HIARI malformation Type I is a congenital disorder characterized by the caudal descent of the cerebellar tonsils below the foramen magnum; the disorder is accompanied by syringomyelia in 50 to 75% of patients. The mean age of patients with this anomaly on presentation is 41 years, and there is a slight female predominance. The most common symptom is suboccipital headache. During the neurological examination, findings of foramen magnum compression syndrome, central cord syndrome, and cerebellar syndrome are remarkable.

A Chiari malformation may remain asymptomatic until the patient has reached adulthood and acute presentation of symptoms occurs. In several clinical and experimental studies it has been shown that essential hypertension is associated with vascular compression of the brainstem, particularly of the rostral ventrolateral medulla oblongata. Nevertheless, two cases of Chiari malformation and neurogenic arterial hypertension have been reported. In this article the authors describe a patient with Chiari malformation Type I and neurogenic arterial hypertension. A simple suboccipital decompression not only provided neurological improvement, but also led to resolution of the hypertension.

In cases of Chiari malformation and concomitant neurogenic arterial hypertension, careful preoperative clinical and neuroimaging assessments may reveal the cause of the arterial hypertension. Resolution of neurogenic arterial hypertension may be expected even in a case of simple suboccipital decompression.

KEY WORDS • arterial hypertension • Chiari malformation • suboccipital decompression

Case Report

History and Examination. This 51-year-old woman was admitted to our institution with dysphagia, tongue paresthesia, and suboccipital headache. She also suffered from hypertension, which had been diagnosed 1 year before her admission to the hospital. A diagnostic workup showed no cardiac, vascular, endocrine, or renal abnormality. The patient was on a medical regimen that included a calcium-channel blocker and an angiotensin-converting enzyme inhibitor. Despite these medications, her blood pressure was approximately 180/120 mm Hg.

The neurological examination demonstrated indistinctness of the left nasolabial sulcus, left hemihypesthesia, and left sensorineural hearing loss. The patient also had a diminished gag reflex, depressed left palatal arc, and a right deviation of the uvula and tongue—symptoms indicative of a compromise of the ninth and 10th cranial nerves. Her left Babinski reflex was positive and her gait was wide-based and ataxic. Both Romberg and L’hermitte signs were also positive.

Magnetic resonance imaging depicted bilateral cerebellar tonsillar descent to 7 mm below the foramen magnum. The

Abbreviations used in this paper: CSF = cerebrospinal fluid; MR = magnetic resonance; MVD = microvascular decompression; VLMO = ventrolateral medulla oblongata.
tonsils obliterated the subarachnoid spaces at the foramen magnum (Fig. 1 upper left). The shape and signal intensity of the bulbus was preserved. No syrinx was identified within the spinal cord. Phase-contrast angiography images of the CSF flow in the sagittal plane were obtained. We were unable to observe any CSF flow at the dorsal portion of the foramen magnum on these images (Fig. 1 upper right). Cerebrospinal fluid flow velocity was measured at the level of the foramen magnum throughout the cardiac cycle. To calculate the CSF velocity, we included as our region of interest all CSF spaces on the axial image and excluded the vertebral arteries. The peak systolic velocity ranged between 0.26 and 4.81 cm/second, and the peak diastolic velocity ranged between 0.09 and 2.85 cm/second. The velocity distribution was not uniform during either systole or diastole. The peak systolic velocities were a mean 2.69 cm/second, and the diastolic velocities a mean 1.82 cm/second.

**Operation.** Taking into consideration the neurological status of the patient, dorsal decompressive surgery was planned and a suboccipital craniectomy with a laminectomy at C-1 was performed. The dura mater was incised and thick arachnoid adhesions in the region of the cisterna magna and

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**Fig. 1.** *Upper Left:* Midsagittal gradient-echo T₂-weighted MR image (TR 21 msec, TE 6.8 msec) depicting the descending tonsils, which obliterate the entire space posterior to the cervicomedullary junction at the foramen magnum. *Upper Right:* Cerebrospinal fluid flow study obtained in the midsagittal plane, demonstrating no flow signal in the posterior portion of the foramen magnum (arrow). *Lower Left:* Postoperative MR image revealing the occipital craniectomy defect (black arrow). The linear hypointense structure (white arrow) represents the Gortex dural graft, which was placed surgically. *Lower Right:* Postoperative CSF flow study confirming reconstitution of posterior CSF flow at the foramen magnum (arrow).
Chiari malformation and arterial hypertension

foramen magnum were dissected. An expansion duraplasty was performed using a Gortex dural graft. The perioperative and early postoperative periods were uneventful.

**Postoperative Course.** Systemic arterial pressure returned to normal immediately after surgery. While the patient recovered from anesthesia, her blood pressure was 130/85 mm Hg. The postoperative neurological examination revealed resolution of the ataxia, left hemihypertension, and symptoms of left ninth and 10th cranial nerve involvement. During the early postoperative period, the woman’s systemic blood pressure was stable at 120/80 mm Hg, and the course of antihypertension medication was ended. There was no subsequent increase in systemic blood pressure. The patient remained drug-free during her 20-month follow up after surgery and was normotensive. Magnetic resonance imaging with CSF flow measurement was repeated 2 days after surgery (Fig. 1 lower). The sagittal images confirmed the occipital craniectomy defect and reappearance of the CSF spaces in the foramen magnum. The velocities were slower than those measured before surgery, and quite uniform during both systole and diastole. The systolic peak velocities ranged between 0.17 and 1.80 cm/second, and the diastolic peak velocities between 0.23 and 1.40 cm/second. The mean systolic and diastolic peak velocities were 1.22 and 0.85 cm/second, respectively.

**Discussion**

A Chiari malformation with or without basilar impression may compress the brainstem and cause a variety of neurological deficits. Compression may be indirectly caused by a relatively small posterior fossa. Neurogenic arterial hypertension in association with Chiari malformation is quite uncommon, and the reason for its occurrence is still unknown.

Both the rostral and caudal portions of the VLMO are crucial for central cardiovascular regulation. The specific activation of rostral VLMO neurons causes an increase in arterial pressure that is mediated by an increase in peripheral resistance, cardiac output, and secretion of catecholamines. The caudal VLMO is a major vasodepressive area in the brainstem. Anatomical and functional data indicate that caudal VLMO neurons inhibit sympathoexcitatory neurons in the rostral VLMO and that inhibition is probably mediated by y-aminobutyric acid.

Clinical observations indicate that neurovascular compression of the left rostral VLMO may cause intractable essential hypertension. Recent reports have demonstrated a close relationship between microvascular compression, especially at the root entry zones of the left glossopharyngeal and vagus nerves, and medically refractory neurogenic arterial hypertension.1,5,8,10,13,15 Recent reports have demonstrated that microvascular compression in the rostral VLMO exists in 74 to 83% of patients with essential hypertension.13 Nonetheless, other MR imaging studies do not support the correlation between microvascular compression of the left rostral VLMO and essential hypertension.5,17–20 Recently, Levy, et al.,15 found evidence of neurovascular compression on preoperative MR images in only 59% of hypertensive patients, whereas they noted intraoperatively obvious neurovascular compression in all the patients. They concluded that current MR imaging technology is not adequate to depict the microvasculature of the brainstem. Microvascular compression was not observed in the case we presented here, although the imaging studies demonstrated dorsal brainstem compression.

Cerebrospinal fluid dynamics can be studied in detail, however. Cerebrospinal fluid flow measurements have long been a focus of interest in the evaluation of patients with Chiari malformations.2,8 In a recent study, CSF flow measurements at the foramen magnum were obtained in healthy volunteers and in patients with Chiari malformation Type I. In the healthy volunteers, the mean peak systolic velocity was 2.4 cm/second and the mean diastolic velocity was 2.8 cm/second.6 In the patients, the peak systolic flow was impaired and characterized by a nonuniform distribution and higher values than those measured in healthy volunteers. The dynamic changes in CSF in the current case support the aforementioned observations.

Investigators in several studies have reported that MVD leads to significant relief in cases of essential hypertension.14,16 Several cases of relief from neurogenic arterial hypertension after simple brainstem decompression without MVD have been reported as well. Resolution of hypertension following odontoidectomy was reported by Dickinson and colleagues6 and Barzo and associates.1 Resolution of neurogenic arterial hypertension was attributed to ventral decompression of the brainstem in these cases. On the other hand, Makhmudov, et al.,16 described a patient with a Chiari malformation whose hypertension resolved after MVD of the lower cranial nerves. Tubbs, et al.,16 recently described a 16-year-old patient with Chiari malformation Type 1 in whom chronic hypertension subsided after posterior fossa decompression.

The patient presented in this study represents the third reported case of Chiari malformation to be associated with neurogenic arterial hypertension. It is noteworthy that this is the first case in which intractable hypertension resolved in an adult patient in response to suboccipital decompression surgery alone. The resolution of arterial hypertension with no attempt to perform MVD may be attributed to indirect decompression of the root entry zones of the lower cranial nerves and the brainstem.

**Conclusions**

The evidence of effective suboccipital decompression and restoration of CSF flow after surgery may reflect decompression of the brainstem and in part the VLMO, although no MVD procedure was performed.

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