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Acute Bilateral Sequential Vestibular Neuritis

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Two cases of bilateral sequential vestibular neuritis demonstrate the significant persistent disequilibrium that follows involvement of the second ear. The etiology for the loss of vestibular function is postulated to be a viral neuritis. Vestibular suppressant drugs are helpful in relieving nausea and vomiting in the acute phase of the disease; however, they are of no therapeutic value for the protracted disequilibrium following involvement of the second ear. An awareness of this disorder as a disease entity will minimize diagnostic and therapeutic frustration on the part of the physician and provide a realistic prognosis for the patient. Unfortunately, the prognosis is for permanent but somewhat lessening disequilibrium with the passage of time and depends in great part on the subject's age.

In a previous report, Schuknecht and Kitamura[1] presented postmortem histologic evidence to verify the occurrence of acute degenerative neuropathy of one or more branches of the vestibular nerves in subjects with acute attacks of vertigo. The characteristic clinical features were severe vertigo without hearing loss, accompanied by objective evidence of vestibular abnormality such as nystagmus and diminished or absent caloric response. The pathologic changes were shown to be atrophy of one or more vestibular nerve trunks, with or without atrophy of their associated sensory organs. It was suggested that these disorders be described by the term "vestibular neuritis."

To further elucidate and document a variant manifestation of this disorder, we present the clinical histories of two subjects who we believe have experienced isolated, bilateral, sequential, acute, severe, degenerative neuropathies of the vestibular nerves.

CASE REPORTS

Case 1

A 70-year-old woman awakened with severe vertigo, sweating, diarrhea, nausea, vomiting, and ataxia without auditory symptoms. She was admitted to the hospital where vestibular suppressive medication was given and extensive medical studies were performed. The results of general physical examination, chemical analyses of the blood, electrocardiogram, skull x-ray films, radioactive brain scan, and blood flow studies were normal. Findings on neurologic examination were reported to be normal except for ataxia. No record was made of nystagmus, and no caloric tests were done. She had no auditory symptoms, and no hearing tests were performed. She was discharged on the eighth hospital day, still requiring assistance when walking. The ataxia gradually subsided over a period of several weeks, after which she felt quite well.

Eighteen months later (at the age of 72 years), she experienced an identical episode of severe vertigo, nausea, vomiting, diarrhea, and ataxia without auditory symptoms. On this occasion she was treated at home with vestibular suppressant medication. The acute vertigo gradually subsided over a period of several weeks, but she continued to feel unsteady and found it impossible to walk in the dark without assistance. She did not complain of blurring of vision on head movement. Vestibular suppressant drugs did not relieve her disequilibrium.

She was first examined by one of the authors six months later. She could walk slowly with short steps and occasional side-steps in both directions. She turned slowly and preferred to have one hand on the wall or furniture for support. Examination revealed a mild spontaneous nystagmus to the left. Caloric tests with 5 ml of ice water injected separately against the tym-

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panic membrane via a 20-gauge needle over a period of 10 seconds with the head inclined posteriorly at an angle of 60°, with observation under Frenzel glasses in a darkened room, gave no response in either ear. She moved her head so slowly that it could not be ascertained whether she experienced oscillopsia. Audiometry showed a mild bilateral sensorineural hearing loss characterized by flat threshold patterns and speech discrimination scores of 100 per cent in both ears. Polytomographic radiologic studies of the temporal bones showed normal bony labyrinths, internal auditory canals, middle ears, and mastoid air cells systems. A diagnosis was made of disequilibrium caused by bilateral sequential vestibular neuritis.

Case 2

A 54-year-old man, while sitting at his desk at work, experienced a sudden severe attack of vertigo associated with nausea and vomiting, for which he was hospitalized. He had no auditory symptoms. Findings on all studies, including brain scan and electroencephalogram, were reported to be normal. No tests of vestibular function were done. He had been blind in the right eye since age 53, following three surgical procedures for detached retina. The vertigo subsided gradually over a period of several days but unsteadiness continued for two months, after which he felt quite well.

Three years later, at age 57, while driving his car, he had another similar episode of severe vertigo, nausea, and vomiting. He was examined in a hospital emergency department and released. He had no auditory symptoms. The vertigo gradually subsided over a period of several weeks; however, ataxia persisted and his vision was blurred with head movements. Ten months later, because of continuing symptoms of unsteadiness and oscillopsia, he was hospitalized for further diagnostic studies. Findings on computed tomographic studies of the temporal bones and brain, electroencephalogram, general neurologic examination, and posterior fossa angiogram were normal. Electronystagmography revealed a 5°/sec right-beating spontaneous nystagmus, asymmetric optokinetic nystagmus, and absence of caloric response in both ears. Both ears showed normal pure-tone thresholds and speech discrimination scores, negative tone decay (0.5 to 2 kHz), negative short increment sensitivity index (SISI) test results, normal tympanometry results, and normal acoustic reflexes. Acoustic reflex tone decay (0.5 kHz) was negative, and evoked-response audiometry results were negative for eighth-nerve and brain-stem disorder. Vestibular suppressant drugs were of no therapeutic value.

He was first examined by one of the authors two months later. He could walk without assistance but felt constantly unsteady and had to turn slowly to prevent falling. He complained of "multiple vision" whenever his head moved voluntarily or involuntarily. No spontaneous nystagmus was observed. There was no response to caloric testing in either ear with 5 ml of ice water injected against the tympanic membranes via a 20-gauge needle over a period of 10 seconds with the head inclined posteriorly at an angle of 60° with observation under Frenzel glasses in a darkened room. Previously recorded examination and test data were available, and no further tests were deemed necessary. A diagnosis was made of disequilibrium caused by bilateral sequential vestibular neuritis.

Three months later he was again hospitalized elsewhere because of continuing unsteadiness and blurring of vision on head movements. Results of electroencephalogram, audiometric studies, and CT scans were again normal. Results of neurologic examination were normal except for the previously observed vestibular disorder.

DISCUSSION

The occurrence of a single acute severe attack of vertigo without associated involvement of the auditory system or central nervous system (CNS) has been well documented. The disorder has been variously referred to as epidemic vertigo,[2] neurolabyrinthitis epidemica,[3] acute labyrinthitis,[4] vestibular paralysis,[5] and vestibular neuronitis.[6] On both clinical and pathologic grounds, the most probable etiology is viral infection. This premise is based on the fact that the disorder occurs as an isolated neuropathy, sometimes in association with influenzal-type diseases, and because of its histopathologic similarity to a reported case of herpes zoster oticus.[7]

In a 1981 study, Schuknecht and Kitamura[1] described three patients who experienced one or more episodes of vertigo sufficiently severe to induce nausea and vomiting and followed by protracted periods of disequilibrium. Two had no caloric responses in one ear, and one had a diminished caloric response in one ear. Postmortem histopathologic studies showed that each of the ears showing the caloric abnormality had atrophy of a branch of the vestibular nerve: the superior division in one, the branch to the lateral canal in one, and the branches to both

superior and lateral canals in one. None of the three ears showed evidence of vascular disease.

Acute vestibular neuritis has a sudden onset that may occur at any time of the day or night and is severe enough to cause nausea, vomiting, and sometimes diarrhea. In contrast to Meniere's disease, the vertigo is prolonged and subsides only gradually over a period of days or weeks, as would be expected for a disease causing death of vestibular neurons. Coates[6] defined vestibular neuritis with the following diagnostic criteria: 1) an acute, unilateral, peripheral, vestibular disorder without associated auditory or CNS manifestations; 2) a single episode of severe prolonged vertigo; 3) diminished caloric response in the involved ear; and 4) complete subsidence of the symptoms within six months.

In unilateral cases, the symptoms of vestibular imbalance (nystagmus, ataxia, tilting, and unsteadiness) gradually subside as CNS compensation for loss of vestibular function takes place. A subsequent similar attack involving the opposite vestibular system is quite another matter. The loss of vestibular function in the second ear results in permanent disequilibrium of varying degrees depending on the magnitude of vestibular neuronal loss, the age of the patient, and the functional integrity of vision, proprioceptive sense, and motor function. Severe bilateral deafferentation of the vestibular system can be expected to cause severe ataxia with slow and incomplete recovery and loss of the vestibulo-ocular reflexes with resulting oscillopsia.

The differential diagnosis includes Meniere's disease, vestibular schwannoma, fenestral fistula, cerebellar infarction, multiple sclerosis, and disequilibrium of aging.

Minimum evaluation includes otologic and neurologic examinations, audiometric evaluation, vestibular tests, and CT scans of the temporal bone and cerebellum.

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