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Case Report

Chiari malformation type I: atypical vestibular presentation with down-beating nystagmus – a case report

Sanjay Helale[1], Divyanshi Malhotra[2]*

[1]Criticare Asia Multi Speciality Hospital, Kurla, Mumbai, Maharashtra, India
[2]Department of Otorhinolaryngology, Khan Bahadur Bhabha Hospital, Kurla (w), Maharashtra, India

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***Correspondence:**
Dr. Divyanshi Malhotra,
E-mail: divyanshi.delhi@gmail.com

ABSTRACT

Arnold-Chiari malformation type I (CM-I) is a neurological disorder characterized by the caudal displacement of the cerebellar tonsils through the foramen magnum into the cervical spinal canal. The most common clinical manifestations include occipital headache and neck pain. However, vertigo is an uncommon presentation and may lead to diagnostic confusion with other vestibular disorders. A 39-year-old female presented with a ten-year history of occipitocervical pain accompanied by recurrent episodes of dizziness. Her symptoms clinically resembled cervicogenic dizziness for which she received treatment with minimal improvement. Further evaluation at a specialized center included videonystagmography, which demonstrated down-beating nystagmus. Magnetic resonance imaging (MRI) of the brain with craniovertebral junction assessment subsequently revealed herniation of the cerebellar tonsils consistent with chiari malformation type I. This case highlights an atypical presentation of CM-I in which vestibular symptoms predominated, while the classic symptom of persistent occipital headache was less prominent. Such atypical manifestations can delay diagnosis and appropriate management. Comprehensive neuro-otological evaluation, including vestibular testing and neuroimaging, plays a crucial role in identifying central causes of vertigo. CM-I should be considered in the differential diagnosis of patients presenting with unexplained vertigo or vestibular symptoms, particularly when associated with occipitocervical pain. Early recognition and appropriate imaging are essential for timely diagnosis and management. Following diagnosis, the patient was planned for foramen magnum decompression surgery.

Keywords: Chiari malformation type I, Down-beating nystagmus, Vertigo, Vestibular disorders, Foramen magnum decompression

INTRODUCTION

Chiari malformation is a neurological condition characterized by structural abnormalities at the base of the skull that cause downward displacement of the cerebellar tonsils or brainstem through the foramen magnum into the cervical spinal canal.[1] It is classified into four major types.[1] Type I, the most common form, is defined by caudal herniation of the cerebellar tonsils more than 5 mm below the foramen magnum without associated brainstem herniation or neural tube defects, and it usually becomes symptomatic during adolescence or adulthood. Type II, frequently associated with myelomeningocele in infants, involves herniation of the tonsils, vermis, and brainstem. Type III is a rare variant characterized by herniation of cerebellar or brainstem tissue into an occipital or upper cervical encephalocele, whereas type IV involves cerebellar hypoplasia or aplasia. Among these, type I is the most prevalent across all age groups.[2]

Chiari malformation type I can present with a wide range of neurological symptoms.[3] The most common manifestations are occipital headache and neck pain, while less frequent symptoms include dysphagia, glosso-pharyngeal neuralgia, and vertigo. This report describes an unusual case in which recurrent vertigo and chronic cervicogenic pain led to the diagnosis of an underlying Chiari malformation. Atypical presentations of Chiari

malformation type I may result in delayed diagnosis and management.[4,5] Additionally, variability in symptom presentation and severity can further complicate recognition.[6,7] This case report highlights a patient initially treated for persistent cervicogenic dizziness who was later diagnosed with Chiari malformation type I, emphasizing the importance of considering this condition in cases of unexplained vestibular symptoms.

CASE REPORT

A 39-year-old female presented to the hospital with a ten-year history of occipitocervical pain accompanied by recurrent episodes of dizziness. The episodes were aggravated by neck movements, particularly flexion and extension, and was relieved when the patient lay down in a neutral position. These episodes were also associated with gait unsteadiness and a sensation of swaying, which significantly interfered with her daily activities. The patient denied associated symptoms such as headache, tinnitus, hearing loss, nausea, or vomiting. For these complaints, she had previously been treated with a cervical collar and vestibular sedatives for approximately two years; however, only minimal symptomatic relief was achieved.

Upon presentation to our center, a comprehensive neuro-otological evaluation was performed. Video nystagmography revealed spontaneous, non-fatigable down-beating nystagmus without latency, which showed mild aggravation on downward gaze (Figure 2). The video head impulse test was within normal limits. No changes in nystagmus were observed during hyperventilation or the Valsalva maneuver. Detailed neurological examination, including cranial nerve assessment and cerebellar evaluation, revealed no abnormalities.

Considering the possibility of a central etiology for the patient's dizziness, magnetic resonance imaging (MRI) of the brain with craniovertebral junction evaluation was performed (Figure 1). Sagittal T1-weighted images demonstrated descent of the cerebellar tonsils more than 5 mm below the foramen magnum, consistent with a diagnosis of Arnold-Chiari malformation type I.

The patient subsequently underwent foramen magnum decompression surgery. The procedure involved bony decompression through removal of a portion of the occipital bone along with the posterior arch of the first cervical vertebra, thereby relieving compression at the craniovertebral junction. The patient experienced immediate postoperative symptomatic relief.

At the three-month follow-up, repeat videonystag-mography demonstrated complete resolution of the previously noted down-beating nystagmus (Figure 4). Clinical and neuro-otological examinations were normal. The patient reported complete resolution of vertigo and occipitocervical pain and was able to resume her routine daily activities independently.

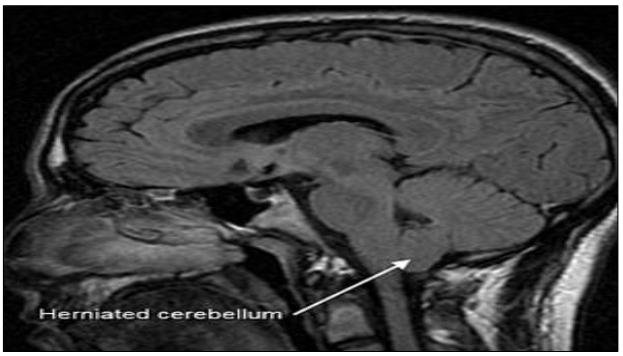

Figure 1: MRI brain with CV junction demonstrating herniation of cerebellar tonsil below the foramen magnum.

Figure 2: Video nystagmography image demonstrating spontaneous down-beating nystagmus suggestive of central vestibular involvement.

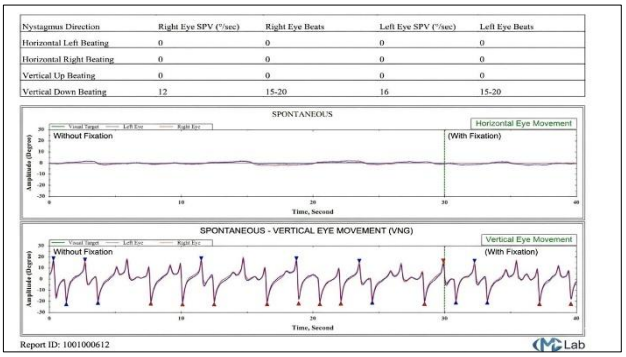

Figure 3: VNG graph showing spontaneous down beating nystagmus.

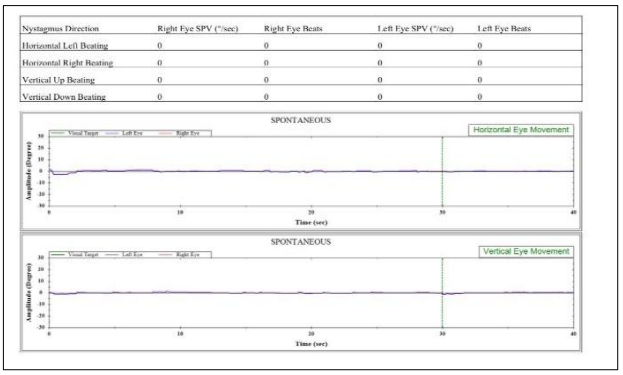

Figure 4: Months post operative VNG graph showing complete resolution of down beating nystagmus.

DISCUSSION

Chiari malformation type I (CM-I) is a structural abnormality characterized by the caudal displacement of the cerebellar tonsils through the foramen magnum into the upper cervical spinal canal.[8] The principal pathophysiological mechanism involves obstruction of the normal cerebrospinal fluid (CSF) circulation at the level of the foramen magnum.[9] This obstruction may result in increased pressure within the posterior fossa, leading to compression of critical neural structures including the cerebellum, medulla oblongata, and lower cranial nerves. In addition, compression of the vestibular nuclei or disruption of vestibulocerebellar pathways may produce vestibular symptoms such as vertigo. In the present case, aggravation of symptoms with neck flexion and extension suggests that dynamic changes in neck posture may further compromise the restricted space at the foramen magnum, thereby exacerbating neural compression or CSF flow disturbances.[10]

Dizziness as the predominant symptom in Arnold-Chiari malformation is relatively uncommon and may pose a diagnostic challenge for clinicians. In this case, videonystagmography (VNG) demonstrated spontaneous down-beating nystagmus, a key indicator of central vestibular pathology (Figure 3). The presence of down-beating nystagmus necessitates careful differentiation between central cerebellar causes, craniovertebral junction abnormalities, and peripheral vestibular disorders such as anterior semicircular canal benign paroxysmal positional vertigo (BPPV).[11]

Cerebellar flocculonodular pathology as a differential diagnosis

Both cerebellar flocculonodular lesions and craniovertebral junction abnormalities may present with central down-beating nystagmus.[12] However, in cerebellar flocculonodular pathology, the nystagmus typically worsens with lateral or downward gaze.[13] These lesions are also commonly associated with additional cerebellar signs such as ataxia and dysmetria, which were absent in the present case.[14,15]

Benign paroxysmal positional vertigo as a differential diagnosis

Anterior semicircular canal BPPV may also produce predominantly down-beating nystagmus, usually accompanied by a subtle torsional component and triggered by positional maneuvers.[16] However, the nystagmus in BPPV generally demonstrates a brief latency period and is fatigable, features that were not observed in our patient.[17]

Role of MRI with craniovertebral junction evaluation

This case highlights the crucial role of MRI of the brain with craniovertebral junction assessment in patients presenting with central down-beating nystagmus. Such imaging is essential for identifying structural abnormalities and distinguishing craniovertebral pathologies from other central causes.[18]

Management of Arnold-Chiari malformation type I

Management of CM-I depends largely on the severity of symptoms and the presence of neurological deficits.[19] Patients with mild or stable symptoms may be managed conservatively, including the use of a cervical collar to limit excessive neck movements.[19] However, in cases with persistent or progressive symptoms, as observed in this patient, surgical intervention such as posterior fossa or foramen magnum decompression is recommended, as described by Kim et al.[20]

The primary objective of foramen magnum decompression is to relieve mechanical obstruction caused by herniated cerebellar tonsils and to restore normal CSF dynamics at the craniovertebral junction.[21] This decompression reduces pressure on the brainstem and cerebellum, thereby alleviating associated neurological symptoms including vertigo and nystagmus.[22] In the present case, the complete resolution of long-standing vertigo and occipitocervical pain following surgery highlights the effectiveness of this intervention in carefully selected patients with CM-I presenting with atypical vestibular manifestations.

CONCLUSION

The case highlights that atypical presentations of Chiari malformation type I can often result in delayed diagnosis and management emphasizing on the need for a comprehensive neuro ontological examination and radiological evaluation and the importance of considering this diagnosis in cases of unexplained vestibular symptoms.

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