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Cervical spinal cord atrophy associated with spina bifida

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We treated three patients with spina bifida who had atrophy of the cervical spinal cord. One patient presented with a gradually progressive paresis of the upper right extremity, and two patients developed tetraparesis. MRI (magnetic resonance imaging) of the cervical spine demonstrated cord atrophy with enlargement of the subarachnoid space. The three patients were treated by releasing the tethered spinal cord in the lumbosacral area. Two of the three patients had neurological recovery soon after the operation, but this was transient and was followed by later deterioration; and the third patient showed no recovery. The transient recovery suggests that the tethering in the lumbosacral region affects the entire spinal cord and causes atrophy of the cervical spinal cord.

Keywords: lumbosacral; spina bifida; cervical spinal cord atrophy; magnetic resonance imaging.

Introduction

Patients with lumbosacral spina bifida occasionally present with symptoms in the upper extremities due to cervical cord lesions.[1,2] Syringohydromyelia and the Arnold-Chiari malformation associated with spina bifida are well known cervical cord lesions.[3] However, from our study of the current literature we could not discover any report describing cervical cord atrophy in spina bifida patients causing cervical myelopathy. We present 3 patients with this rare lesion.

Case reports

Case 1

A six day old girl underwent surgery to repair a meningomyelocele. At the age of 6, she presented with numbness and weakness in the right hand. Motor power was graded 2–3/5 in the intrinsic muscles of the right hand, with atrophy. Sensation was disturbed in the whole of the right arm and hand. The deep reflexes in the upper extremities were absent. MRI showed atrophy of the whole of the cervical cord with enlargement of the subarachnoid space and loss of the intumescentia cervicalis. Neither a syrinx nor an Arnold-Chiari malformation was demonstrated (Fig 1). We suspected that there was a relationship between cord tethering and cervical cord atrophy, and thought that this could be the proximate cause of the paresis of the upper extremity. The patient underwent a second operation to release the filum terminale in the lumbosacral area, but there was no improvement postoperatively.

Case 2

A 28 year old woman became unable to walk after falling down. She had a flaccid tetraparesis with a sensory level at C5. The muscle power of the upper extremities improved for a few months but she required crutches for walking. At the age of 37, progressive muscle weakness of the upper extremities was noted. The diagnosis of spina bifida occulta was first made based on the results of myelography. MRI of the cervical spine demonstrated cervical cord atrophy as in case 1. Neither syringomyelia nor the Arnold-Chiari malformation were found. The patient underwent surgery to remove a lipoma and to release the spinal cord in the lumbosacral area. There was partial recovery of muscle power in the upper extremities soon after the operation. At the age of 45, however, tetraparesis is continuing to progress very slowly. The right hand is useless and she needs a wheelchair.

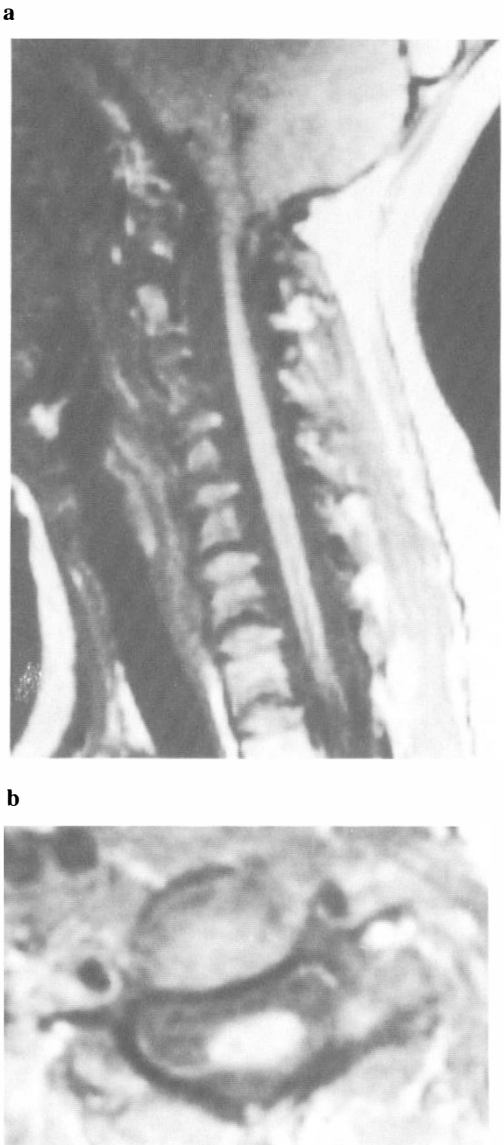

Figure 1 Case 1. T1-weighted sagittal (a) and axial at C6 (b) MRIs showing atrophy of the cervical spinal cord with enlarged subarachnoid space. Intumescentia cervicalis is not observed.

Case 3

A 3 month old girl developed an acute flaccid tetraparesis without any evidence of trauma. A diagnosis of spina bifida occulta was made, and surgery was performed to remove a dermoid cyst and to release the spinal cord. The symptoms resolved soon after the operation. At the age of 6 years, progressive tetraparesis began to develop slowly. Motor power was graded 2–3/5 in the intrinsic muscles of the hands, and she currently needs crutches for walking. MRI of her cervical spine showed cervical cord atrophy without syringomyelia or an Arnold-Chiari malformation.

Discussion

In the cases presented, MRI was helpful in demonstrating cervical spinal cord atrophy and excluding a syrinx or the Arnold-Chiari malformation. The value of MRI in detecting cervical lesions in patients with spina bifida has already been shown for the diagnosis of syringohydromyelia and the Arnold-Chiari malformation.[4] The differential diagnosis should include cervical cord atrophy as a possible cause of cervical myelopathy in patients with spina bifida. The scarcity of previous literature on cervical cord atrophy associated with spina bifida may be due to a failure to recognise the pathology. Careful examination by MRI can increase the ability to diagnose the condition.

The cause of the cervical cord atrophy is not known. The most common cause of spinal cord atrophy is mechanical compression as is seen in spinal spondylosis.[5] However, in the 3 patients that we report, MRI studies did not show cord compression. We speculate that the possible pathomechanisms of cervical cord atrophy are (1) cord tethering, (2) a dysraphic condition, and (3) a spontaneous rupture of the syrinx. In our study, 2 of the 3 patients showed improvement after release of the tethered spinal cord. This suggests that the most probable cause is the tethering, which is known also to affect the lumbar and lower thoracic cord.[6] We feel that the traction force exerted by the tethering affects the whole spinal cord and causes dysplasia of the cervical cord. Case 2 suggests that such conditions are a predisposing cause of spinal cord concussion following minor trauma.

Surgical treatment for cervical spinal cord atrophy is a difficult problem. An operation to release a tethered spinal cord where there is progressive paralysis is recommended by many neurosurgeons.[6,7] We thought that such surgical release in the lumbosacral area

might be effective for cervical myelopathy, but our 3 patients did not show satisfactory long term improvement. The reasons are unclear, but one possibility is that there is retethering by adhesions in the operated region.[8]

References

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