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Cervical meningocele with tethered cervical cord in a Chinese infant

華裔嬰兒出現頸部脊膜膨出並伴有頸髓栓系

Cervical meningocele and myelomeningocele are rare spinal dysraphic lesions. Unlike lumbosacral 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. We report a case of cervical meningocele in an 8-month-old girl. The tethering band, confirmed intra-operatively, was not evident on imaging. Untethering of the cord was performed together with resection of the sac and repair of the dura. Tethered cord should be suspected in the presence of cervical meningocele and intact neurology. It should be carefully looked for using high-resolution magnetic resonance imaging or computed tomography. Treatment aims to prevent future neurological deterioration, and should include careful intradural exploration with untethering of the cord.

頸部脊膜膨出與脊髓脊膜膨出同屬罕見的先天性脊柱裂。與腰骶椎部病變不同，嬰兒頸椎脊膜膨出一般不伴有神經功能缺損。因此，在影像檢查中頸髓栓系的微細特徵可能會受到忽略。本文報告一名8個月大的女嬰患有頸部脊膜膨出並接受手術切除和修補。術中發現有頸髓栓系，但術前影像未能明確顯示。作者認為即使神經功能正常，醫務人員應留意所有頸部脊膜膨出患者是否伴有頸髓栓系，並應採用高解像度磁力共振成像或電腦斷層掃描進行仔細檢查。手術中要進行硬脊膜內探查並鬆解脊髓栓系，從而預防將來的神經功能損害。

Key words:
Meningocele;
Meningomyelocele;
Neck;
Neural tube defects;
Spinal dysraphism

關鍵詞：
脊膜膨出；
脊髓脊膜突出；
頸部；
神經管缺陷；
先天性椎弓缺損

Hong Kong Med J 2005;11:113-5

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Introduction

Cervical meningocele and myelomeningocele 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.[1-3]

Diagnosis of cervical meningoceles and myelomeningoceles are obvious at birth: a mass protrudes from the posterior midline of the neck. 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. Most patients have no neurological deficit at presentation and treatment has been mainly cosmetic: superficial ligation of the dural fistula and resection of the sac only. Patients 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.[4]

We report a case of an infant with cervical meningocele-associated tethered cervical cord, who underwent intradural exploration, untethering of the cord, and sac resection.

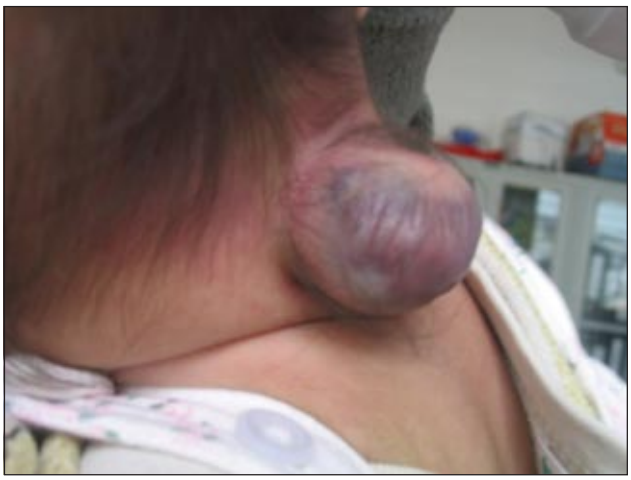

Fig 1. External features of cervical meningocele, with a wide, sessile base covered with full thickness skin

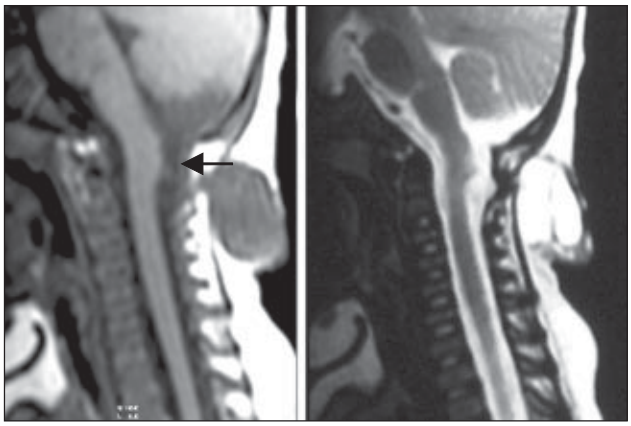

Fig 2. Preoperative magnetic resonance images of the cervical meningocele
T1-weighted sagittal magnetic resonance image shows dorsal tented bulge of the cervico-medullary junction. Tethering stalk is observed (arrow)

Case report

The patient was an 8-month-old girl from an orphanage in Mainland China. 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 orphanage. Her perinatal history was unknown.

In August 2004, aged 8 months, she was admitted to the author's unit 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 (Fig 1). 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 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. Growth parameters were normal, including head circumference. Developmental assessment revealed normal developmental milestones, although there was a slight delay in gross motor aspects. She was able to sit without support and stand with support.

Plain cervical spine X-ray revealed no gross lamina or bony defects. Magnetic resonance imaging (MRI) of the cervical spine showed a fluid-filled septated cyst extending from the subcutaneous tissue of the posterior neck to the skin, compatible with a cervical meningocele (Fig 2). The left lamina of C1 was absent and that of C2 was smaller, suggesting that the origin of the meningocele sac was at the level of C1. A posterior tented bulge of the cervical cord was also present at the cervico-medullary junction, and there was a posterior tenting of dorsal dural sac at the rostral end. A low-signal connection between the posterior bulge of the cord and the dorsal dural sac, which could represent the tethering stalk, was identified (Fig 2). 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.

Excision of the cervical meningocele and untethering of the cervical cord was performed under general anaesthesia. The patient was positioned prone and her head stabilised using a head-frame. A suboccipital vertical midline incision was made passing the skin around the base of the meningocele down to the C3 level. The edges of the meningocele with the overlying skin and layers were dissected, and the sac confirmed. The left C1 lamina was absent. The neck of the meningocele was narrow and atresic and could be traced entering the spinal canal via the C1 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. Opening of the sac revealed whitish fibrous tissues that connected the dorsal cervical cord to the sac of the meningocele. 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 untethered. No split cord malformation was noted. The

dura was repaired and the overlying layers closed. Postoperative recovery was uneventful.

Discussion

Despite the paucity of reported cases,[4-7] 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. Two separate mechanisms of tethered cervical cord have been suggested. The first mechanism is a taut fibroneurovascular stalk that extends from the dorsal column of the cord to the dorsal dura of the sac. This phenomenon was named limited dorsal myeloschisis by the author. The second, less common mechanism occurs in a cervical myelomeningocele that contains a type II split cord malformation. In this situation, a median fibrous septum between the two hemicords tethers them to the dorsal dura.[5]

In this case, two features of the MRI scan suggested a tethered cervical cord. The first—a posterior tented bulge of the cord at the cervico-medullary junction—was more apparent in the sagittal images wherein a change in the longitudinal contour was more easily visualised. This feature has been previously reported.[4,5] The second feature was a posterior tenting of the dorsal dura just anterior to the 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. Computed tomography is reported to be more sensitive than MRI in localising a fibroneurovascular stalk and median fibrous septum of split cord malformations, and delineating the details of such malformations.[5]

Conclusion

Posterior tenting of the cervico-medullary junction and the dorsal dura, suggestive of tethered cord, were demonstrated on the MRI in a Chinese infant with cervical meningocele. Cord tethering was confirmed intra-operatively and released. Tethered cord should be highly suspected in cervical meningocele with intact neurology. Careful investigation should include high-resolution MRI or computed tomography. The definitive treatment aims to prevent future neurological deterioration, and should include thorough intradural exploration with untethering of the cord.

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