Anterior cervical myelomeningocele: a rare malformation of the spinal cord

Case report

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Cervical myelomeningocele (MMC) is an uncommon congenital malformation of the spinal cord and accounts for a small proportion of neural tube defects. These lesions mostly occur in the dorsal part of the body. Only a single case of anterior cervical MMC has been previously reported. The authors report a second case of anterior cervical MMC diagnosed when the patient began to experience symptoms of bilateral hand weakness in adulthood. In this patient, MR imaging of the cervical spine showed an anterior cervical MMC at the C6–7 level with hydrocephalus, thinning of the genu and trunk of the corpus callosum, maldevelopment of the cerebellar tonsils, and expansion of the fourth ventricle, posterior cranial fossa, and subarachnoid space. A CT scan and a 3D CT reconstruction of the cervical spine clearly demonstrated contiguous fusions of multiple lower-cervical vertebrae and neural arches, which was consistent with Type III Klippel-Feil syndrome. The patient was advised to undergo operative treatment to prevent the progression of her neurological deficit. However, after being notified of the potential neurological risks, the patient declined surgery and opted for conservative treatment with a hard neck collar. At 4 months’ follow-up, the patient’s neurological deficit remains stable with the MMC left untreated. The authors presume that the possible pathogenesis of anterior cervical MMC may greatly differ from that of posterior lesions. This lesion could also be associated with multiple other spinal abnormalities, which highlights the importance of comprehensive preoperative radiological examinations. (DOI: 10.3171/2011.10.SPINE11484)

Key Words • cervical • myelomeningocele • malformation • spinal cord

Cervical myelomeningocele is a rare congenital lesion, accounting for only 1%–5% of all neural tube anomalies.8,9 This lesion mostly occurs in the dorsal part of the body and is typically characterized by a skin-covered mass in the posterior midline. In our review of the literature, we found only one reported case of an anterior cervical MMC.3 Here, we report a second case, which was diagnosed when the patient began to experience symptoms in adulthood.

Case Report

History and Presentation. This 23-year-old woman presented to our outpatient clinic with a 1-month history of rapidly progressive weakness in both hands. She was generally in a good state of health and had been completely asymptomatic until the recent onset of weakness. Physical examination revealed mild limitations of neck extension and flexion. A comprehensive and careful neurological examination was performed, and the only neurological abnormality detected was decreased grip strength in both hands.

Imaging Studies. The patient had undergone several radiological evaluations, including MR imaging and CT (with 3D CT reconstruction of the cervical spine), in a local hospital before her first visit to our outpatient clinic. Sagittal MR images showed an anterior MMC at C6–7 with hydrocephalus, thinning of the genu and trunk of the corpus callosum, maldevelopment of the cerebellar tonsils, and expansions of the fourth ventricle, posterior cranial fossa, and subarachnoid space. At C6–7, the cord was kinked to form an anterior loop and was anteriorly prolapsed into a ventrally herniated meningeal cyst (Fig. 1). Axial MR images revealed an anterior meningocele protruding through the defects of the vertebral body and containing a portion of the cervical spinal cord. Part of the cervical spinal cord was located in the spinal canal while part was located anteriorly outside the vertebral body (Fig. 2). The CT scan and the 3D CT reconstruction of the cervical spine clearly demonstrated contiguous fusions of multiple lower cervical vertebrae and neural arches (Fig. 3), which was consistent with Type

Abbreviation used in this paper: MMC = myelomeningocele.
III Klippel-Feil syndrome according to the classification of Guille et al.5 No further imaging examinations of other organs were performed because the patient had no complaint related to any other organ system and no other abnormality was detected through physical examination. Dynamic cervical radiographic examination was also not performed in this patient for fear of deterioration of neurological deficit.

Treatment. Because of the rapid progression of the patient’s neurological deficit, we recommended surgical treatments to prevent further deterioration. A single-stage combined anterior-posterior procedure was proposed, with the posterior surgery consisting of C5–7 laminotomy and instrumentation placement and the anterior surgery consisting of C5–7 corpectomy with or without duraplasty. After being notified of the potential neurological risks, however, the patient declined surgery and opted for conservative treatment with a hard neck collar. At 4 months’ follow-up, the patient’s neurological deficit remains stable.

Discussion

Cystic dysraphic lesions of the cervical spine are rare conditions, with only a few cases reported in the literature.11,13 These lesions can be divided into several categories that include cervical myelocystocele, cervical meningocele, and cervical MMC.5,90 Differentiation of the subtypes of cystic dysraphic lesions is based on the distinct pathomorphological features of these lesions.2,9 The classic morphological feature of MMC is a cystic protrusion of meninges through a defect of the spinal column with the herniated spinal cord as its contents.10 Magnetic resonance imaging is an excellent tool for diagnosing spinal abnormalities because it can accurately demonstrate the morphological properties of these lesions.2,9,14 Because the protruding meningeal cyst with the kinked spinal cord inside is clearly shown in the MR imaging of cervical spine in our case, we believe that the lesion should be diagnosed as a cervical MMC.

Anterior cervical MMCs represent an extremely rare subset of spina bifida cystica lesions in contrast to the more commonly occurring posterior type. The case reported by Balachandran3 is the only previously reported example. The possible pathogenetic mechanism of anterior cervical MMC was not discussed in that report, and it may greatly differ from that of posterior cervical MMC. The posterior lesions are caused by delayed primary neurulation, which prevents the neural tube from closing dorsally.7,15 The incomplete fusion of dorsal tube leads to the failure of neural ectoderm to separate from the cutaneous ectoderm. Consequently, a fibroneurovascular stalk arises from the dorsal surface of the cervical cord and penetrates through the dorsal dural opening to connect to the skin.5,7 This delayed neurulation hypothesis, however, could not explain the development of anterior cervical MMC. The process of dorsal fusion of the neural tube may be normal in the anterior lesion, given that no dorsal herniation of neural structures occurs. The more acceptable mechanism of anterior cervical MMC is the maldevelopment of the cervical spine.
of mesoderm surrounding the neural tube and notochord, from which the vertebral column and the membranes of the spinal cord are developed. In our patient, we presumed that the maldevelopment of the mesoderm could lead to the vertebral body defect and anterior dysplasia of the meninges. As the CSF forms, it gradually squeezes the dysplastic anterior wall of meninges and subsequently provokes the anterior protrusion of meninges and spinal cord through the vertebral body defect, thus producing the anterior MMC.

It is well documented that posterior cervical MMC is often associated with other spinal abnormalities, such as hydrocephalus, hydromyelia, diastematomyelia, Chiari malformation, Klippel-Feil syndrome, and so on. Similar to the patients with posterior cervical MMC, our patient had multiple associated abnormalities of the nervous system. The sagittal MR images demonstrated hydrocephalus, thinning of the genu and trunk of the corpus callosum, maldevelopment of the cerebellar tonsils, and expansion of the fourth ventricle, posterior cranial fossa, and subarachnoid space. The CT scan and 3D CT reconstruction of the cervical spine clearly showed Type III Klippel-Feil syndrome. In Balachandran's case, Klippel-Feil syndrome was also detected. Magnetic resonance imaging is recommended for patients with cervical MMC to depict the morphological properties of the lesions and detect any other associated spinal anomalies, since failure to evaluate the anatomical details of the lesion and associated anomalies may result in unsuccessful treatment. Computed tomography and 3D CT reconstruction may be helpful in identifying complex associated abnormalities of bony structures.

There are 2 limitations that should be mentioned in our report. First, although the neurological symptoms of our patient had not progressed at the 4-month follow-up visit, we acknowledge that further long-term follow-up is needed to assess the natural history of the anterior cervical MMC more accurately. Second, our patient did not undergo a brain CT scan, which might have comprehensively demonstrated hydrocephalus of each ventricle, especially the lateral ventricles, which could not be well visualized in the sagittal MR images.

Conclusions

Our case is of great interest because of the extremely uncommon occurrence of anterior cervical MMC. To the best of our knowledge, this is the second case reported in the English literature. The possible pathogenesis of anterior cervical MMC may greatly differ from that of posterior cervical MMC. Although etiologically different from the posterior lesion, anterior cervical MMC could also be associated with multiple other spinal abnormalities, which highlights the importance of comprehensive preoperative radiological examinations.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Jiang, Qiu. Acquisition of data: Jiang, Liu, Qiu. Drafting the article: Jiang. Critically revising the article: Qiu, Zhu. Reviewed final version of the manuscript and approved it for submission: all authors. Administrative/technical/material support: Qian, Zhu. Study supervision: Qiu.

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