Clinical Case Report

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Combined hyperactive dysfunction syndrome of the cranial nerves complicated by essential hypertension

A case report

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Abstract

Rationale: Combined hyperactive dysfunction syndrome (HDS) refers to a special type of HDS characterized by a combination of trigeminal neuralgia (TN), hemi facial spasm (HFS), and/or gloss pharyngeal neuralgia (GPN). Rostra ventrolateral medulla (RVLM) plays a crucial role in central cardiovascular regulation, and neurovascular compression of the RVLM has been identified as a contributor to essential hypertension.

Patient concerns: A 65-year-old female with a facial tic and pain located in the root of the tongue and throat on the same side; the systolic and diastolic blood pressure was approximately 170 and 100 mmHg.

Diagnosis: The patient was diagnosed with combined HDS (HFS-GPN) and essential hypertension. Brain magnetic resonance 3-dimensional time-of-flight imaging and digital subtraction angiography revealed vertebrobasilar artery compressed the left RVLM and contacted with the root entry zones of multiple cranial nerves.

Interventions: The patient was treated with microvascular decompression surgery

Outcomes: The symptoms were completely relieved, and blood pressure was well-controlled.

Lessons: The pathological association of hypertension and HDS should be highlighted, and microvascular decompression is an effective approach for relieving the hypertension.

Abbreviations: GPN = gloss pharyngeal neuralgia, HDS = combined hyperactive dysfunction syndrome, HFS = hemi facial spasm, RVLM = rostra ventrolateral medulla, TN = trigeminal neuralgia.

Keywords: combined hyperactive dysfunction syndrome, essential hypertension, microvascular decompression, rostra ventrolateral medulla

1. Introduction

Hyperactive dysfunction syndrome (HDS) refers to a clinic pathological condition characterized by overactivity in the cranial nerves, including trigeminal neuralgia, hemi facial spasm (HFS), and gloss pharyngeal neuralgia (GPN). HDS is caused by arterial cross-compression at the root entry/exit zone (REZ) of the cranial nerves. Previous studies indicated that a combination of these symptoms, which is also known as combined HDS, is extremely rare. It is noticed that patients with combined HDS exhibited an increased incidence of essential hypertension comparing with patients with a single HDS.[1]

Rostra ventrolateral medulla (RVLM) plays a crucial role in central cardiovascular regulation, and neurovascular compression of the RVLM has been identified as a contributor to essential hypertension. The microvascular decompression of the RVLM has been shown to be effective for treating relevant hypertension.[2] The pathological contacts of vessels and the root entry or exit zone of cranial nerves close to the brain stem may be the common cause of combined HDS and essential hypertension.

In the present study, we report a case with concomitant combined HDS of the cranial nerves and essential hypertension, which was successfully treated with microvascular decompression surgery. Patient has provided informed consent for publication of this case.

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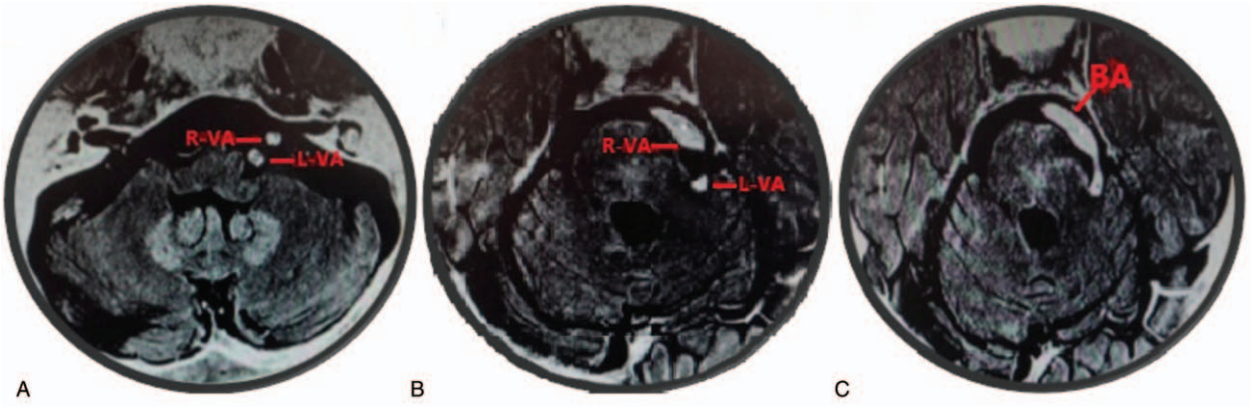

Figure 1. Magnetic resonance 3D-TOF imaging revealed that the right tortuous vertebral artery (R-VA) deviated to the left side, and the left vertebral artery (L-VA) compressed the RVLM (A and B). The basilar artery (BA) contacted with the root entry zones of multiple cranial nerves (A–C).

2. Case report

A 65-year-old female with a crowded posterior fossa presented with a 5-year history of left HFS. No specific treatment was performed. Three years after onset, the patient developed GPN and hypertension. HFS occurred first in this case, and then GPN occurred on the same side for this patient with combined HDS (HFS-GPN). Oral carbamazepine (800 mg/day), irbesartan and nifedipine were prescribed. The pain was partially relieved, and the systolic and diastolic blood pressures were controlled at approximately 170 and 100 mmHg, respectively. Chest X-ray revealed cardiomegaly. Doppler echocardiography revealed left ventricle hypertrophy with decreased diastolic function, while blood flow and pulmonary artery pressure remained within normal ranges. The electrocardiogram and laboratory examinations on admission were all normal. Brain magnetic resonance three-dimensional time-of-flight (3D-TOF) imaging (Fig. 1) and digital subtraction angiography (Fig. 2) demonstrated that the right tortuous vertebral artery (R-VA) deviated to the left side, and the left vertebral artery (L-VA) compressed the RVLM and the basilar artery contacted with the root entry zones of multiple cranial nerves.

Microvascular decompression was performed via the superior portion of the cerebellopontine angle cistern. Intraoperatively, it was found that the vertebrobasilar artery compressed the root entry zones of multiple cranial nerves and the RVLM in a crowded posterior fossa (Fig. 3). The offending vessels were suspended with bioadhesive paste. Thus, the compression of the vertebrobasilar artery on multiple cranial nerves and the RVLM were relieved.

The postoperative course was uneventful. Facial tic and pain located in the root of the tongue and throat completely disappeared at 12 hours after the surgery. At postoperative one month, the systolic and diastolic blood pressures were stabilized at 140 and 90 mmHg, respectively. During a follow-up period of 12 months, no clinical recurrence was noted.

3. Discussion

Jannetta was the first to propose the neurovascular compression of the ventrolateral medulla at the exit zone of cranial nerves could lower blood pressure which suggested the association between hypertension and HDS.[3] In a previous study involving 201 patients affected with hemifacial spasm who underwent MVD, 23.8% of the patients had hypertension, and after MVD, BP had returned to normal in 28 patients; 14 of these patients were able to maintain normal BP without any antihypertensive treatment.[4] Li retrospectively analyzed published studies and proposed that MVD may be an effective approach for patients with severe, unstable, and refractory hypertension.[5] In addition, relevant meta-analyses have confirmed the correlation between hypertension and HDS.[6] Legrady retrospectively investigated 13 resistant hypertensive patients, in which systolic and diastolic blood pressures and pulse pressure decreased in all patients following the decompression, and these changes were more remarkable in patients with multiple compressions.[7]

This case is a 65-year-old female with a crowded posterior fossa, who was diagnosed with combined HDS of the cranial nerves and concomitant hypertension. As presented in the previous literature, combined HDS shows a tendency to occur in older female patients and it is associated with an increased incidence of hypertension.[8] The aging process could accelerate arteriosclerotic changes as well as hypertension and facilitate the development of arterial elongation and tortuosity, which would increase the risk of combined HDS due to neurovascular compression. In Chinese population, the posterior fossa in females is more crowded than that in males.[1] This anatomic feature may contribute to the crowdedness between cranial nerves and vascular structures, which increases the risk of neurovascular compression.[9] Therefore, we speculated that hypertension may be a cause for, rather than a result of, combined HDS.

Previous studies showed that neurovascular pulsatile compression at the RVLM is a possible cause for essential hypertension.[10] Hypertension is characterized by sympathetic overactivity, which is associated with an enhancement in angiotensin receptor type I (AT1R) in the rostral ventrolateral medulla (RVLM).[11] Aorta found that plasma norepinephrine levels were significantly higher in the neurovascular compression group than in the non-neurovascular compression group.[12] Norepinephrine could enhance angiotensin II responses.[13] Furthermore, an animal experiment indicated that Angiotensin II may activate RVLM neurons via AT1R, and stimulate glutamic acid release in the context of pulsatile compression on the RVLM, thereby eliciting sympathetic activation and pressor effects.[14]

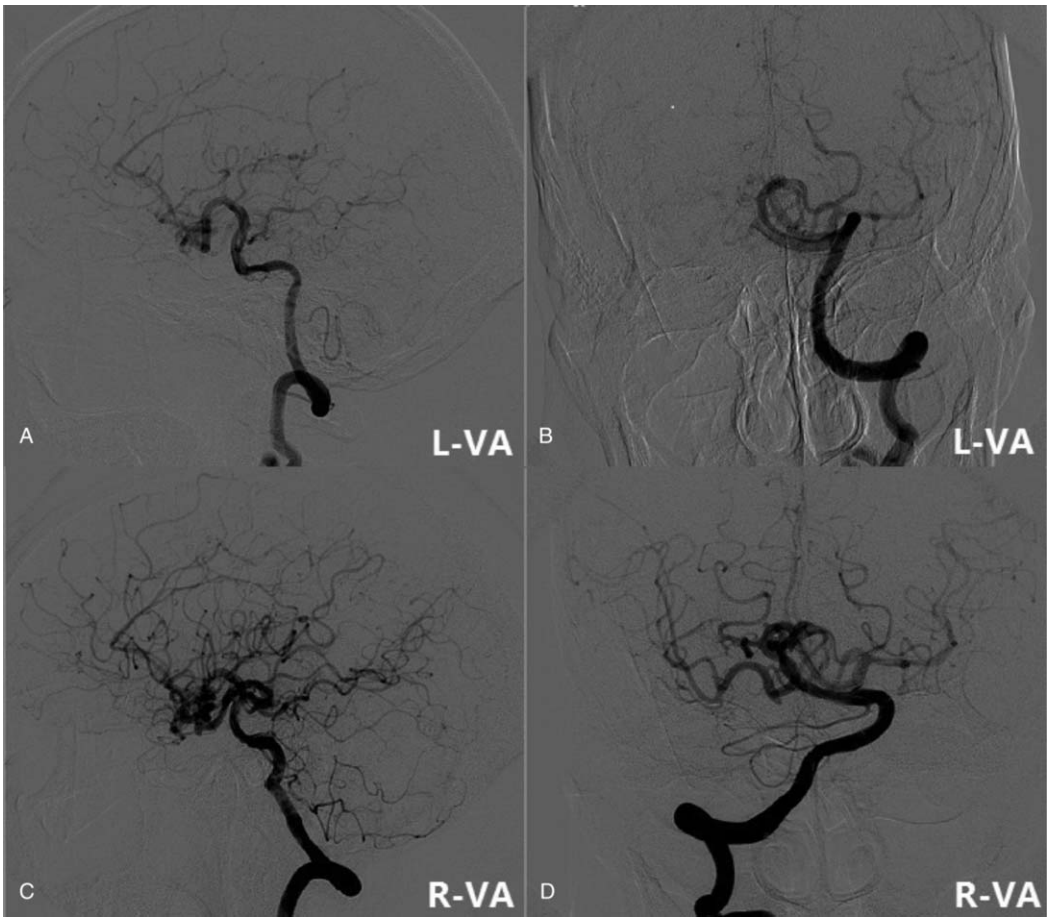

Figure 2. Digital subtraction angiography demonstrated that the left vertebral artery (L-VA) compressed the RVLM (A and B). The right tortuous vertebral artery (R-VA) and basilar artery extended to the suprasellar cisterna level (C and D).

The C1 cells are glutamatergic neurons located in the rostral ventrolateral medulla (RVLM) as critical for respiratory-sympathetic entrainment and the development of experimental hypertension.[15] Long-term repeated neuropathic pains or convulsions of HDS enhanced tonically active glutamatergic input to the rostral ventrolateral medulla (RVLM) which contributes to high level of blood pressure or even refractory hypertension.[16] In the present case, we found that the right tortuous vertebral artery (R-VA) deviated to the left side, and the left vertebral artery (L-VA) compressed the RVLM and the basilar artery contacted with the root entry zones of multiple cranial nerves. The hypertension was successfully treated with

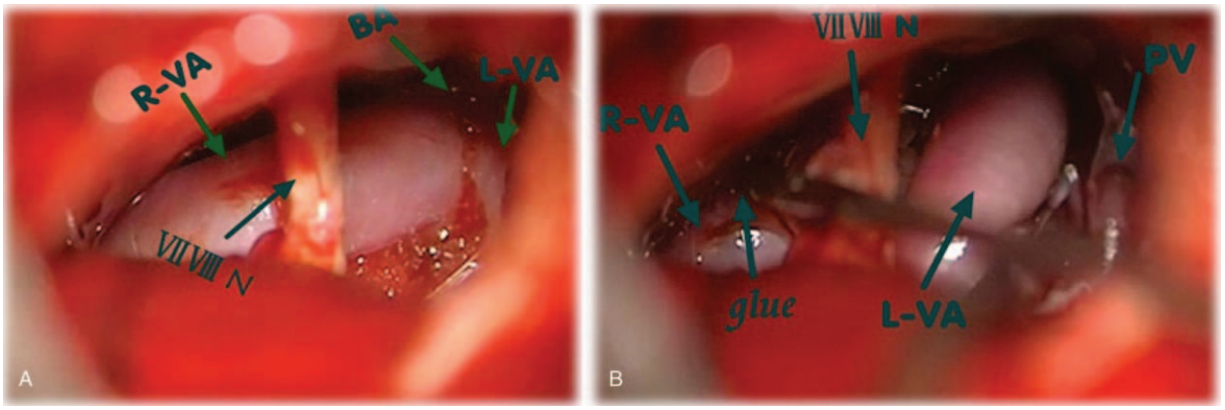

Figure 3. Intraoperatively, it was found that the right vertebral artery (R-VA) deviated to the left side, and the left vertebral artery (L-VA) compressed the RVLM in a crowded posterior fossa (A). The offending vessels were suspended with bioadhesive paste, and the compression of multiple cranial nerves and the RVLM was completely relieved (B).

microvascular decompression, suggesting a pathological association between hypertension and HDS.

It showed that the hypertension ratio of patients with combined HDS was significantly higher than those patients with single HDS.[9] Another important issue is whether the severity of hypertension in patients with combined HDS is similar to that in patients with single HDS. First, the clinical manifestations of combined HDS, such as spasm and neuralgia, can increase sympathetic nervous system activity, promoting the release of norepinephrine and repinephrine. Second, the pathological mechanism of combined HDS is more complicated and serious, such as abnormal elongation and dilatation of the vertebrobasilar artery and the over-release of vascular active substances induced by the neurovascular compression of the RVLM. The definitive difference between combined HDS-complicated hypertension and single HDS-complicated hypertension still needs further clinical and experimental studies.

4. Conclusion

Age, gender, and hypertension might be the important causative factors for combined HDS. Neurovascular compression at the root entry zones of multiple cranial nerves and the RVLM is a possible cause for essential hypertension. Posterior fossa crowdedness and arterial tortuosity might increase the severity of neurovascular compression. Microvascular decompression is an effective approach for patients with coincidence of combined HDS of the cranial nerves and essential hypertension. Considering combined HDS tends to occur in elderly patients, surgeons should pay more attention to certain potential postoperative complications.

Author contributions

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