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We describe the case of a 34-year-old patient presenting with the unique combination of bilateral vestibulopathy in combination with noise- and pressure-induced nystagmus. Bilateral vestibular dysfunction was demonstrated by pathological results in video-based head impulse testing as well as in caloric testing. In contrast, cervical vestibular-evoked myogenic potentials were normal, demonstrating normal sacculus function. Due to the positive Tullio phenomenon, semicircular dehiscence syndrome was excluded. Recently, this symptom combination was related to the histopathological finding of vestibular atelectasis.

**Key Words:** Bilateral vestibulopathy, noise- and pressure-induced nystagmus, vestibular atelectasis.

**Level of Evidence:** NA.

**INTRODUCTION**

Bilateral vestibulopathy is a severely cumbersome impairment as it leads to severe gait disturbance with postural instability and unsteady gait. Here we describe the case of a young patient presenting with the unique combination of bilateral vestibular dysfunction in combination with noise- and pressure-induced nystagmus.

**CASE REPORT**

The 34-year-old male patient presented in our outpatient clinic for vertigo. His complaints started about 4 years before with a short intensive vertigo attack during the start of an airplane flight that began after the plane took off. In the following years he suffered from short vertigo attacks once a month up to once a week. In parallel, he recognized short eye movements provoked by loud noises or by changes of air pressure (e.g., when opening a window or driving a car into a tunnel). The Tullio phenomenon was positive in both ears, but was more pronounced in the right-hand side than on the left-hand side, so that the patient used his phone only on the left ear. The short vertigo attacks ceased for 1 year, and then he suffered from gait disturbance aggravated in darkness. His hearing was normal.

In clinical examination the patient showed bilateral pathological head impulse test, a positive Romberg test, and an unsteady gait and otherwise normal neurological status. Loud noises and Valsalva maneuver consistently evoked short horizontal nystagmus for seconds. Caloric testing showed a bilateral vestibular hypoxcitability. A video-based head impulse test revealed dysfunction of all six semicircular canals. In contrast, cervical vestibularevoked myogenic potential (cVEMP) did not show signs of saccular dysfunction. Hearing tests were normal.

Because of the positive Tullio phenomenon, high-resolution computed tomography of the temporal bone was performed but did not show superior semicircular canal dehiscence. Also, the other semicircular canals were examined in detail, but did not show any pathology. In addition, 3T magnetic resonance imaging with three-dimensional reconstruction of the vestibular organs did not show any pathology. Figure 1 shows some of the clinical findings.

**DISCUSSION**

The concept of vestibular atelectasis was initially introduced 1988 by Merchant and Schuknecht. In temporal bone histopathologic samples, a collapse of the walls of the ampullae and utricle not explained by histological artifacts was recognized in rare cases. The authors presumed that the collapsed membranes interfered with the motion mechanics of the cupulae and otolithic membranes.

Recently, Wenzel et al. described four patients with a similar symptom constellation as our patient. All patients showed the combination of bilateral vestibular dysfunction and sound- and/or pressure-induced nystagmus. Due to the symptom of noise-induced nystagmus, labyrinthine dehiscence was excluded in all patients. In addition, cVEMP revealed to be normal in contrast to...
semicircular canal dysfunction in head impulse testing and caloric testing.

Although the finding of a collapse of the walls of the ampullae and utricle can only be a (postmortem) histological diagnosis, Wenzel et al. related this rare and unique symptom constellation to the diagnosis of vestibular atelectasis based on theoretical considerations. It has been hypothesized that the collapse of the membranous labyrinth may approach the stapes closer to the vestibule, so that as a consequence, motions of the stapes may stimulate the otolith organs.

The etiology of it is still unknown. Our patient did not meet the diagnostic criteria for Ménière’s disease or perilymphatic fistula and did not show paraclinical signs for autoimmune syndromes or the intake of toxic agents or drugs. Physiological studies have shown even a tiny dehiscence can cause canal neurons to respond to sound and vibration. However, a semicircular canal dehiscence may be unlikely here because of the bilateral nature of the symptoms. Conspicuous is the bilateral occurrence in all of the patients described so far, so a genetic origin of the syndrome has been discussed.

It is notable that the first symptoms occurred after the takeoff of an airplane. As the causative agent for this condition is unclear, it could be speculated if a change of cabin pressure is able to cause the membranous labyrinth to collapse against the stapes footplate, which is found bilaterally in a surprising number of temporal bones, and thus may cause the Tullio phenomenon? Actually, no specific therapy is known for this condition. We subjected the patient to multimodal vestibular rehabilitation.

**CONCLUSION**

Bilateral vestibulopathy generally causes a significant limitation of quality of life due to postural imbalance and unsteadiness of gait. The combination of bilateral vestibulopathy with noise- and pressure-induced nystagmus is a characteristic but rare condition and may be related to vestibular atelectasis, although this relationship may be controversial.

Fig. 1. Clinical findings of the case. Bilateral semicircular canal dysfunction as shown by video-based head impulse test; normal sacculus function in cervical vestibular-evoked myogenic potential (cVEMP) testing. Normal findings on both sides in computed tomography (CT) of the temporal bone and magnetic resonance imaging (MRI) of the inner ear and cerebellopontine angle. SD = standard deviation.
BIBLIOGRAPHY