

Case Report

Posterior cervical meningocele with tethered cord: A rare form of spinal dysraphism

Sunil Upadhyaya^{1*}, Rajesh Nair^{*1}, Lakshman I Kongwad¹, Vinod Kumar¹, Bhavna Nayal²

¹Department of Neurosurgery, Manipal University, Manipal

²Department of Pathology, Manipal University, Manipal

Abstract

Meningocele is a complex congenital form of spinal dysraphism secondary to neural tube defects that occur during the third to fourth week of gestation. This form of spina bifida cystica (open type) requires early diagnosis and surgical intervention. Untreated, they develop the dreaded combination of spontaneous/traumatic rupture and encephalitis and the eventually fatal sepsis, secondary to florid meningitis. Cervical meningocele and myelomeningocele are rare spinal dysraphic lesions. There is often no neurological deficit in infants with cervical lesions, thus the subtle features of cervical cord tethering may be overlooked on imaging. Here, a case of cervical meningocele with tethered cord is reported in a 17-day-old girl. The tethering band, confirmed intra-operatively, was evident on imaging. Treatment aims to prevent future neurological deterioration, and should include careful intradural exploration with detethering of the cord.

Keywords: Spinal dysraphism, Cervical meningocele, Spina bifida.

INTRODUCTION

Spinal meningocele, by definition, are protrusions of the dura and arachnoid mater through defects in the spinal

column, with the spinal cord remaining within the spinal canal (Senoglu, 2008). These are much less frequent than myelomeningoceles, usually posterior, but can occur anteriorly in association with 'sacral agenesis' or in antero-lateral position at lumbar, thoracic or cervical levels (Senoglu, 2008).

Established facts

1. Meningocele is a complex congenital form of spinal dysraphism secondary to neural tube defects that occur during the third to fourth week of gestation.
2. Early diagnosis and prompt treatment ensures excellent outcome.

Cervical meningocele and myelomeningocele (Figure 1) are rare lesions that comprise only a small proportion of neural tube anomalies. Previous studies have reported that only 3.9% to 8.0% of spina bifida cystica occurred in the cervical region (Fisher, 1952) (Doran, 1961) (Barson, 1970). Diagnosis of cervical meningoceles and myelomeningoceles are obvious at birth: a mass protrudes from the posterior midline of the neck.

Novel insights

- ✚ Tethered cord should be highly suspected in cervical meningocele/meningomyelocele with intact neurology.
- ✚ Prompt investigation should include high-resolution MRI or computed tomography.
- ✚ In our case, the dorsal tenting of the cervical cord at the C5-6 level, which was suggestive of tethered cord, was demonstrated on MRI.
- ✚ The definitive treatment aims to prevent future neurological deterioration, and should include thorough intradural exploration with untethering of the cord.

Children with cervical tethered cord can be asymptomatic (Table 1) and often come to medical attention for a midline cutaneous lesion on the back. Although cutaneous lesion normally associated with spinal dysraphism may occur in upto 5% of healthy children. (McAtee, 1994) (Senoglu, 2008)

Unlike their lumbosacral counterparts, the base of these cervical lesions is covered with full thickness skin, and typically do not have a cerebrospinal fluid fistula. Treatment is mainly cosmetic: superficial ligation of the dural fistula and resection of the sac only. Patients

*Corresponding Author Email: rajeshnair39@yahoo.com;
Sunil19upadhyaya@gmail.com

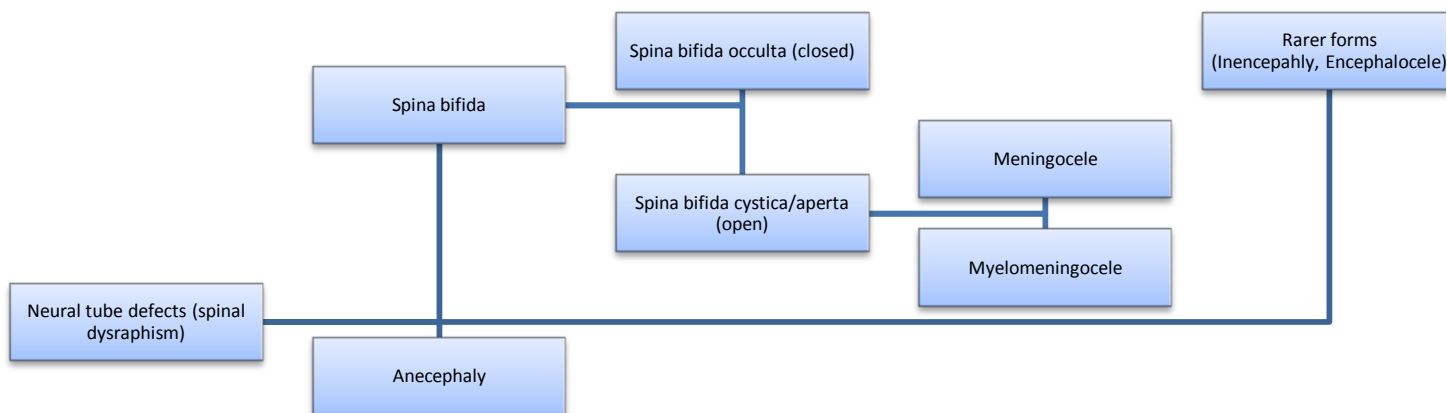


Figure 1. Classification of spinal dysraphism

Table 1. Classification of open and closed spinal dysraphism

Open spinal dysraphisms	Closed spinal dysraphisms	
	With subcutaneous mass	Without subcutaneous mass
Myelomeningocele	<i>Lumbosacral</i>	<i>Simple dysraphic states</i>
Myelocele	• Lipomas with dural defect	• Intradurallipoma
Hemimyelomeningocele	✓ Lipomeningocele	• Filarlipoma
Hemimyocele	✓ Lipomyelocele	• Tight filumterminale
	• Terminal myelocystocele	• Persistent terminal ventricle
	• Meningocele	• Dermal sinus
	<i>Cervico-thoracic</i>	<i>Complex dysraphic states</i>
	• Nonterminal myelocystocele	• Disorders of midline notochordalintegration
	• Meningocele	✓ Distematomyelia
		✓ Neurenteric cysts
		✓ Dorsal enteric fistula
		• Disorders of segmental notochordal formation
		✓ Caudal agenesis
		✓ Segmental spinal dysgenesis

nonetheless experience progressive neurological deterioration due to the untreated tethered cervical spinal cord.

The ensuing neurological deficits are disabling, and affect mainly the fine motor functions of the hands. Prophylactic resection of all the tethering bands and septa is thus advocated (McAtee, 1994) (Seichi, 1993). We report the case of an infant with cervical meningocele and associated tethered cervical cord, who underwent intradural exploration, detethering of the cord, and sac resection.

Case report

The patient was a 17-day-old girl born to a 26-year old G2P1Ab1 lady. The mother had a prior history of spontaneous abortion secondary to anencephaly in the fetus, diagnosed during her anomaly scan in the 2nd

trimester. In her present pregnancy, the infant had a cystic posterior neck mass had been present since birth and was 3.0 cm wide x 2.0 cm long x 2.0 cm high at the time of admission to the neonatal intensive care unit (NICU). Her perinatal history was unremarkable. She had regular antenatal checkups (ANCs) with adequate and monitored folate, iron and calcium supplementation during the entire pregnancy. At her 7th month ANC, she was informed about a swelling in the back of the neck of the growing foetus. The infant was delivered via an elective LSCS in view of maternal anaemia and referred to a higher centre for definitive treatment.

In July 2014, aged 1 day, she was admitted to the hospital for management of the neck mass. Physical examination revealed a posterior midline neck protrusion measuring 3.5 cm wide x 3.0 cm long x 2.0 cm high (Figure 2). The sac was round, fluctuant, and had a wide, sessile base covered with full thickness skin. The apex of the sac was covered with a tough purplish



Figure 2. Preoperative clinical image showing a posterior midline neck protrusion measuring 3.5 cm wide x 3.0 cm long x 2.0 cm high (a), brilliantly trans-illuminant (b & c) with a purplish hue (d) at the summit, suggestive of a cervical meningocele/meningomyelocele.

membrane that comprised about 30% of the total surface area of the sac. There was no sign of cerebrospinal fluid leakage. The infant had no focal neurological deficit and was playful and interested in her surroundings. She had spontaneous movements in all four extremities and spontaneous voiding and was feeding well. Growth parameters were normal, including head circumference. Developmental assessment revealed normal developmental milestones, although there was a slight delay in gross motor aspects. Plain cervical spine X-ray revealed no gross lamina or bony defects.

MRI revealed a defect in the posterior elements of C5 vertebra. Herniation of meninges was seen through the defect causing external swelling of size ~3.0 x 2.8 cm (AP x CC) at this level with no post contrast enhancement of the mass. Cord was seen tethered to the defect; however neural placode was well within the spinal canal with an intact skin over the swelling. A low-signal connection between the posterior bulge of the cord and the dorsal dural sac, which could represent the tethering stalk, was identified (Figure 3). The spinal cord was otherwise normal, with no Chiari malformation, split cord malformation, or low-lying cord. Brain MRI revealed no hydrocephalus or other anomaly.

Operative Procedure

Excision of the cervical meningocele and detethering of the cervical cord was performed under general anesthesia. The patient was positioned prone and her head stabilized using a horse-shoe frame. An elliptical incision, oriented horizontally, was made passing the skin around the base of the meningocele down to the C6 level. The edges of the meningocele with the overlying skin and layers were dissected, and the sac confirmed.

The neck of the meningocele was narrow and atretic, with trapped cerebrospinal fluid within the lumen of the meningocele, and could be traced entering the spinal canal via the C5 lamina defect. The meningocele sac was transected, with the proximal part exposed, and no outflow of cerebrospinal fluid was detected. A linear durotomy was made at the neck of sac after C4-C6 laminectomy. Opening of the sac revealed whitish fibrous tissues that connected the dorsal cervical cord to the sac of the meningocele (Figure 4). These fibrous tissues were taut, further confirming the suspicion of tethered cord. The fibrous tissues on the dorsal cord were attached at the rostral end of the sac. All were subsequently excised and the spinal cord detethered. No split cord malformation was noted. The dura was

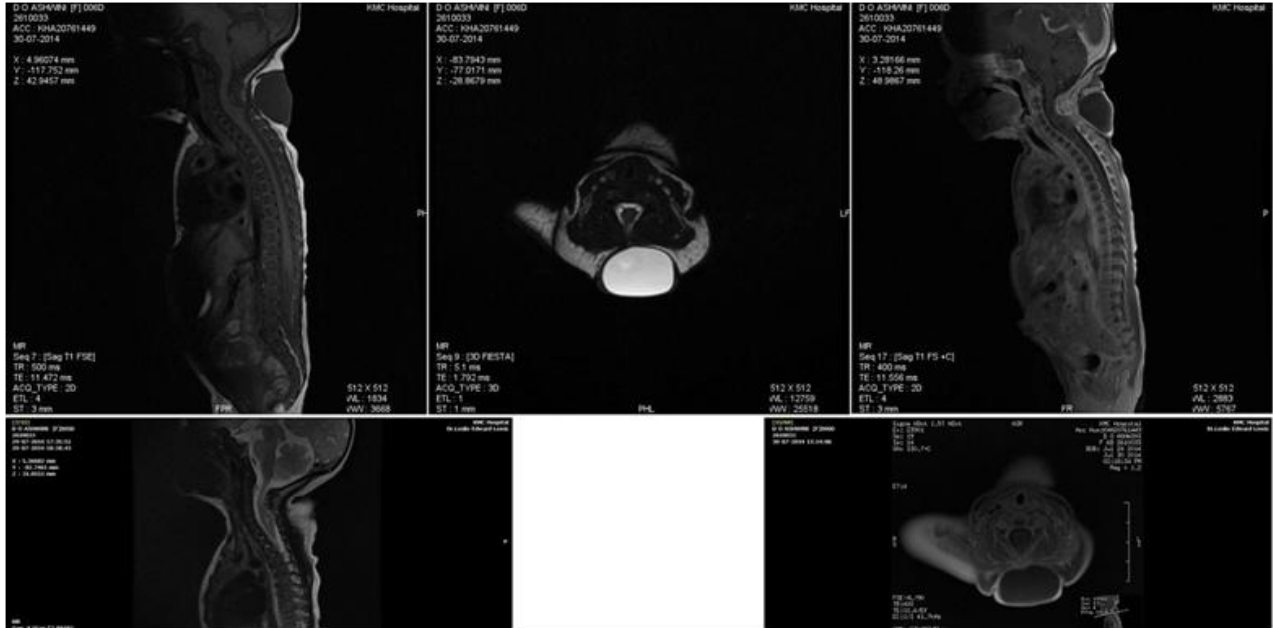


Figure 3. Preoperative magnetic resonance imaging (MRI) revealed a defect in the posterior elements of C5 vertebra (T1 weighted image). Herniation of meninges was seen through the defect causing external swelling of size ~3.0 x 2.8 cm (AP x CC) at this level with no post contrast enhancement of the mass. A low-signal connection between the posterior bulge of the cord and the dorsal dural sac, which could represent the tethering stalk, was identified.



Figure 4. Intraoperative images

- a. Meningocele sac opened which showed trapped CSF with no communication with the underlying dural tube.
- b. Meningocele sac with the skin tag and tethering stalk dissected till underlying normal dural tube was seen.
- c. Normal looking proximal and distal dural tube was dissected and dura exposed to show the tethered stalk between the meningocele sac and normal cord below.
- d. Meningocele sac along with fibrous tract excised with underlying tented cord dissected and laid free into the dural tube.

repaired and the overlying layers closed. Postoperative recovery was uneventful (Figure 5).

DISCUSSION

Despite the paucity of reported cases, (Eller, 1987) (Pang, 1993) (Meyer, 2003) (Myles, 2002) it is evident that tethered cervical cord is closely associated with cervical myelomeningocele and meningocele. If left untreated, the tethered cervical cord is likely to cause gradual neurological deterioration over the years with motor function in the upper extremities being primarily

affected.

Most posterior meningoceles are not 'pure' in that they contain aberrant nerve roots adherent to the inner wall, occasional ganglion cells or even glial nodule that may represent a diverticulum of the central canal of the spinal cord.

Two separate mechanisms of tethered cervical cord have been suggested.

- The first mechanism is a taut fibro-neurovascular stalk that extends from the dorsal column of the cord to the dorsal dura of the sac. This phenomenon was named limited dorsal myeloschisis, an additional embryological defect (Pang and Dias 1993).



Figure 5. Postoperative clinical image showing a well healed scar on followup.

- The second, less common mechanism occurs in a cervical myelomeningocele that contains a type II split cord malformation, a dorsal band of connective tissue, nodules within the sac, an ependymal lined cavity (Steinbok and Cochrane 1995). In this situation, a median fibrous septum between the two hemicords tethers them to the dorsal dura. (Pang, 1993)

In this case, two features of the MRI scan suggested a tethered cervical cord. The first—a posterior tented bulge of the cord at lower end of C5, which was more apparent in the sagittal images wherein a change in the longitudinal contour was more easily visualized. This feature has been previously reported.(Eller, 1987) (Pang, 1993) The second feature was a posterior tenting of the dorsal dura just anterior to the C5 lamina defect. Extending a straight line joining the two most tented points on the posterior bulge of the cord and the dorsal dura backward would pass through the lower edge of the lamina defect into the meningocele sac, and indicate the line of tension to the tethered cord. Preoperative imaging (especially magnetic resonance imaging {MRI}) helps determine the extent of surgical exploration required however computed tomography is reported to be more

sensitive than MRI in localizing a fibro-neurovascular stalk and median fibrous septum of split cord malformations, and delineating the details of such malformations.(Pang, 1993) (Meyer, 2003) (Myles, 2002)

Since the lesion is covered with skin, a surgical correction can be done electively, although in many instances the defect is repaired before the newborn leaves the hospital. Intraoperative, the sac usually narrows down to a neck, which is usually adherent to the normal cord via a fibrous stalk. Excess dura is usually trimmed and closed with absorbable sutures. The redundant skin is used for tension-free closure. The prognosis for these babies with a 'closed' NTD is generally excellent.

CONCLUSION

Tethered cord should be highly suspected in cervical meningocele/meningomyelocele with intact neurology. Prompt investigation should include high-resolution MRI or computed tomography. In the present case, the dorsal tenting of the cervical cord at the C5-6 level, which was

suggestive of tethered cord, was demonstrated on MRI. Cord tethering was confirmed intra-operatively and released. The definitive treatment aims to prevent future neurological deterioration, and should include thorough intradural exploration with untethering of the cord.

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How to cite this article: Upadhyaya S, Nair R, Kongwad LI, Kumar V, Nayal B (2015). Posterior cervical meningocele with tethered cord: A rare form of spinal dysraphism. *Int. Inv. J. Med. Med. Sci*. Vol. 2(2): 23-28