

Cervical meningocele with tethered cord in a seven-years old child: Case Report

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Citation

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Abstract

Meningoceles are typically associated with other congenital spinal anomalies, and are usually diagnosed in childhood. Cervical meningoceles are extremely uncommon congenital spinal anomalies. Most patients have no neurological deficit at presentation and treatment has been mainly cosmetic. Treatment aims to prevent future neurological deterioration, and should include careful intradural exploration with untethering of the cord. We report a case of cervical meningocele in a 7-year-old girl. The tethering band, confirmed intra-operatively. Untethering of the cord was performed together with resection of the sac and repair of the dura. Early intervention is recommended even in cases with normal neurological examinations in order to prevent deterioration. These cases should be operated as soon as possible after birth.

INTRODUCTION

Cervical meningoceles and myelomeningoceles account for only 4% to 8% of spina bifida cystica lesions. Cervical meningoceles comprise only a small proportion of neural tube defects [123]. A meningocele is a single, posteriorly herniating cystic sac. The sac is cerebrospinal fluid filled and is lined by arachnoid and dura [4]. There may be a band of tissue that tents the posterior aspect of the cervical cord and extends into the wall of the sac either to the base or the dome [5]. A small number of reports have noted that meningoceles may feature associated tethering of the cervical spinal cord [126].

In this report, we presented a 7 year-old patient harboring a meningocele in the upper cervical region with neurological deficit and tethered cord. The lesion was treated surgically.

CASE HISTORY

A 7-years-old girl suffering from a posterior mass in the cervical region was admitted to our clinic (Figure 1).

Figure 1

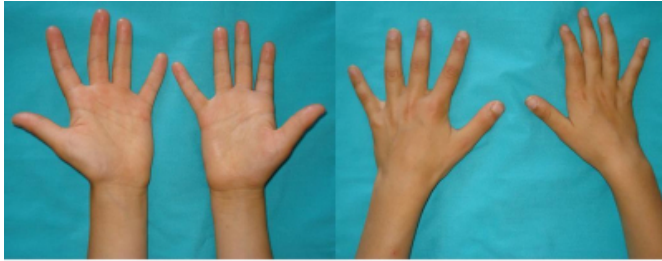
Figure 1: A 7-year-old child with a cervical meningocele. External features of cervical meningocele, with a wide, sessile base covered by full thickness skin and by squamous epithelium at the dome.



On physical examination, the only abnormality detected was a 5-cm-diameter sac in the posterior cervical area. It was covered at the base by full-thickness skin and covered on the dome by thick squamous epithelium (Figure 1). Neurological examination revealed right monoparesis, and atrophy (Figure 2).

Figure 2

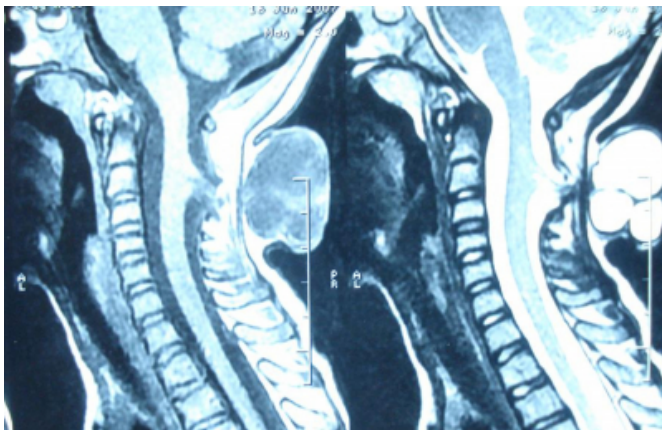
Figure 2: Right upper extremite shows atrophy.



Patient had no urinary dysfunction. The results of routine laboratory tests were unremarkable. The magnetic resonance imaging (MRI) of the cervical spine revealed a meningocele at C2-3 level and the spinal cord was tethered transversely to the posterior dura mater at same levels (Figure 3).

Figure 3

Figure 3: Magnetic resonance images of the cervical meningocele T1 and T2-weighted sagittal magnetic resonance images shows dorsal tented bulge of the upper cervical region. Tethering stalk is observed.



The spinous process of C2 was absent. The origin of the meningocele sac was at the level of C2. Patient had no urodynamic dysfunction. The patient was operated under general anesthesia in prone position, her head stabilized using a head-frame and the meningocele was corrected without any complication. Cord tethering was confirmed intra-operatively and released. Postoperative neurological examination of the patient was unchanged. Patient was discharged on the fourth postoperative day.

DISCUSSION

The recurrence risk after the birth of an infant with isolated spina bifida is 3-5%. Recurrence may be for spina bifida or another type of spinal abnormality [7].

Folic acid is involved in the synthesis of nucleic acids composing DNA, genes, and chromosomes; furthermore, folic acid is metabolized to tetrahydrofolic acid (THFA), which is one of three coenzymes (B6 and B12) that converts homocysteine to methionine [78]. If folic acid or THFA decreases, homocysteine will increase. Such increase is linked to an increase risk of early pregnancy loss, pregnancies complicated by cleft lip and spina bifida [789]. A lack of folic acid can explain elevated plasma homocysteine levels. In addition, it can explain up to 50% of the protective effect of folic acid against spina bifida[2].

There is clear evidence that a large proportion of spina bifida is preventable by periconceptional folic acid intake[78]. The timing of folic acid supplementation is considered as significant factor to prevent spina bifida which is more effective when treatment is initiated at least 1 month prior to conception[6]. In September 1992, the United States Public Health Service (USPHS) issued the recommendation that all women of child-bearing age who are capable of becoming pregnant need to consume 0.4 mg of folic acid daily, and for women who already have had an spina bifida-affected pregnancy should receive 4 mg of folic acid every day starting 1-3 months prior to the planned conception and continuing throughout the first 3 months of pregnancy [78910].

There are only a few published series of cervical meningocele in the literature. MRI is the imaging modality of choice for suspected cervical meningocele, prior to operation. It helps distinguish the subtypes, as well as identify the associated anomalies [11]. Cervical meningoceles differs from typical lumbar meningocele, in that the nerve fibres within cervical meningoceles are a vertical outgrowth from the cord and traverse the sac. In contradistinction, the neural placode in the lumbar region is generally fixed, flattened and exposed [1].

The cervical region is the most mobile part of the spinal column, consequently when the cord here becomes tethered, any movement—even one in the normal range of motion—can exert stretching forces on the cord [212]. Stretching can lead to strain that exceeds the compensatory properties of the spinal cord and results in neurologic deterioration [2].

In presented case, the patient showed neurologic deterioration and this was a clear clinical sign for cervical tethered cord. Our case presented with atrophy and monoparesis. Presenting with atrophy is rare. During the operation, we detected fibrovascular bands that were

tethering the cord dorsally. We also found that the arachnoid membrane near the meningocele neck was thickened. The incidence of retethering caused by postoperative adhesions at the repair site after initial tethered spinal cord surgery is not uncommon.

The tethered cervical cord is likely to cause gradual neurologic deterioration eventually, with motor function in the upper extremities mainly affected [16131415]. Therefore, whenever a cervical meningocele is detected, early intervention is recommended even in cases with normal neurological examinations in order to prevent deterioration.

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