Case Report

# Schwannoma of the vestibulocochlear nerve presenting as isolated paroxysmal vertigo

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## ABSTRACT

We report a patient with a schwannoma of the eighth cranial (vestibulocochlear) nerve who presented with isolated episodes of paroxysmal vertigo and positive Dix-Hallpike maneuver, and without the common features of hearing loss, disequilibrium and tinnitus. There are no previous reports of paroxysmal episodes of vertigo as the sole manifestation of schwannoma of the vestibulocochlear nerve. Hence, recurrent paroxysmal vertigo should therefore prompt the physician to rule out schwannoma of the vestibulocochlear nerve as a potential cause even in the presence of normal hearing tests.

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Patients presenting with a history of progressive unilateral sensorineural hearing loss, usually accompanied by tinnitus, imbalance or both should be highly suspected of suffering from schwannoma of the vestibulocochlear nerve (VCN) (formerly called acoustic neurinoma).[1] We present a case of VCN schwannoma presenting with episodes of paroxysmal vertigo as a single neurological manifestation.

**Case Report.** A 46-year-old right-handed man was admitted following 2 episodes of spinning vertigo, which lasted approximately 4 minutes each. The vertigo was not related to any specific position or movement of the head. The episodes were followed by sweating, nausea and vomiting. There was no tinnitus or hearing loss. During his admission, he had one similar episode. There was no history of fever, weight loss or nocturnal sweating. Nine years ago he had a one-week episode of ataxia, tinnitus and oscillopsia for which no investigations were carried out. He had diabetes, which was well controlled with diet. He stopped smoking 6 years ago. Neurological examination showed an alert patient with normal coherent speech and cognitive functions. Neuro-ophthalmologic examination revealed normal visual acuity, fields and fundi. The pupils were reactive to light and near stimuli. Eye movements were normal and there was no spontaneous, or gaze-evoked nystagmus. Post head shake nystagmus was absent. There was a positive response on the Dix-Hallpike maneuver in the right head-hanging position, consisting of an acute-onset nystagmus fatigable with repeated maneuvers and associated with dysphoria. Hyperventilation provoked nystagmus was absent. Sensation in the face was intact. Otoscopy was normal. There was no sensorineural hearing loss. The bulbar functions were intact. There were no sensory or motor deficits in the upper and lower extremities, all tendon reflexes were normal and symmetric, and plantar responses were flexor. Blood pressure was 144/73 mm Hg. Cardiopulmonary examination was normal. There were no cervical bruits or lymphadenopathies in the cervical, supraclavicular or axillary areas. Although the clinical impression was that of benign paroxysmal positional vertigo, the atypical presentation (lack of a history of positioning vertigo) prompted further evaluation, including

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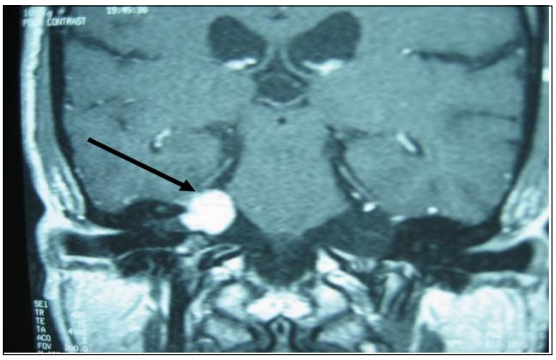

**Figure 1** - Coronal MRI scan (gadolinium-enhanced-T1-weighted image) showing right-sided schwannoma of the vestibulocochlear nerve (arrow).

neuroimaging. All serum laboratory and hematological values were within the normal range, except for HbA1C, which was 8.1%. Screening for pheochromocytoma was negative. Cardiac Holter evaluation was normal. Pure tone audiometry showed normal hearing thresholds in both ears. Speech discrimination was 98% in both ears. Brain CT scan without contrast on admission was reported as normal. However since a fossa posterior pathology could not be ruled out, an MRI (T1 and T2 with gadolinium contrast enhancement) was performed, which revealed a contrast enhancing tumor (approximately 1.5 x 1.5 cm in size) at the right cerebellopontine angle with extension and widening of the right internal auditory canal (**Figure 1**) suggesting VCN schwannoma. There were no supratentorial, cerebellar or brainstem lesions. An MR angiography was normal. Brainstem auditory evoked response (BAER) was normal. The patient underwent total excision of the tumor, but unfortunately suffered permanent sequelae (ipsilateral facial palsy and deafness). The tumor proved histologically to be schwannoma.

**Discussion.** Episodes of paroxysmal vertigo are commonly found as presentations of Ménière disease, vestibular neuronitis, perilymph fistula, benign paroxysmal positional vertigo, peripheral vestibular paroxysmia, vestibular migraine and vertebrobasilar insufficiency. However, in the presence of other symptoms of VCN schwannoma (for example, hearing loss, tinnitus or disequilibrium) this type of vertigo has been reported in less than 5% of patients.[1] In addition, only one case of acute paroxysmal positioning vertigo with positive Dix-Hallpike maneuver has been reported associated with a large contralateral VCN schwannoma.[2] Therefore, recurrent paroxysmal vertigo as the sole manifestation of this type of cerebellopontine angle tumor can be considered to be extremely rare. Progressive unilateral hearing loss occurs in approximately 70% of patients with VCN schwannoma, while tinnitus and vertigo or disequilibrium present both in approximately 50% of cases.[1] Overall, approximately 21% of patients with VCN schwannoma present with atypical initial features.[1] Normal hearing including strict audiological criteria (mean pure-tone thresholds normal for age and speech discrimination, and no presenting symptoms of hearing loss) has been reported in up to 11% of patients with VCN schwannoma.[1] Therefore, any patient with a history of a combination of atypical presentations such as; headaches, otalgia, facial nerve weakness, fullness of the ear, unilateral facial pain, altered sensation in unilateral face, painful unilateral eye or unexplained vomiting and vertigo (either episodic or single acute, or positional vertigo) presents a high suspicion index. Surprisingly, tumor size does not relate to the duration of disease and bulky tumors frequently present with minimal neurological deficit or symptoms.[3] This is probably explained by the nature of the expansion of the tumor. Tumor enlarging within the internal auditory meatus is more prone to produce early manifestations such as hearing loss, tinnitus and vertigo. However, despite this, postoperative success will be greater when the intervention is carried out if the tumor is small.[4] The relative small size of the tumor and its location - predominantly filling the cerebellopontine space - probably explained the mild and unusual presentation, and added to the fact that both audiometric testing and BAER were normal in our patient. The latter has been reported negative in up to 40% of patients with small NCV schwannoma.[5]

In summary, we report a patient with recurrent paroxysmal attacks of vertigo and positive Dix-Hallpike maneuver as the sole manifestation of VCN schwannoma. This unique case demonstrates that even the presence of a negative neurotological evaluation, the features presented in this case should prompt clinicians to rule out VCN schwannoma.

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