Clinical Report

Improvement of Severe Retrocochlear Hearing Loss and Vestibular Disorders Caused by Ruptured Brainstem Cavernous Angioma

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We report a case of hemorrhage from brainstem cavernous angioma that caused various symptoms including neurootological, gustatory and facial sensory disturbances. A man experienced nausea and numbness of his right ear. It was confirmed that the symptom were derived not from a peripheral lesion but from retrocochlear and brainstem impairment based upon the findings of the DPOAE, ABR, VEMP, caloric test, optokinetic nystagmus etc. We detected an area of retrocochlear lesion that was responsible for both the auditory and vestibular disturbances based upon detailed neurootological examination in addition to MRI findings, and showed that the impairment was improved one year later without specific treatment.

Key words: hearing loss, dysgeusias, facial palsy, intracranial hemorrhage, auditory brainstem response (J. Nihon Univ. Med. Ass., 2010; 69 (6): 361–366)

Introduction

We experienced a case of hemorrhage from cavernous angioma which revealed various brainstem symptoms about neurootological, gustatory and facial disturbances. Especially, in the early stage of a brainstem hemorrhage, it is usually difficult to evaluate auditory function because those patients commonly are in critical life condition at that time. Also, it is uncommon that a patient who suffered retrocochlear hearing loss shows severe deafness on its pure tone audiogram examination as in this case because the auditory nerve runs bilaterally in brainstem after olivary nucleus. We followed up the case for over one year and reported the improvement of symptoms.

Case report

A 40-year-old male developed right tinnitus so he visited a nearby clinic the next day, (day 1). He was diagnosed as having a cold. On day seven the patient was experiencing nausea and numbness of his right ear so he called an ambulance in which he was then transported to a neighborhood hospital. A brain computed tomography (CT) scan revealed a high-density area from the right dorsal brainstem to the right pedunculus cerebellaris, he was immediately admitted to the hospital. Magnetic resonance imaging (MRI) was performed at the hospital and arteriovenous malformation at the brainstem was suspected. He was transferred to the Neurosurgery Department of our hospital on day 16 for observation and treatment. Both his past medical history and that of his family’s were unremarkable.

On day 17 of the admission, the patient was seen by an otolaryngologist. The patient’s eardrums were normal but pure tone audiometry revealed that the right ear had little hearing (Fig. 1a), and distortion product oto-acoustic emissions (DPOAEs) showed good re-

Fig. 1a Pure tone audiometry on day 17.

The left ear was normal and the right ear showed almost deafness.
response in both ears without laterality (Fig. 2). On the same day, the stapedius reflex test showed defects in the ipsilateral response on the affected side and the contralateral response on the healthy side.

Self-recording audiometry on day 21 revealed that he was unable to hear continuous sound and discrimination of speech sound was markedly impaired (Fig. 3). An auditory brainstem response (ABR) demonstrated that after 100 dB stimulation of the right ear, the I wave was recognized but the II and later waves were not (Fig. 4).

Facial palsy was absent, but the sense of pain, touch, and cold was impaired on the right side of the face, in particular, from the right upper eyelid to the right corner of the mouth. Electrogustometry and the filter-disc method revealed that the patient’s sense of taste was impaired in the regions on the right side covered by the tympanic nerve, glossopharyngeal nerve, and major petrosal nerve, but was normal on the left side.

The patient did not have vertigo, but he developed horizontal rotary spontaneous nystagmus directed to the left side with a small amplitude and low frequency on day 17. The optokinetic nystagmus (OKN) test on day 21 demonstrated that the maximum velocity in the slow phase was reduced to 42 degrees/sec on the right side and 43 degrees/sec on the left side (normal range, about 70–90 degrees/sec), and the nystagmus induction rate was markedly reduced. A caloric test with a cold wind at 12 degrees centigrade showed a good re-
The visual suppression test showed 56% on the right and 60% on the left. Vestibular evoked myogenic potential (VEMP) revealed a good response on both sides upon stimulation at 105 dB. The stepping test revealed remarkable unsteadiness to the right and left with his eyes closed, but there was no specific tendency of deviation. Cerebellar symptoms, other cranial nerve symptoms, and the Horner sign were not observed.

A brain CT on day 17 showed a high-density area at the dorsal to lateral side of the right medulla to the right pedunculus cerebellaris. An MRI on day 22 revealed an angioma with high-intensity signals at the center and low-intensity signals at the surrounding rim on the T2-weighted image, suggestive of macrophages incorporating hemosiderin, which was a typical image of a hemorrhage from a cavernous angioma (Fig. 5). Cerebral angiography on day 20 demonstrated no images of afferent vessels, nidus, or staining, and right vertebral arteriography suggested arteriovenous malformation in the arterial phase. In contrast, a number of fine veins were aggregated at the right pedunculus cerebellaris in the venous phase, and had merged to the superior petrosal sinus as the efferent vein, which suggested the presence of venous angioma (Fig. 6).

**Fig. 3** Speech discrimination test on day 21.
Speech discrimination of the right ear was markedly reduced.

**Fig. 4a** ABR of the right ear on day 22.
Upon stimulation at 100 dB, the I wave was recognized but the II and later waves were lost.

**Fig. 4b** Right ARB one year after onset.
The II and later waves appeared and the latency of the I–V waves was prolonged to 6.3 ms.
Fig. 5  Brain MRI on day.
T2-weighted image shows a hematoma with high-intensity signals at the core and low-intensity signals at the surrounding rim.

Fig. 6  Vertebral arteriography on day 20.
There were no abnormal findings in the arterial phase but aggregated vascular images were observed at the right pedunculus cerebellaris in the venous phase.

Clinical course

According to the findings of the MRI and cerebral angiography, the patient was diagnosed with concurrent brainstem cavernous angioma and venous angioma. After a thorough discussion with the patient and his family, conservative therapy and rehabilitation was chosen over surgical treatment. Hearing in the right ear was 65 dB at the frequency of (500 Hz + 1000 Hz + 2000 Hz)/3 (dB) on day 21 (Fig. 1b), which improved to 20 dB at the same frequency on day 70 (Fig. 1c). However, right tinnitus persisted. The speech discrimination test carried out four months after onset revealed improvement in the maximum speech articulation of the right ear to 95% (45 dB).

Sensory impairment of the face had almost disappeared by day 70 and the gustatory sense improved in the regions covered by the tympanic nerve, glossopteryngeal nerve, and major petrosal nerve by day 61. With regard to balance, spontaneous nystagmus disappeared by day 51, but the stepping test revealed lopsidedness to the right. Unsteadiness to the right and left with his eyes closed disappeared by day 70.

An MRI taken 10 months after onset showed hypointensity signals on T1- and T2-weighted images. A scar of an old hemorrhage was observed but there was no evidence of any recurring bleeding or edema.

Pure tone audiometry one year after onset demonstrated that the hearing threshold of the right ear showed improvement by about 10 dB at 1–8 kHz, compared with the left side. On the same day, ABR was examined by stimulation of the right ear at 100 dB. The II and later waves, which were lost on day 22, appeared and the latency of I–V waves was prolonged to 6.3 ms (Fig. 4b).

The OKN test revealed an improvement in the rate of nystagmus induction on both sides, and the maximum velocity in the slow phase was 61 degrees/sec on the right and 65 degrees/sec on the left. In terms of subjective auditory symptoms, right tinnitus disappeared and only a mild sense of the right ear being blocked persisted.

Discussion

There are four pathological types of cerebral vessel malformation; a) cerebral arteriovenous malformation (AVM); b) cavernous malformation (CM); c) capillary telangiectasia; and d) venous malformation2. It has been reported that brain CM occurs in 0.4–0.9% of the population and accounts for 8–15% of all vascular malformations3–7. Of these, brainstem cavernous malformation (BCM) accounts for about 20%8. Intracranial hemorrhaging occurs in about 0.25–1.1% of all CMs annually3, 7, 9, and in about 5% of all BCMs2. BCM often appears with hemorrhaging10, and once it bleeds, re-bleeding is likely11–13, and previous reports showed a recurrent bleeding rate of 20–30%6, 8. CM often accompanies venous malformation such as venous angioma, and venous malformation occurs concurrently in 8–16% of BCMs8, 14, 15.

Porter, et al. reported that among 100 patients with BCM, their clinical symptoms consisted of headache (36%), dizziness (24%), nausea and vomiting (18%), and trigeminal neuralgia (4%), and their neurological findings included cranial nerve symptoms (69%), sensory disorder (39%), motor disorder (38%), cerebellar ataxia (30%), articulation disorder (12%), and no symptom (3%)9. It is widely known that not only CM but also aneurysm at the distal area of the anterior inferior cerebellar artery is a cause of brainstem hemorrhaging. In cases of mild hemorrhaging symptoms at
onset may include perception deafness and rotatory vertigo\(^6\). MRI is the most effective imaging modality for the diagnosis of CM. As CM causes repeated hemorrhages, MRI findings characteristically change from a hematoma pattern to a mosaic one. Zabramski, et al. categorized MRI findings of CM into four types: Type 1, T1: hyperintensity core, T2: hyper- or hypo-intensity with a surrounding hypointensity rim; Type 2, T1: reticulated mixed signal core, T2: reticulated mixed signal core with a surrounding hypointensity rim; Type 3, T1: iso- or hypo-intensity, T2: hypointensity with a hypointensity rim that magnifies the size of the lesion; and Type 4, T1: poorly visualized or not visualized at all, T2: poorly visualized or not visualized at all\(^7\).

The present case exhibited high intensity on the T1-weighted image and a mixture of low and high intensity at the core and low intensity at the surrounding rim on the T2-weighted image, which corresponded to the findings of Type 1.

Due to the risk of recurrent bleeding, it is desirable to resect BCMs\(^{15,18,19}\), but conservative therapy is expected for venous angioma as long as it is asymptomatic because it is involved in the normal venous circulation\(^{12,20,21}\). Radiosurgery may be selected according to the risk of the surgery and the patient’s condition. The efficacy of radiosurgery for BCM has been proposed and some reports showed a significant reduction in recurrent bleeding rates\(^{13,23}\), while it was reported that mortality was as high as about 3% because of radiation injury in cases where the CM is located at the brainstem and deep area\(^{24}\). With Consideration to the age of the patient at presentation, the risk of symptom aggravation of recurrent bleeding and the necessity of surgery were fully explained to the patient and conservative follow-up was selected. A follow-up MRI confirmed absorption of the hematoma, and no sign of recurrence has been observed for one year.

Neurological findings observed in this case are summarized as follows.

Auditory disturbance: DPOAE to examine cochlear function was normal. Since the I wave was present but the II and later waves were lost on ABR, a marked elevation in the hearing threshold of the right ear on pure tone audiometry suggested retrocochlear hearing impairment. Since MRI demonstrated that a hematoma had formed at the site where the cochlear nerve entered the brainstem, it was suspected that there was impairment at the site where the cochlear nerve entered the brainstem and in the acoustic conduction system at the lower brainstem where the acoustic nerve ran from the cochlear nucleus to the superior olivary nucleus.

Balance disturbance: Horizontal rotary nystagmus directed to the left which was observed at the first visit to our department (day 17) is not contradictory to the findings of right brainstem injury, and it was thought that poor OKN induction was attributable to brainstem injury by edema and hematoma. The Caloric test and VEMP, which is the vestibulo-collic reflex arc consisting of saccule—inferior vestibular nerve—vestibular nucleus—internal vestibulospinal tract—accessory nerve spinal tract—sternocleidomastoid muscle, showed a good response on both sides, suggesting that there was no injury to the peripheral vestibular nerve.
Good results on the visual suppression test implied that there was no injury from the paramedian zone of the pons to one hemisphere of the cerebellum.

Gustatory and facial sensory impairment: Based on the location of the hematoma detected by MRI, gustatory impairment was attributable to injury to the nucleus of the solitary tract, while facial sensory impairment seemed to be caused by injury to the trigeminal nucleus.

Taken together, the presumed injury sites at the brainstem in the present case are shown in Fig. 7. Hemorrhage from cavernous angioma injured the brainstem with a hematoma or compressed the brainstem with edema. It was suspected that the hemorrhage injured the region from the cochlear nucleus to the corpus trapezoideum and their adjacent nucleus of the solitary tract, trigeminal nucleus, and inferior pedunculus cerebellaris at the same time.

Conclusion

According to detailed neurootological examination’s evidence in addition to MRI findings, we detected the area of retrocochlear lesion about both of auditory and vestibular disturbance. Retrocochlear hearing loss caused by brainstem hemorrhage showed severe deafness in the early stage, but the patient completely recovered after one year. Vestibular, gustatory and facial sensory disturbance recovered as well. Therefore, we supposed that the symptom were occurred because of the brainstem compression of hematoma caused by hemorrhage and they naturally recovered due to the diminishing of hematoma.

REFERENCES