Received: 17.8.2010 **Accepted: 26.10.2010**

Case Report

Is Chiari malformation a cause of systemic hypertension and sinus bradycardia?

A case report and literature review

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Abstract

A middle aged woman, having a history of diastolic hypertension and sinus bradycardia since one year ago, was referred to our center with a sudden occipital headache after shouting. To evaluate the cause of headache the brain MRI was performed reporting a slight cerebellar tonsillar herniation of about one centimeter below the foramen magnum. After the patient was diagnosed to have type I Chiari malformation, a surgery procedure was done and the symptoms were recovered after that.

Type I Chiari malformation is a disease mostly caused by congenital displacement of cerebellar tonsils through the foramen magnum. The most common symptom is headache, rarely reported with hypertension or sinus bradycardia.

KEYWORDS: Arnold-Chiari Malformation, Bradycardia, Hypertension, Headache Disorders.

JRMS 2011; 16(1): 115-118

Type I Chiari malformation (CMI), is a disease with different congenital or acquired etiologies that characterized by caudal cerebellar tonsillar herniation through the foramen magnum into the cervical canal. This disease involves both sexes with the male to female ratio of approximately 2:3 and its prevalence is unknown.[1-3] The age of patients varies from 10 months to 65 years with an average of approximately 35 years old.

The most common symptom is headache with a range from short "cough induced" to continues one which seems to be related to the compression of neural and/or dural structures by the herniated tonsils.[4-6] Moreover, the size of tonsillar herniation cannot predict the severity and type of headaches. However, there is not a pathogonomic type of headache for CMI, severe paroxysmal, occipital and/or posterior cervical pain associated with Valsalva maneuver named "cough headache" is more seen in these patients.[4,7] Other rare presentations of CMI are sinus bradycardia [8] and hypertension.[9-11]

In this case study, we represented a woman with a resistant hypertension, sinus bradycardia, and some neurological symptoms that relieved by decompressive surgery.

Case Report

History

A forty year old woman was referred to our center with a sudden paroxysmal headache after shouting. The history of occasional classic migraine-typed headache with visual aura responding to common analgesic drugs of at least two years duration was found, although she did not have such shouting-induced headache before that. Moreover, there had been a history of diastolic hypertension which was resistant to 25 mg of captopril three times a day since one year ago. The blood pressure was self-monitored at home ranging from 90 to110 mmHg in diastolic ones. Also, she complained frequent sinus bradycardia in the 35-50

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beats per minute range with unknown source, any relation to medications, and concurrent with onset of hypertension which sometimes was treated with intravenous atropine. In past medical history, elective cesarean section and hernioplasty were dominant. She did not have any history of sleep apnea or any signs and symptoms related to increased ICP.

Examination

The general physical examinations were normal except for pulse rate of 55 beats per minute and diastolic hypertension of 95 mmHg. In neurologic examination, there was no evidence of meningeal irritation, and cranial nerves, mental status, sensory and motor system functions were normal. Also, deep tendon reflexes were mildly exaggerated (+ 3) in all limbs. The brain MRI was done to rule out the structural causes of cough-induced headaches. According to MR imaging, Chiari I malformation was discovered in the patient (Figure 1).

Surgery Procedure

The skin was incised from inion to C7 spinous process with a midline incision. The occiput, C1 and C2 laminas, and foramen magnum were explored, and then craniectomy and C1 laminectomy was done. After dura exploration and adhesion dehiscent, thick tonsillar bands were burned with bipolar and dura was expanded with femoral muscle fascia. (Figure 2)

Post Surgery Follow-Up

In the six-month follow up period, the patient did not have any neurological symptoms such as headache. She had no more hypertension and sinus bradycardia which was measured by herself or a physician. Follow up MRI was presented in figure 2.

Discussion

In our view, the hypertension and sinus bradycardia of this patient could be related to CMI, because not only these presentations were unresponsive to medications, but also were resolved after the surgery. In addition, there were three case reports showing resolution of hypertension in CMI patients after the decompressive surgeries, though these cases had no sinus bradycardia.[9,11] Selmi et al represented a case with profound sinus bradycardia and CMI, without hypertension.[8] Now, we can claim that this case is the first having both sinus bradycardia and diastolic hypertension with CMI malformation.

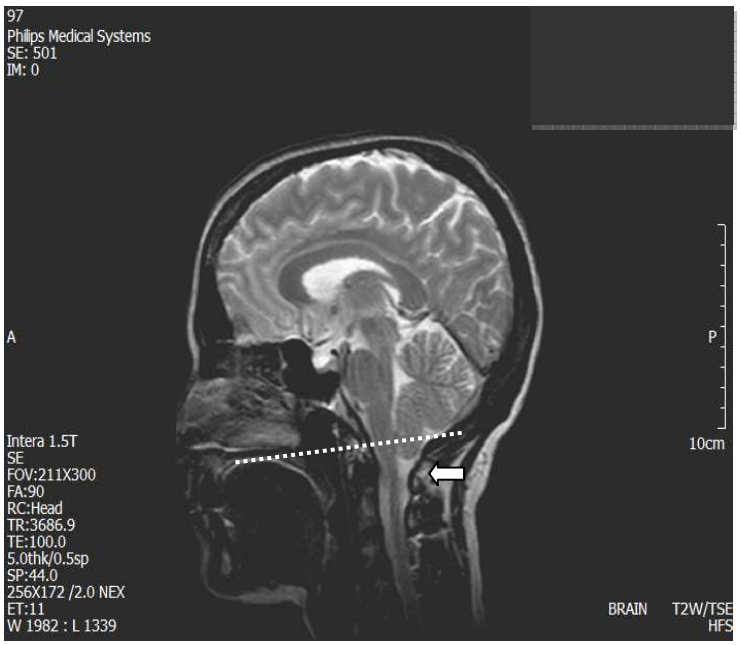

Figure 1. (Before the surgery) Midsagittal T_2-weighted MRI shows tonsillar herniation of about one centimeter (the arrow) below the hard palate-foramen magnum line (the line). Also, no Syrinx and no hydrocephaly or deformity of 4th ventricle is noticed.

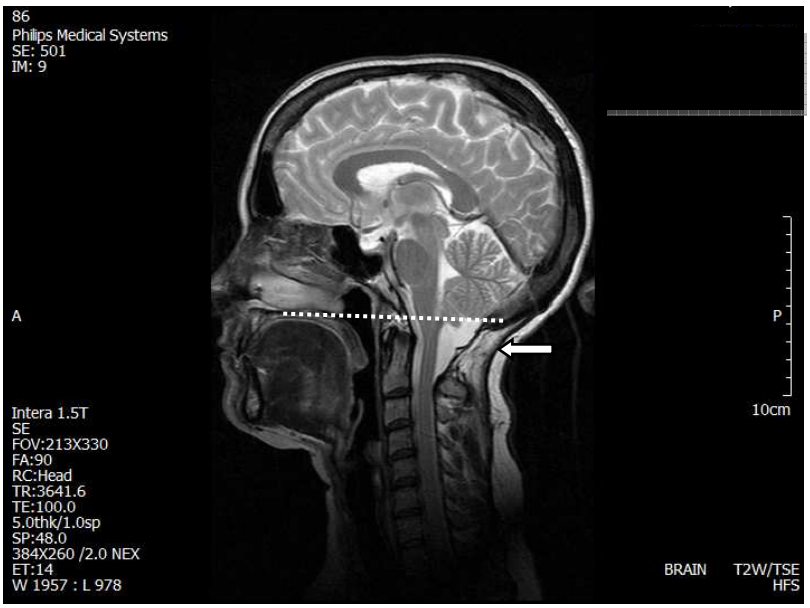

Figure 2. (After the surgery) The occipital craniectomy scar (the arrow) and tonsillar upward migration above hard plate-foramen magnum line (the line), with dilation of foramen magnum and reduction of stenosis were seen. No syrinx, stenosis, and compression were found in the MRI study after the surgery.

Though the exact mechanism of hypertension in CMI patients is unknown, some hypotheses exist such as activation of rostral ventrolateral medulla oblongata (VLMO) neurons by compression which causes an increase in peripheral vascular resistance, cardiac output, and secretion of catecholamines.[11-13] The caudal VLMO is a nucleus working against the vasomotor activity, so it has anti hypertensive effects.[14] Another tonic vasomotor center within the rostral VLMO is the reticularis rostroventrolateralis (RVL) nucleus being responsible for basal levels of sympathetic tone.[15] Kleineberg et al showed the neurovascular compression of the rostral VLMO in MRI and MR angiography of patients with essential hypertension,[16] however this study's finding was not confirmed by others.[17,18] By reviewing the course of events in this case, it could be concluded that the compression of this area was responsible for the patient's hypertension, since the surgical decompression led to subsequent normalization of blood pressure.

CSF flow is another explanation for hypertension in the patient [19] but there was not any suitable equipment measuring CSF flow from the foramen magnum in our center.

We proposed that compression of rostral port of VLMO and subsequently tension of dorsal nucleus of the vagus and the ambiguous nucleus is responsible for sinus bradycardia in absent of increased ICP because the rostral ventrolateral medulla is a regulatory center for cardiac, vascular, and endocrinologic control of blood.[13-20]

Conclusions

Chiari I malformation should be considered in differential diagnosis of patients with resistant hypertension, sinus bradycardia, and sudden cough-induced headache; therefore, ruling this diagnosis out by imaging techniques such as MRI seems to be essential. Finally, clinical presentations of CMI like hypertension and sinus bradycardia can be resolved by decompressive surgery.

Acknowledgements

All authors are grateful to Dr. Marashi, radiology specialist, who helped us in MRI findings.

Conflict of Interests
Authors have no conflict of interests.

Authors' Contributions
MG designed the study and gathered information about the patients before and after the surgery and wrote the case report part except the surgery procedure. KG provided assistance in the design of the study and also wrote the introduction, the discussion and the conclusion. VS coordinated between authors of this study and reviewed related articles. MR, the neurosurgeon of the patient, wrote the surgery part of the report. All authors have read and approved the content of the manuscript.

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