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Short Communication

A Complex Presentation of Vestibular Paroxysmia in an Adolescent With Wolff- Parkinson-White Syndrome

Manuel Nunez, MD [a], Michal T. Ruprecht, BS [b], Alex S. Aguirre, MD [a], Alcy Torres, MD [a, *]

[a] Boston University Chobanian & Avedisian School of Medicine, Boston, Massachusetts
[b] Wayne State University School of Medicine, Detroit, Michigan

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ABSTRACT

Vestibular paroxysmia is an episodic vestibular disorder resulting from compression or irritation of the eighth cranial nerve. This disorder is a rare and difficult diagnosis in children. We report the case of a 16-year-old adolescent male with a history of syncope and coronavirus disease 2019 infection four months prior who presented with intermittent episodes of *vertigo* and unsteadiness several times a week. These events started abruptly, and he appeared frozen. However, he remained conscious and was able to answer questions. He subsequently resumed normal activity in less than a minute without seizure stigmata or postictal period. His general and neurological examinations were unremarkable. Extensive diagnostic evaluation yielded negative results, except for an electrocardiogram consistent with Wolff-Parkinson-White syndrome. However, his symptoms persisted after cardiac ablation, suggesting they were not related to this arrhythmia. Following unsuccessful trials with various medications, his symptoms resolved with carbamazepine. Early recognition and appropriate treatment of this condition could substantially improve the quality of life for affected individuals.

Background

Vertigo is characterized by a sensation of motion without actual head or body motion. The condition can be categorized into a spectrum of vestibular syndromes, one of which is vestibular paroxysmia.[1] Clinicians have historically made efforts to distinguish between dizziness and vertigo, but there has been a shift in this tendency.[1] Despite introducing the term vestibular paroxysmia in 1994, its prevalence among the pediatric population is unknown. This diagnosis requires the presence of all diagnostic criteria.[2] Past reports were associated with arterial compression of the vestibular nerve.[3]

Case study description

A 16-year-old male with a history of syncope and coronavirus disease 2019 infection four months ago presented with acute onset, 20- to 30-second-long episodes of dizziness, unsteadiness, and swaying. These episodes gradually increased in frequency to about one to four per week. During these episodes, the patient denied any changes of consciousness or triggered by positional changes. An electrocardiogram demonstrated Wolff-Parkinson-White (WPW) syndrome. These events were not related to changes in the cardiac rhythm. His symptoms persisted despite successful catheter ablation.

The patient denied headaches, vision, hearing changes, or a history of head trauma. There is a family history of migraines in both parents, but no vertigo. Additionally, he had no history of clinical seizures, and general, neurological, and vestibular examinations—including the Dix-Hallpike maneuver—were all unremarkable.

Two electroencephalograms, including a 24-hour study capturing three typical events, showed no evidence of epileptiform activity. Magnetic resonance imaging of the brain, including the temporal bones and magnetic resonance angiography, were normal; specifically there was no vascular compression of the vestibular nerve. A diagnosis of vestibular migraine was considered, but treatment with topiramate did not alleviate the patient's symptoms. He was then started on venlafaxine, which provided some anxiety relief, but the paroxysmal vertiginous events persisted, hindering the patient's quality of life. The vestibular testing

* Communications should be addressed to: Dr. Torres; Boston University Chobanian & Avedisian School of Medicine; 801 Albany Street; Boston, MA 02118.
E-mail address: artorres@bu.edu (A. Torres).

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showed no evidence of benign paroxysmal positional vertigo. His final diagnosis was noncompressive vestibular paroxysmia. Our patient was treated with carbamazepine 100 mg twice daily (3 mg/kg/d) and demonstrated an immediate and lasting response despite low serum levels of carbamazepine.

Discussion

The differential diagnosis of this patient included several possibilities. The episodes were paroxysmal and stereotyped, thus we contemplated seizures as an etiology. However, the preservation of consciousness and the lack of other seizure stigmata or postictal period did not support this diagnosis, and his electroencephalograms were unremarkable. Although vertigo is an uncommon presentation in patients with WPW syndrome, the timing of this simultaneous presentation made it difficult to ignore. Considering that early treatment of WPW syndrome is a life-saving procedure, he underwent a successful ablation, but his symptoms persisted. Despite his normal neurological examinations, a detailed imaging of the brain and the blood vessels was obtained, but they did not show a specific etiology. We also considered the possibility of a migraine-equivalent syndrome. Nevertheless, given that the patient's symptoms lasted less than one minute, he did not fulfill the vestibular migraine diagnosis criteria.[4] The lack of lingering symptoms despite family history of migraine in both parents and poor response to migraine treatment made this diagnosis less likely. A postinfectious process was also in our differential. The patient had a coronavirus disease 2019 infection four months before his initial presentation, and cranial nerve abnormalities have been reported, such as olfactory nerve dysfunction.[5] However, considering how frequently this occurs, it is difficult to prove a direct relationship. Finally, his presentation is consistent with vestibular paroxysmia, which is usually caused by vascular compression of the eighth cranial nerve in adults. However, this characteristic was not found in our patient. This case suggests and supports that carbamazepine may be effective in treating this condition.[2,6] The patient's fluctuating course was not consistent with typical vestibular neuritis, and the lack of pain was not typical for neuralgia.

Conclusions

Vestibular paroxysmia is a rare but treatable condition in pediatric neurology. Our case illustrates the importance of including this condition in the differential diagnosis of a pediatric patient with recurrent, brief episodes of vertigo. We argue that management of vestibular paroxysmia with carbamazepine is effective and that early detection could improve a patient's quality of life.

CRediT authorship contribution statement

Manuel Nunez: Writing – original draft, Resources, Methodology. **Michal T. Ruprecht:** Writing – review & editing. **Alex S. Aguirre:** Writing – review & editing, Resources. **Alcy Torres:** Writing – review & editing, Writing – original draft, Visualization, Validation, Supervision, Resources, Methodology, Investigation, Formal analysis, Conceptualization.

Declaration of Competing Interest

We know of no conflicts of interest associated with this publication, and there has been no significant financial support for this work that has influenced its outcome.

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