Benign Paroxysmal Positional Vertigo Diagnosed in a Patient with Idiopathic Cervical Dystonia

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ABSTRACT

Idiopathic cervical dystonia (CD) is a focal dystonia characterized by involuntary contraction of the muscles of the neck causing twisting movements and abnormal head and neck postures. Its etiology is unclear; however, intracerebral neuronal circuit pathologies are highly considered. On the contrary, benign paroxysmal positional vertigo (BPPV) is the most common peripheral vertigo diagnosed. We present a patient with CD suffering from severe vertigo who was diagnosed with BPPV. This is a very unique case representing the co-occurrence of a rare central nervous system pathology and a peripheral inner ear disease. The cause and effect relation between two pathologies is under scope. We present a 55-year-old woman with CD attended our clinic with severe vertigo and nausea. Left posterior semicircular canal BPPV (PSCC BPPV) was diagnosed. Involuntary head and neck muscle contractions caused by CD appear to have contributed to the development of BPPV in this case. However, increased neuronal activities causing CD could also have led to BPPV in the vestibular nuclear level. Further investigations are essential. Peripheral vestibular pathologies and their association with CD are not clearly determined in the literature so far. To our knowledge, there are no other cases reported regarding this co-occurrence.

Keywords: Vertigo, cervical dystonia, focal dystonia, dizziness

INTRODUCTION

Idiopathic (primary) cervical dystonia (CD) is a coercive undesirable pathology characterized by involuntary sustained or intermittent contractions of the muscles of the neck causing twisting movements and abnormal head and neck postures (1-3). Adult onset primary dystonia is the most common form of focal dystonia (4). Prevalence is reported as 20–137 cases per million (4). It is more dominant in women, with an average age of onset in early and mid-40s.

Primary dystonia is a pure dystonia, and unlike secondary cases, no associating neurological pathologies are observed. Its etiology is unclear; however, dysfunction of motor control network is strongly considered. Extrapyramidal system, especially the basal ganglia, involvement was suggested (1, 2, 5). In some specific forms of dystonia, including CD, gross thalamic volume reduction was detected (4). Diagnosis is still based on history, neurological examination, and exclusion of other pathologies (3).

In the past, some researchers evaluated the central vestibular system in patients with CD. In 1989, Huygen et al. (1) detected vestibular hyperreactivity in patients with dystonia. However, they were unable to find a specific relation regarding the etiopathogenesis of vestibular symptoms in these patients. Cervical dystonia is thought to be a result of systems-level disorder; therefore, vestibular pathologies may be associated due to proximity of vestibular nuclei and pathways. Though very limited data have been reported so far, the co-occurrence of central vestibular pathologies and CD may be more common than expected. However, the authors of this current report present a unique patient with CD, diagnosed with posterior semicircular canal benign paroxysmal positional vertigo (PSCC BPPV), a peripheral vestibular pathology.

To our knowledge, there are no other cases reported in the literature regarding this co-occurrence.

CASE REPORT

A 55-year-old woman was referred to our clinic with a complaint of vertigo and nausea for 5 days. She claimed that she had not experienced such symptoms previously. Her complaints were aggravated with head rotations especially when turning left. No additional otological problems including hearing loss, tinnitus, ear fullness, or pressure sense were mentioned. She was diagnosed with CD 3 years ago. Since the diagnosis, botulinum toxin (Botox;
Allergan, USA) has been injected into her cervical muscles every 6 months, and through this therapy, symptoms were relieved but not terminated. No other history of co-morbid systemic or neurological diseases, head trauma, infections, surgery, or allergy was noted. She mentioned no previous vestibular disorders and/or migraine.

On her physical examination, involuntary continuous tonic muscle contractions of the neck resulting in right side laterocollis were observed. She was attempting to control the involuntary head movements by sensory tricks (gently touching her chin). Otherwise, tremor occurred during every head rotation toward all sides. Her otorhinolaryngological examination was in normal limits, except the left Dix–Hallpike maneuver; a torsional left up-beating nystagmus with vertigo was recorded. Nystagmus initiated after approximately 10 s of latency and lasted nearly 6 s. Left PSCC BPPV was diagnosed, and for canalith repositioning, Epley maneuver was performed. Temporal high-resolution computed tomography, magnetic brain imaging, and audiotoympanometric evaluations were within normal limits. In her next visit 3 days following the Epley maneuver, she was disease-free with no complaints of vertigo and nausea.

Written informed consent was obtained from the patient participating in this case study.

**DISCUSSION**

Benign paroxysmal positional vertigo is the most common type of vertigo. Its major symptom is sudden vertigo attacks induced by alterations in head position. The pathophysiology is related with loose otoconia dislodged from the utriculosaccular region. These free floating otoconia respond to specific gravitational forces and displace the cupula of PSCC (canalolithiasis). In 1969, Schuknecht proposed another form of this theory named as “cupulolithiasis” and demonstrated the attached otoconia to the cupula of PSCC of the affected side (6–8). The etiology may be idiopathic (50%–70% of all cases) or secondary to infections, migraines, Meniere’s disease, otologic, non-otologic surgery, prolonged bed rest, trauma, magnetic resonance imaging, vascular and metabolic pathologies, and hormonal changes (7, 9, 10).

In 2013, Hallberg et al. (11) reported an 18-month-old infant with both torticollis: postural imbalance and torsional nystagmus. They observed abnormal cervical vestibular evoked myogenic potentials (c-VEMP) indicating left side saccular dysfunction.

Accordingly, authors of this report suggest two possible mechanisms of action. Primarily, similar to prolonged bed rest, abnormal sustained head and neck postures caused by involuntary neck movements may initiate the detachment of otoconia through the semicircular canals. She had a tendency to have a right side deviating head and neck position. Therefore, right side PSCC BPPV appears to be more likely. However, the etiology of developing a left PSCC BPPV remains obscure, making this case more featured.

The other possible pathophysiology may be related with increased neuronal activity in the affected cerebral regions of patients with CD. Thus, the vestibular pathways and the nuclei may be involved, leading to saccular dysfunction and BPPV. Münchau et al. (5) suggested that a subclinical basal ganglia lesion coupled with a lesion affecting the systems that control head and neck movements (including the vestibular system) causes cervical dystonia. Stimulation of the nucleus of Cajal, which is closely related to vestibular nuclei, initiates rotational head movements toward the side of the stimulus (5). On the contrary, recent findings show that vestibular reflexes are mildly impaired in this pathology. The suppression of click evoked vestibulocollic reflex obtained from the sternocleidomastoid muscle is considered as a consequence of the long duration of dystonia, not a primary co-existing abnormality (5, 12). Another study by Rosen gren et al. (13) found intact vestibular evoked myogenic potentials, suggesting that vestibular dysfunction is not a result of CD itself.

In our case, being a middle-aged woman is a probable risk factor for BPPV; however, there were no other accompanying chronic diseases, trauma, infection, neurotologic pathologies, or identified metabolic disorders, or history of surgery that could have provoked BPPV.

She was not referred to physical therapy, which could have initiated BPPV, and it was noted that Botox® injection was performed into both sternocleidomastoid, trapezius and occasionally paravertebral muscles, in appropriate dosages applied by the neurologists in the erect sitting position that is not expected to provoke otococinal displacement. Her last botulinum injection was in April 2016: 4 months prior to her vertigo attack. No evidence was found in the literature regarding a possible peripheral vestibular adverse effect of Botox® injection.

**CONCLUSION**

Benign paroxysmal positional vertigo and CD togetherness is an original clinical case. Abnormal vestibular functions and involuntary cervical muscle contractions may co-occur, but the association of peripheral vestibular pathologies with CD has not been reported in the English literature so far.

In our case, cervical dystonia is believed to induce BPPV due to sudden involuntary head and neck muscle contractions. The contribution of central vestibular pathologies remains controversial; however, the general view is that the combination of basal ganglia defects with vestibular deficits may contribute to CD. It is worth mentioning that in our case, the patient’s CD started much earlier than her vestibular symptoms. Further, her vestibular symptoms were not permanent. Apparently, PSCC BPPV in our patient is a consequence of abnormal postural movements causing otoconial dislocation.

To justify the involvement of vestibular nuclei and their possible effects on saccule, causing BPPV in patients with CD requires further analysis on wide patient series, including VEMP measurements.

**Informed Consent:** Written informed consent was obtained from the patient who participated in this study.

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REFERENCES


2. Zoons E, Tijssen MA. Pathologic changes in the brain in cervical dystonia pre- and post-mortem - a commentary with a special focus on the cerebellum. Exp Neurol 2013; 247: 130-3. [CrossRef]


8. Schuknecht HF. Cupulolithiasis. Arch Otolaryngol 1969; 90(6): 765-78. [CrossRef]


13. Rosengren SM, Colebatch JG. Vestibular evoked myogenic potentials are intact in cervical dystonia. Mov Disord 2010; 25(16): 2845-53. [CrossRef]