

Case Report

Chiari type 1 malformation presenting as an atypical benign paroxysmal positional vertigo in a 16 year old patient

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ABSTRACT

Dizziness and vertigo are very common symptoms and can be difficult to distinguish clinically. Vertigo is a specific subtype of dizziness in which a patient experiences the illusion of motion (often rotatory) of either themselves or their surroundings. Vertigo can be either peripheral (arising within the labyrinth) or central (brain). It is important to identify central aetiologies as they can be fatal if missed. This case report narrates a 16-year-old male who presented complaining of vertigo related to changes in head position which was initially misdiagnosed as a benign paroxysmal positional vertigo (BPPV). It was several years before he was correctly diagnosed as having Chiari type 1 malformation on MRI scan. He underwent a surgical decompression with complete alleviation of his symptoms. All cases of atypical vertigo should be evaluated in detail. Patients with failure of initial management should undergo imaging to identify any central pathology.

Keywords: Chiari malformation, Benign paroxysmal positional vertigo

INTRODUCTION

Dizziness and vertigo are very common symptoms and can be difficult to distinguish clinically. Vertigo is a specific subtype of dizziness in which a patient experiences the illusion of motion (often rotatory) of either themselves or their surroundings. Vertigo can be either peripheral (arising within the labyrinth) or central (brain). It is important to identify central aetiologies as they can be fatal if missed. Chiari malformations are one of the central causes of vertigo. These malformations are a group of congenital hindbrain and spinal cord abnormalities characterised by herniation of the posterior cranial fossa through the foramen magnum into the upper cervical vertebrae.^{1,2} They were classified into four types pertaining to the degree of herniation by Chiari in 1896.^{1,3} Type 1 is associated with downward displacement of cerebellar tonsils without herniation of the cerebellum. Type 2 is the most common type. It is associated with herniation of cerebellum and the brainstem through the

foramen magnum and is associated with meningomyelocoele in majority of cases. Type 3 has the same features as type 2, along with herniation of entire cerebellum and the fourth ventricle through the foramen magnum and a bony defect in the occipital bone. It is the most dangerous form of malformation. Type 4 is known as cerebellar hypoplasia in which the cerebellar tonsils are located in a normal position but parts of the cerebellum are missing.

We report a case of Chiari type 1 malformation which was initially misdiagnosed as a case of BPPV. Once correctly diagnosed, he underwent neurosurgical intervention for the control of his symptoms.

CASE REPORT

A 16-year-old male presented to the outpatient clinic with complaints of positional vertigo (worst on tilting his head to the right). Interestingly he had had this symptom since

he was 5 years old and was initially diagnosed with BPPV. At the age of 5 years, he had episodes of occasional vertigo, paleness and nausea. During these attacks he would remain sitting or standing still, not being touched, until it resolved. The episodes lasted 2-3 minutes and within 15-20 minutes would completely abate. There was no apparent trigger and episodes could occur 2-3 times a day but then be absent for 1-2 weeks at a time. His past medical history, systemic examination including cranial nerves and cerebellar signs were unremarkable. The only positive family history was that his father suffered from longstanding unilateral hearing loss. At this point, Cinnarizine was trialled but on clinical review it was found to have had no effect. The episodes remained short-lived and resolved within 1-2 minutes. Another antihistamine was trialled and it appeared he entered remission and was discharged.

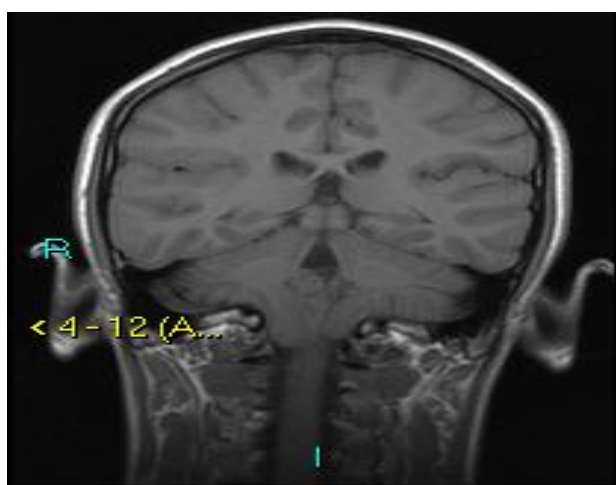


Figure 1: MRI T1 weighted coronal image showing right cerebellar herniation.



Figure 2: MRI T2 weighted sagittal image showing 8mm cerebellar herniation.

At the age of 9 years, he presented to a nearby ENT department with history of repeated attacks of vertigo which were short-lived. He described movement specific

vertigo— worst getting out of bed in the morning and turning right, looking upwards and reading. Rotatory vertigo lasted a few seconds with associated nausea. He had an associated right temporal headache which would last a few hours in the morning. There were no other symptoms and no family history of migraine. He had a strongly positive Dix-Hallpike test and right Epley's manoeuvre was performed in the clinic. He was then discharged from the clinic.

He presented to our department at the age of 16 years with a clear history of the room spinning while turning to the right, experiencing nausea without vomiting and alleviation of symptoms by tilting his head to the contralateral side. Triggers included emotions (becoming excited), exercising, banging his head or bending over. On examination he had normal findings on otoscopy and neurology examination. However when he bent his head to his right, it triggered his vertigo and he had immediate persistent vertigo with nystagmus. His pure tone audiometry and tympanometry were normal. In view of persistent vertigo, MRI was requested which revealed an inferior herniation of the cerebellar tonsils by 8mm suggesting a type 1 Chiari malformation. He was referred to the neurosurgery department and underwent a surgical decompression with complete alleviation of symptoms post operatively.

DISCUSSION

Vertigo has a complex aetiology and can be classified as peripheral or central. BPPV is a disorder characterised by rotatory vertigo associated with changes in head positions and is thought to be due to displacement of otoconia in the semicircular canals. Vertigo is short lived, fatiguable with no nausea or vomiting. Patients with history of head trauma, labyrinthitis and central nervous system related dizziness have the largest recurrence rates inspite of canalolith repositioning manoeuvres.⁴

According to a recent study in 2015⁵, the pathophysiology of type 1 Chiari malformation is simply the obstruction of the normal pulsatile movement of CSF across the foramen magnum. Chiari type 1 malformations present late in adult life.⁶ As per Meadows et al up to 30% of patients with tonsillar herniation of 5 to 10 mm may be asymptomatic.⁷ The most common clinical symptom is pain, including occipital pain, neck pain, back pain, and upper limb pain. Other clinical manifestations include involvement of cranial nerves, sensory loss, numbness and tingling, muscle weakness, and ataxia.⁸ Involvement of the trigeminal nerve causes neuralgia whereas involvement of 8th to 12th cranial nerves can cause hearing loss, tinnitus, dysphagia and change in voice. Positional nystagmus is common. It is most commonly down beating or lateral beating. Persons with Chiari may develop vertigo after spending some time with their head inclined on their trunk. Thus the Chiari can cause central vertigo.

In this patient, the main symptom was episodic vertigo with changes in head position and hence was misdiagnosed as benign paroxysmal positional vertigo. Failure of improvement with canalolith repositioning manoeuvres should raise a suspicion of some other cause of vertigo and should be evaluated with imaging. MRI scan is considered a gold standard investigation for the diagnosis of central causes of vertigo.⁹

Similar cases have been reported in the past.^{10,11} A study in Helsinki university hospital retrospectively analysed 439 otological patients and found the prevalence of Chiari- malformation was 0.9% and concluded the possibility of this should be considered in patients with atypical benign positional vertigo.¹²

CONCLUSION

All cases of atypical vertigo should be evaluated in detail. Patients with failure of initial management should undergo imaging to identify any central pathology.

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