Neurovascular compression syndrome of the brain stem with opsoclonus-myoclonus syndrome combined with vestibular paroxysmia and autonomic symptoms

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Summary

We describe a rare case of neurovascular compression syndrome (NVCS) of the brain stem and opsoclonus-myoclonus syndrome (OMS) complicated with vestibular paroxysmia (VP) and autonomic symptoms. Moreover, we discuss the case with respect to the available information in medical literature. A 36-year-old man with vertigo and nausea, and difficulty standing, was transported by an ambulance to our hospital. He had VP, opsoclonus, cervical myoclonus, anxiety, and restless legs syndrome. Magnetic resonance imaging showed that the dolichoectatic vertebral artery was in contact with the postero-lateral side of the pontomedullary junction. He was diagnosed with NVCS of the brain stem (most likely of the input to the vestibular nucleus) associated with contact with the dolichoectatic vertebral artery. Combination therapy using multiple antiepileptic drugs, such as low-dose carbamazepine, clonazepam, and lacosamide, improved his clinical symptoms. He was finally able to walk and was discharged on day 42 after admission. He is being routinely followed-up since then. Further research is needed to confirm the validity of the combination therapy.

Keywords: Neurovascular compression syndrome, brainstem, opsoclonus myoclonus syndrome, vestibular paroxysmia, autonomic symptoms

1. Introduction

Opsoclonus-myoclonus syndrome (OMS) (1-3) is a rare disease with opsoclonus, cerebellar ataxia, and myoclonus of the trunk, limbs, and neck as the primary symptoms. The probable underlying etiologies eliciting the symptoms include viral encephalitis, malignant tumors, metabolic disorders, and degenerative diseases. Particularly, OMS as paraneoplastic nerve syndrome (PNS), wherein, various neurological symptoms manifest because of the "remote effect" of malignant tumors, is an important etiology. Neuroblastoma is present in about half of the children diagnosed with OMS, and in adults, OMS can be seen as a complication of lung cancer, breast cancer, and ovarian cancer.

While, vestibular paroxysmia (VP) may manifest when arteries in the cerebellar pontine angle cause a segmental, pressure-induced dysfunction of the eighth nerve. There are some case reports of neurovascular compression syndrome (NVCS) presenting with VP (4-7). We encountered a rare case of NVCS of the brainstem complicated with vestibular paroxysmia (VP) and autonomic symptoms combined with OMS. Here, we present the clinical course of the disease and report the findings in light of the existing scientific literature.

2. Case Report

A 36-year-old man with no remarkable medical history and chief complaints of vertigo and nausea, visited an ophthalmologist. Magnetic resonance imaging (MRI) showed no abnormality, and he was prescribed betahistine, kallidinogenase, adenosine triphosphate disodium hydrate granules, and alprazolam for the aforementioned symptoms. A few days later, his vertigo progressed and he was unable to walk, and...
was transported to our hospital by an ambulance. At hospitalization, he had clear consciousness, blood pressure was 110/62 mmHg, and body temperature was 36.2°C. He had vertigo and nausea, but without facial nerve palsy and tinnitus. He showed no decline in hearing or hearing sensitivity, no sense of ear obstruction, and no difference in hearing ability. Spontaneous nystagmus at rest was noted (horizontal nystagmus was omnidirectional, with biphasic horizontal nystagmus at bilateral lateral gaze, horizontal rotational nystagmus at upward gaze, and swaying omnidirectional saccadic abnormal eye movement during eye-opening in the dark [opsoclonus]). Marked trunk ataxia was found with no quadrupeds. He was unable to sit or stand. He had no other obvious neurological deficits. Momentary involuntary movements of the neck and shoulders (cervical myoclonus) were found. He also had myoclonus, restless leg syndrome, erectile dysfunction. Blood tests showed no abnormal findings (such as tumor markers) other than an LDL level of 182 mg/dL.

There were no remarkable findings in the auditory brainstem response. MRI at hospitalization indicated contact of the left vertebral artery (VA) with the dorso-lateral area of the ponto-medullary junction (Figures 1A and 1B). A marked bending or meandering of the vertebral artery was observed (Figure 1C). Chest-pelvic computed tomography showed no neoplastic lesion.

Based on the presented symptoms and imaging findings at admission, we suspected OMS and VP associated with NVCS caused by the left VA compressing the brainstem. Based on this initial diagnosis, the patient was administered carbamazepine (CBZ) 400 mg/day from day 1. Although the OMS and VP persisted, the seizure frequency decreased marginally. On day 2, rosuvastatin 5 mg and brotizolam 0.25 mg (oral) were started for managing the dyslipidemia and insomnia, respectively. Etizolam 1.5 mg/day was started on day 10. Lacosamide (LCM) 100 mg/day was started on day 12 which was subsequently increased to 200 mg/day on day 19. When the frequency of the OMS symptoms and VP decreased, clonazepam (CZP) (1.5 mg/day) was introduced to treat the restless leg syndrome. Erectile dysfunction, insomnia, and restless legs syndrome alleviated and the patient was able to walk, and was discharged on day 42. At 12 months post discharge, his OMS occasionally presented as seizures when talking with others. He continued all the previously mentioned prescription drugs except for etizolam and brotizolam because the mental symptoms, such as insomnia and anxiety, disappeared. He has since been in the outpatient follow-up.

Written informed consent was obtained from the patient for the publication of this case report and accompanying images, and the study design was approved by the appropriate ethics review board.

3. Discussion

To the best of our knowledge, although there have been reports of "spino-bulbo-spinal like" reflex because of the pressure of the dolichoectatic VA on the lower brainstem (4), this is the first reported case of OMS with VP and autonomic symptoms combined with NVCS of the brainstem. The main symptoms of VP (5-10) usually lasts for less than one minute and occur more than 30 times a day. Spontaneous or non-rotatable vertigo attacks occur spontaneously and repeatedly over a short period. VP is often caused by direct pulsatile compression of the eighth cranial nerve by an artery at the cerebello-pontine angle.

Neurovascular compression of the eighth cranial nerve is observed in more than 95% of cases presenting VP after MRI. Particularly, the involvement of the anterior inferior cerebellar artery loop is the highest, followed by the posterior inferior cerebellar artery, vertebral artery (VA), or veins. Treatment strategies involve administration of low-dose carbamazepine (200-600 mg/day) or oxcarbazepine (300-900 mg/day), which is equally effective in children. Additionally, lamotrigine, phenytoin, gabapentin, topiramate,
Therapeutically, low dose CBZ alone did not significantly reduce seizure frequency; therefore, we added LCM, a novel sodium channel blocker, and CZP, a drug for myoclonus, restless legs syndrome, and erectile dysfunction. VP and autonomic symptoms were thought to have improved significantly. Careful follow-up will be continued in the future.

In conclusion, we report a rare case of NVCS of the brainstem (most likely of the input to the vestibular nucleus) with VP and autonomic symptoms combined with OMS. Low dose CBZ, LCM, and CZP combined therapy contributed to the marked decrease of seizure frequency.

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References


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