, Romero R, Hobbins JC (1989) Amniotic band syndrome: reevaluation of its pathogenesis. *Am J Obstet Gynecol* 160:1030–1033
6. Mohamed MA, Abdurehman P, Jose J (2008) A case of tethered cord associated with amniotic band syndrome. *J Assoc Physicians India* 56:1000
7. Moreman P, Fryns JP, Vandenberghe K, Lauweryns JM (1992) Constrictive amniotic bands, amniotic adhesions and limb–body wall complex: discrete disruption sequences with pathogenetic overlap. *Am J Med Genet* 42:470–479
8. Paletta CE, Huang DB, Sabeiro AP (1999) An unusual presentation of constriction band syndrome. *Plast Reconstr Surg* 104:171–174
9. Prabhu VC, Tomita T (1999) Spinal cord tethering associated with amniotic band syndrome. *Paediatr Neurosurg* 31(4):177–182
10. Soldado F, Aguirre M, Peiro JL, Carreras E, Arevalo S, Fontecha CG, Velez R, Barber I, Martinez-Ibanez V (2009) Fetoscopic release of extremity amniotic band with risk of amputation. *J Pediatr Orthop* 29:290–293
11. Torpin R (1968) Fetal malformations caused by amnion rupture during gestation. Charles C Thomas, Springfield, pp 1–76
12. Walter JH Jr, Goss LR, Lazzara AT (1998) Amniotic band syndrome. *J Foot Ankle Surg* 37:325–333