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Complete Resolution of Hypertension after Decompression of Chiari I Malformation

ABSTRACT—The Chiari I malformation has not been previously linked to sustained hypertension. Other forms of medullary compression have, however, been shown to be associated with elevated arterial pressure. This association has been demonstrated through numerous studies that have implicated compression of the rostral ventrolateral medulla, usually by vascular structures, as a factor responsible for hypertension. We present a case of a young man with a 2-year history of hypertension who was found to have a Chiari I malformation. No other cause for hypertension was found. After subsequent surgical decompression, his hypertension resolved immediately. No other therapeutic options were activated. This is the first reported case linking Chiari I malformation and sustained hypertension. This case presents further evidence linking medullary compression and elevation of hemodynamic tone.

The Chiari I malformation has been associated with a variety of symptoms. It commonly presents with pain, weakness or numbness of one or more limbs, loss of temperature sensation, and unsteadiness. Clinical signs at presentation often include those associated with foramen magnum compression syndrome, central cord syndrome, or cerebellar syndrome.¹ Although a case has been reported in which a patient with Chiari I malformation experienced central dysregulation of arterial pressure leading to paroxysmal blood pressure oscillations, chronic hypertension has not been linked in the past to this malformation.² Several studies, however, have shown an association between hypertension and vascular compression of neural structures at the level of the medulla.^{3–7} This association suggests that other

causes of medullary compression might produce blood pressure elevation through a similar mechanism. This report describes a patient with a Chiari I malformation who experienced hypertension, along with other signs and symptoms of the malformation, which resolved almost immediately after surgical decompression.

CASE REPORT

A 20-year-old man presented with a 5-year history of occipital headaches, hypertension of at least 2 years duration, obesity since the onset of puberty, and sleep apnea. His headaches were intense but brief, typically lasting about 5 minutes, and were becoming more fre-

quent and severe. They were usually associated with nausea and relieved by sleep. The patient's blood pressure had recently been monitored daily by a registered nurse for 1 week, during which time it was found to be within the range of 160 to 210 mm Hg systolic over 80 to 115 mm Hg diastolic. He was taking isradipine 10 mg twice daily (bid) and atenolol 100 mg once daily to control his hypertension. He had undergone a sleep study 18 months before presentation that showed numerous episodes of central apnea in addition to obstructive hypopnea. He was using a home continuous positive airway pressure (CPAP) unit at 15 cm H₂O, with some improvement in symptoms. He had been aware of having a nystagmus for about 8 months and described the recent onset and progression of vertigo, oscillopsia, Lhermitte's phenomenon, and dysphagia.

Examination

The patient was found to be an obese white male with a weight of 154 kg and a height of 182 cm. His blood

pressure at presentation was 153/81. Cranial nerve examination was unremarkable except for a downbeating nystagmus, which was present in primary gaze. The nystagmus was noted to increase and to develop a downward oblique configuration in lateral gaze and to dampen with convergence. The patient was also noted to have generalized hyperreflexia. The remainder of the physical examination was unremarkable. The patient had undergone an MRI examination 3 weeks before presentation, which showed a Chiari I malformation with the cerebellar tonsils located at the level of the middle of the body of C2 (Fig. 1). A syrinx was present that extended from the lower third of the body of C2 to just below the C3–C4 disc space.

Operation

Surgery was performed the day after presentation. A suboccipital craniectomy with C1–C2 laminectomy was performed. The cerebellar tonsils were found to have herniated down to the level of C3 and showed the

Figure 1. (A) T₁-weighted sagittal MRI showing Chiari I malformation with tonsillar herniation to the C2–C3 interspace. A syrinx at the C4 level is also noted. (B) T₁-weighted axial MRI at the foramen magnum, showing cerebellar tonsillar herniation.

characteristic indentation at the level of the foramen magnum. Bilateral tonsillectomies were performed to decompress the foramen of Magendie, allowing cerebrospinal fluid (CSF) to flow freely from the fourth ventricle. Adhesions were released, and a duraplasty was performed using the patient's fascia lata. There were no complications.

Postoperative Course

The patient's stay in the intensive care unit (ICU) was uneventful, and he was transferred to the ward the following day. His neurological examination was initially unchanged and he was discharged 3 days after surgery. His blood pressure was elevated 160/86 while in the ICU but was measured consistently at 140 to 145/75 to 80 when discharged to the ward. At follow-up examination 6 weeks after surgery, the patient was found to be doing extremely well. His blood pressure was 145/78 mm Hg, and he was no longer taking any blood pressure medications. He had lost considerable weight, weighing 14 kg less than at presentation. He reported that he was no longer snoring, and his nystagmus was much improved. His neurologic examination indicated no other abnormalities. Repeat MRI examination showed that the syrinx had resolved. The cerebellar tonsils were noted to be located at the level of the tip of the odontoid, with the location of the fourth ventricle within normal limits (Fig. 2). Follow-up examination at 4 years showed no return of the hypertension despite weight gain beyond his original weight. He remains off all medications.

DISCUSSION

We believe that this patient's hypertension was caused by his Chiari I malformation, not only because his blood pressure returned to normal after surgical decompression, but also because both clinical and physiologic studies have supported the hypothesis that medullary compression can lead to elevated arterial pressure, as well as other hemodynamic changes.³⁻⁹ In addition, the ventrolateral medulla has been shown to be an important modulator of vasomotor tone.¹⁰⁻¹⁷ After reviewing the course of events in this case, we believe that compression of this area was responsible for the patient's hypertension, and that surgical decompression allowed the subsequent normalization of his blood pressure.

The ventrolateral medulla can be functionally subdivided into rostral and caudal sections. The caudal ventrolateral medulla is associated with vasomotor inhibition.¹⁸ In contrast, the nucleus reticularis rostroventrolateralis (RVL), found within the rostral ventrolateral medulla, is thought to be a tonic vasomotor center, responsible for producing basal levels of sympathetic tone. This nucleus receives projections from the nucleus trac-

Figure 2. Postoperative sagittal T₁-weighted MRI. The fourth ventricle is in a more normal anatomic location, and the cerebellar tonsils are at the level of the tip of odontoid.

tus solitarius, which relay afferent cardiopulmonary information to the RVL. It also contains adrenergic neurons that terminate in the intermediolateral cell column of the spinal cord, allowing modulation of arterial pressure and other hemodynamic parameters.¹⁹ The ability of the RVL to affect vasomotor tone has been illustrated by the fact that stimulation of the rostral ventrolateral medulla electrically or with L-glutamate has been shown to raise arterial pressure, as has pulsatile compression of this area on the left side.^{16,20} These findings illustrate a plausible mechanism by which medullary compression caused by the abnormal position of the cerebellar tonsils, which are displaced inferiorly in cases of Chiari I malformation, might lead to hypertension.

Much of the research linking hypertension to medullary compression has focused on unilateral left-sided compression. This is because, through both afferent and efferent pathways, the left vagus nerve is the primary structure responsible for maintaining hemodynamic control.^{5,8,9} In this case, however, the compression was found to be symmetric. Both the preoperative MRI and intraoperative findings indicated that the level

of the cerebellar tonsils was equal bilaterally and that the compression of the medulla did not seem to affect one side more than the other. Although this does not contradict the hypothesis that left-sided compression is responsible for hemodynamic changes, we cannot exclude the possibility that right-sided compression may be responsible for hemodynamic changes, nor can we exclude the possibility that right-sided compression played a role in this case.

It could be argued that this patient's hypertension was the result of his obesity rather than medullary compression by the Chiari I malformation, and that the resolution of his hypertension was attributable to his postoperative weight loss rather than surgical decompression. It is well known that obesity predisposes to elevated arterial pressure, leading physicians to recommend weight loss as first-line therapy in the treatment of hypertension. We feel, however, that in this case the normalization of the patient's blood pressure was not the result of weight loss. This is demonstrated by the fact that at follow-up approximately 6 weeks after surgery, although his hypertension had completely resolved, the patient was still 10 kg heavier than when his elevated blood pressure was initially noted, 2 years before presentation, and at 4-year follow-up evaluation, his blood pressure was still normal despite a weight of 160 kg, 6 kg heavier than preoperatively.

The literature does not contain evidence demonstrating a link between Chiari I malformation and hypertension, indicating that this occurrence must be rare. There is, however, ample evidence linking hypertension with other forms of medullary compression.^{21,22} This case provides an interesting example of the wide variety of signs and symptoms with which Chiari I malformations may present and sheds further light on the phenomenon of hypertension associated with medullary compression.

REFERENCES

1. Paul KS, Lye RH, Strang FA, Dutton J. Arnold-Chiari malformation. *J Neurosurg* 1983;58:183-188
2. Arcaya J, Cacho J, Del Campo F, Grande J, Maillo A. Arnold-Chiari malformation associated with sleep apnea and central dysregulation of arterial pressure. *Acta Neurol Scand* 1993;88:224-226
3. Dickinson LD, Papadopoulos SM, Hoff JT. Neurogenic hypertension related to basilar impression: Case report. *J Neurosurg* 1993;79:924-928
4. Fein JM, Frishman W. Neurogenic hypertension related to vascular compression of the lateral medulla. *Neurosurgery* 1980;6:615-622
5. Jannetta PJ, Segal R, Wolfson SK. Neurogenic hypertension: Etiology and surgical treatment. I. Observation in 53 patients. *Ann Surg* 1985;201:391-398
6. Kleineberg B, Becker H, Gaab MR. Neurovascular compression and essential hypertension. *Neuroradiology* 1991;33:2-8
7. Naraghi R, Gaab MR, Walter GF, Kleineberg B. Arterial hypertension and neurovascular compression at the ventrolateral medulla. *J Neurosurg* 1992;77:103-112
8. Jannetta PJ, Segal R, Wolfson SK, Dujovny M, Semba A, Cook EE. Neurogenic hypertension: Etiology and surgical treatment. II. Observations in an experimental nonhuman primate model. *Ann Surg* 1985;202:253-261
9. Segal R, Gendell HM, Canfield D, Dujovny M, Jannetta PJ. Hemodynamic changes induced by pulsatile compression of the ventrolateral medulla. *Angiology* 1982;33:161-172
10. Blessing WW, Reis DJ. Inhibitory cardiovascular function of neurons in the caudal ventrolateral medulla of the rabbit: Relationship to the area containing A1 nonadrenergic cells. *Brain Res* 1982;253:161-171
11. Caverson MM, Ciriello J, Calaresu FR. Direct pathway from cardiovascular neurons in the ventrolateral medulla to the region of the intermediolateral nucleus of the upper thoracic cord: An anatomical and electrophysiological investigation in the cat. *J Auton Nerv Syst* 1983;9:451-475
12. Ciriello J, Caverson MM. Bidirectional cardiovascular connections between ventrolateral medulla and nucleus of solitary tract. *Brain Res* 1986;367:273-281
13. Ciriello J, Caverson MM, Polosa C. Function of the ventrolateral medulla in the control of the circulation. *Brain Res Rev* 1986;11:359-391
14. Dampney RAL, Goodchild AK, Robertson LG, Montgomery W. Role of ventrolateral medulla in vasomotor regulation: A correlative anatomical and physiological study. *Brain Res* 1982;249:223-235
15. Dormer KJ, Bedford TG. Cardiovascular control by the rostral ventrolateral medulla in the conscious dog. *Prog Brain Res* 1989;81:265-277
16. Ross CA, Ruggiero DA, Park DH, Joh TH, Sved AF, Fernandez-Pardal J, Saavedra JM, Reis DJ. Tonic vasomotor control by the rostral ventrolateral medulla: effect of electrical or chemical stimulation of the area containing C1 adrenaline neurons on arterial pressure, heart rate, and plasma catecholamines and vasopressin. *J Neurosci* 1984;4:474-494
17. Ross CA, Ruggiero DA, Reis DJ. Projections from the nucleus tractus solitarii to the rostral ventrolateral medulla. *J Comp Neurol* 1985;242:511-534
18. Blessing WW, Li Y-W. Inhibitory vasomotor neurons in the caudal ventrolateral region of the medulla oblongata. *Prog Brain Res* 1989;81:83-97
19. Ruggiero DA, Cravo SI, Arango V, Reis D. Central control of the circulation by the rostral ventrolateral reticular nucleus: Anatomical substrates. *Prog Brain Res* 1989;81:49-79
20. Reis JR, Granata AR, Tong HJ, Ross CA, Ruggiero DA, Park DH. Brain stem catecholamine mechanisms in tonic and reflex control of blood pressure. *Hypertension* 1984;6(suppl II):7-15
21. Levy EI, Clyde B, McLaughlin MR, et al. Microvascular compression of the left medulla oblongata for severe refractory neurogenic hypertension. *Neurosurgery* 1998;43:1-9
22. Segal R. Microvascular decompression of the left lateral medulla oblongata for severe refractory neurogenic hypertension. *Neurosurgery* 1999;44:232-233