Chiari malformation with compression of the medulla oblongata by the vertebral arteries

Case report

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A unique case is reported of Chiari malformation and compression of the medulla oblongata by both vertebral arteries. A 39-year-old woman complained of unsteady gait and motor weakness of the legs, and magnetic resonance imaging revealed the malformation and compression. Vascular decompression of the vertebral arteries was performed using synthetic (Gore-tex) vascular strips following posterior fossa decompression.

KEY WORDS • Arnold-Chiari malformation • vertebral artery • medulla oblongata • vascular decompression

It is generally known that the symptoms in Chiari malformation are due to the compression of the medulla oblongata by the depressed cerebellar tonsils. We report a case of Chiari malformation with compression of the medulla oblongata by the vertebral arteries. Decompression of both vertebral arteries was carried out using vascular slings following decompression of the posterior fossa.

Case Report

This 39-year-old woman complained of an irregular gait and a loss of strength in both legs. Her medical history revealed nothing of interest. Her elder sister had syringomyelia.

In the autumn of 1984, the patient became aware of an inability to run during a school sports day. Loss of strength in both legs gradually progressed during 1985, and she noticed that her gait had become irregular and that she could not walk in a straight line. During 1987, she experienced occipital pain, numbness of the right arm, and vertical nystagmus of both eyes. In 1988, she noted a gradual decrease in strength on the left side of her body that led to a tendency to drag her left leg. She was admitted to the Department of Neurology on February 6, 1989, and was transferred to the Neurosurgical Department on February 14.

Examination. On admission, the patient's level of consciousness was normal. A leftward-protruding scoliosis was evident. There was no atrophy of the legs, but loss of strength of the muscles on the left side, especially of the left leg, was identified. A slight accentuation of the deep reflexes was noted at all locations, but was particularly marked for the left leg. A positive Babinski sign was seen bilaterally, particularly on the left side. Ocular symptoms included a down-beat nystagmus bilaterally. Disturbances of gait due to trunk ataxia were found, but there were no deficits in coordination or perception.

Laboratory studies revealed no biochemical abnormalities, and neurophysiological examination showed normal auditory brain-stem responses and no significant abnormalities of the somatosensory evoked potentials. Plain skull and cervical x-ray films showed mild platybasia and basilar impression. Leftward scoliosis of the thoracic vertebrae was seen. Computerized tomography (CT) scans suggested atrophy of the cerebellum, and metrizamide-enhanced CT showed a protruding cerebellar tonsil at the foramen magnum. Axial magnetic resonance (MR) imaging demonstrated atrophy of the cerebellum and medulla oblongata, the latter being deformed into a clover-leaf shape at the level of the foramen magnum. Findings suggesting compression
both of the medulla oblongata due to medial deviation of both vertebral arteries were also seen (Fig. 1 left). The cerebellar tonsil leading to the foramen magnum as well as a flow-void signal suggesting compression of the medulla by the vertebral arteries were observed on sagittal MR images (Fig. 2 left). Bilateral vertebral arteries were also observed medially deviating the third segment of both vertebral arteries (Fig. 3 left).

Operation. On February 27, 1990, posterior fossa decompression with laminectomy of the C-1 and C-2 vertebrae was performed. After the dura mater was cut, the cerebellar tonsil was found to have invaginated through the foramen magnum to the C-1 level. Moreover, there was bilateral compression of the medulla oblongata by the vertebral arteries. For this reason, two strips of synthetic (Gore-tex) vascular graft material were cut and formed into loops which were passed around the vessels through a slit in the dura. The vertebral arteries were then pulled laterally, thus decompressing the medulla (Fig. 4).

Postoperative Course. A postoperative vertebral angiogram demonstrated improvement in the medial displacement of the third segment of both vertebral arteries (Fig. 3 right). Sagittal MR imaging showed decompression of the posterior fossa and disappearance of the flow-void signal seen on the preoperative MR image and thought to have been due to the compression of the medulla by the vertebral arteries (Fig. 2 right). Axial MR imaging demonstrated the laterally transposed flow-void signal of the vertebral arteries (Fig. 1 right). The patient is currently under observation as an outpatient and improvements in gait and nystagmus have been realized.

Discussion

Although there is no established hypothesis concerning the mechanism of onset of Chiari malformation, the suggestion of developmental disturbances during the fetal stage has been proposed by Barry, et al., and is consistent with the associated congenital abnormalities of such cases. Among these are abnormalities of the vascular system, such as the looping and descent of the posterior inferior cerebellar artery with associated entrance into the spinal canal at the cerebellum. A less well-known abnormality is the looping of the vertebral arteries within the spinal canal with associated inferior deviation of the cerebellum. However, a case of Chiari malformation in which the medulla oblongata is compressed by abnormal coursing of the vertebral arteries has not been reported previously.

Symptoms of Chiari malformation are predominantly headache and neck pain, followed by sensory and gait disturbances, cerebellar ataxia, and lower cranial nerve abnormalities. Saez, et al., have classified these neurological symptoms into the following six syndromes: foramen magnum compression, paroxysmal intracranial hypertension, central cord disturbances, cerebellar dysfunction, spasticity, and bulbar palsy. The first three syndromes are usually noted on examination, with symptoms due to foramen magnum compression the most frequently reported. A Chiari malformation is thought to be brought about by disturbances of the cerebellum, lower brain stem, and cervical spine at the so-called “craniovertebral junction.”

In our case, in addition to such mechanisms, we believe that bilateral compression of the medulla oblongata by the vertebral arteries may also have been involved.

Posterior fossa decompression is usually the surgical therapy undertaken for Chiari malformation. In our case, both a posterior fossa decompression, including a C-1 and C-2 laminectomy, and vascular decompression with withdrawal of both vertebral arteries were performed. The patient’s symptoms improved postoperatively.

There have been only two previous reports of surgical treatment of compression of the medulla oblongata by
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the vertebral artery. The first was by Kim, et al., in 1985, in which improvements in hemiplegia were noted following microvascular decompression of the medulla by lifting off the vertebral artery. The second case was that of Maruyama, et al., reported in 1989, in which an elongated and tortuous vertebral artery compressed the ventral medulla oblongata, leading to progressive quadriplegia and mild sensory disturbances below the neck region. Neurovascular decompression was performed. In both of these cases, the perforating branches of the vertebral artery were avoided and two Dacron slings were placed between the first dentate ligament and the bifurcation of the posterior inferior cerebellar artery, the end of which was sutured to the dura mater at the edge of the foramen magnum. The vertebral artery was dissected from a portion of the medulla oblongata and a sponge placed between the two structures for decompression. In our case, we passed a synthetic vascular graft around the vertebral arteries in order to pull them laterally from the medulla oblongata. We believe that we achieved decompression without putting undue stress on the vessels involved.

In neither of the two previously reported cases was there an associated Chiari malformation. Although uncertainties remain with regard to the causal relationship between the Chiari malformation and the abnormal course of the vertebral arteries in the present case, we believe it is important to keep the presence of such vascular anomalies in mind and, moreover, to note the extreme effectiveness of MR imaging in delineating compression of the medulla oblongata by the vertebral artery.

References


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