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Vestibular Neuropathy Accompanying Auditory and Peripheral Neuropathies

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Objective: To define the incidence of measurable vestibular disorders in patients with auditory and peripheral neuropathies.

Design: Descriptive study of the case features of auditory neuropathy in 14 patients, 8 of whom had concomitant peripheral neuropathies.

Setting: University referral center.

Patients: Fourteen patients aged from 10 to 75 years and diagnosed as having auditory neuropathy, 8 of whom had concomitant peripheral neuropathies.

Main Outcome Measures: Incidence of abnormal vestibular caloric test results and the relationship of such incidence to clinical variables including the ages of the subjects, the presence of a concomitant periph-

eral neuropathy, vestibular symptoms, and audiological findings.

Results: Abnormal vestibular caloric test results occurred in 9 of the 14 patients. These 9 patients were on average older (35.6 years) than patients with normal caloric responses (17.8 years). Seven of the 9 patients with abnormal caloric responses had concomitant peripheral neuropathies compared with only 1 of the 5 patients with normal caloric responses. None of the 14 patients experienced symptoms of vestibular disorder.

Conclusions: Asymptomatic vestibular disorders are common in patients with auditory neuropathy when a peripheral neuropathy is also present. The reason for the abnormal vestibular test results is likely a neuropathy of the vestibular nerves.

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NEUROPATHIES of the vestibular nerves are not commonly recognized.¹ They are typically found in patients with generalized neuropathic disorders who develop symptoms of vestibular dysfunction.²⁻⁷ However, the incidence of asymptomatic vestibular neuropathies in patients with peripheral or cranial neuropathies is not known because vestibular testing is not usually performed for these patients. We examined vestibular functions in 14 patients with auditory neuropathy (and some with concomitant peripheral neuropathies) who did not express symptoms of vestibular disorder.

Auditory neuropathy is a term used to describe hearing loss in the presence of preserved cochlear outer hair cell functions (ie, otoacoustic emissions and cochlear microphonics) and severe abnormalities of auditory brainstem responses (ABRs) beginning with Wave I, the component generated by the cranial

nerve, or nerve VIII, within the cochlea.⁸ The hearing loss is of gradual onset and affects the ability to discriminate speech to a degree that is out of proportion to the pure tone loss. Starr et al⁸ describe 10 patients with auditory neuropathy, 3 of whom showed horizontal nystagmus on lateral gaze, and 2 others had absent responses to caloric vestibular tests (electronystagmography). These 5 patients were asymptomatic with regard to vestibular dysfunction (ie, no dizziness, unsteadiness worsened by darkness, or oscillopsia). All 5 patients had evidence of a peripheral neuropathy suggesting that the abnormal auditory and vestibular test results were part of a generalized neuropathic disorder affecting both components of cranial nerve VIII.⁷ In the present study, we report the incidence of abnormal peripheral vestibular test results in 14 other patients with auditory and peripheral neuropathies when symptoms of vestibular dysfunction were absent.

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SUBJECTS AND METHODS

The subjects were 14 patients ranging in age from 10 to 75 years and included 7 men and 7 women. Bilateral auditory neuropathies were identified in all subjects by severe abnormalities of ABRs beginning with Wave I (the component generated by cranial nerve VIII within the temporal bone) in the presence of normal cochlear function reflected by preserved otoacoustic emissions and/or cochlear microphonics.⁸ Eight subjects had concomitant peripheral neuropathies confirmed by clinical evaluation and abnormal nerve conduction velocity, and 4 subjects had abnormal sural nerve biopsy results. The characteristics of the subjects are described in the **Table**. Subjects were questioned regarding symptoms of vestibular dysfunction including dizziness, unsteadiness worsened by darkness, and instability of the visual environment. We did not use subjective assessment scales for vestibular impairment because of the paucity of subjective complaints consistent with vestibular disorder.

Vestibular function was measured using the following protocol. Electrodes were placed at the outer canthi of the eyes and a ground placed at the upper forehead (except for 1 subject with disconjugate eye movements who had electrodes placed around the left eye only). Calibration of the voltage change for a defined eye movement of 10° was measured by having patients move their fixation by saccades along a standard visual display of small dots placed at 10° intervals. As part of the calibration process, the saccades of each of the 14 subjects were tested by evaluating the rapid horizontal eye movement between 2 dots placed 20° apart. The results were studied for hypermetric or hypometric saccades, saccadic slowing, or other movement irregularities. Five of the initial 14 subjects studied (subjects 1, 8, 11, 12, and 14) were also tested with a central vestibular battery consisting of sinusoidal tracking, optokinetics, and gaze in addition to saccades. The results in each case were within the normal range. Because the other subjects also had no vestibular complaints, only the caloric tests were conducted for the remaining subjects. Caloric stimulation was provided through a closed-loop system with water irrigating through balloons placed in the external ear canal at temperatures of

30°C and 44°C for a duration of 45 seconds per incident. After irrigation, recordings were made for approximately 2 minutes while the subjects recited a list of states or common names as a mental diversion. The slow phase of nystagmus was measured in degrees per second, and the computations analyzing the strength of response of each ear and the preponderance of the responses were represented by the percentage of the total response. The criteria for abnormality included an asymmetry of response greater than 20%, a preponderance of response greater than 26%, or responses averaging less than 4° per second. Two subjects with absent ocular responses to caloric stimulation (subjects 1 and 2) were tested with ice-water irrigation. We did not use additional vestibular tests such as the sensory organization test or oscillatory head movements because these methods were not available at our institution.

Peripheral neuropathy was found in 8 patients based on clinical findings of absent ankle jerks, elevated thresholds to sensory testing, and abnormal nerve conduction study results. Four patients also had abnormal sural nerve biopsy findings. Abnormalities for nerve conduction studies were defined as sensory sural nerve conduction velocity of less than 40 meters per second; nerve action potential amplitudes of less than 8 µV; peroneal motor nerve conduction velocity of less than 40 meters per second; and/or compound motor potential amplitudes of less than 1 mV. The other 6 patients had normal clinical examination results for peripheral nerve function including preserved Achilles tendon reflexes and normal appreciation of a 128-Hz vibration tuning fork in the toes, which was supplemented by normal sural nerve conduction study results in 3 of these 6 patients. The audiological evaluation results for all patients met the criteria of auditory neuropathy with abnormal neural function evidenced by absent or abnormal ABR beginning with Wave I in the presence of preserved cochlear microphonics and/or otoacoustic emissions. Six patients had audiogram results with ascending thresholds; 4 had bilaterally severe hearing losses; 4 had mild-to-moderate losses; 1 (subject 12) had unilateral auditory neuropathy secondary to a viral infection; and 1 (subject 8) had hearing loss dependent on the temperature.⁹ Approval by the institutional review boards was obtained for the study of these patients with auditory neuropathy.

RESULTS

VESTIBULAR TESTS

Of the 14 patients tested (Table), 9 (subjects 1-9) had abnormal eye movement responses to caloric stimulation, and 5 had normal, symmetrical caloric responses (subjects 10-14). In the abnormal group, 5 had absent responses (subjects 1, 2, 4, 5, and 6), and 3 had asymmetrical responses (1 [subject 7] had an absent response in the right ear with reduced response in the left; 1 [subject 8] had left ear responses reduced with respect to the right; and 1 [subject 9] had a right-beating preponderance). Saccadic test results were within the normal range for all subjects. All results for the 5 subjects tested with the sinusoidal tracking test, the optokinetic test, and the gaze test were within normal range.

PERIPHERAL NERVE AND VESTIBULAR TESTS

The patients with vestibular dysfunctions had certain distinctive clinical features. Seven of the 9 patients with abnormal vestibular test results had peripheral neuropathies, whereas 4 of the 5 subjects with normal vestibular test results were without evidence of a peripheral neuropathy. The etiologies of the peripheral neuropathies included hereditary motor and sensory neuropathy in 5 patients, olivopontocerebellar atrophy in 1, spinocerebellar degeneration in 1, and an undefined etiology in 1. Sural nerve biopsy results were consistent with an axonal neuropathy in 3 patients and a combined axonal/demyelinating neuropathy in 1. The 5 patients with hereditary motor and sensory neuropathy belonged to 2 kindreds, one of which had a recessive inheritance due to a genetic disorder at 8q24;¹⁰ the other family had a

Characteristics of Subjects With Auditory Neuropathy*

Subject No./ Sex/Age, y	Caloric Test Results/Gait	Peripheral Neuropathy/ Family History/ Vestibular Symptoms	Pure-Tone Average, dB		Word Recognition, %	
			AD	AS	AD	AS
1/M/32	ABN/WNL	+/-/-	55	60	CNT	CNT
2/F/75	ABN/WNL	+/+/-	65	58	4	8
3/F/54	ABN/ABN	+/+/-	20	23	76	76
4/F/47	ABN/ABN	+/-/-	5	15	96	100
5/M/33	ABN/WNL	+/+/-	75	63	0	0
6/F/30	ABN/ABN	+/+/-	105	86	0	0
7/M/21	ABN/ABN	+/+/-	13	35	20	10
8/F/15	ABN/ABN	-/-/-	15	18	96	76
9/M/13	ABN/ABN	-/+/-	63	67	0	0
10/M/11	WNL/WNL	-/+/-	97	82	0	0
11/F/23	WNL/WNL	-/-/-	18	28	0	0
12/M/10	WNL/WNL	-/-/-	7	82	100	0
13/M/28	WNL/WNL	-/-/-	48	64	40	0
14/F/17	WNL/WNL	+/-/-	67	70	0	0

*ABN indicates abnormal; WNL, within normal limits; +, positive findings; -, negative findings; AD, right ear; AS, left ear; and CNT, could not test.

dominant mode of inheritance. Five of the 9 patients with abnormal vestibular test results had gait ataxia attributable to their peripheral neuropathies; whereas all 5 patients with normal vestibular test results had a normal gait. Of the 2 patients who had abnormal caloric responses without having peripheral neuropathies, 1 (subject 8) had a temperature-sensitive auditory neuropathy associated with a demyelinating disorder of the auditory nerve.⁹ The other subject (subject 9) had an isolated auditory neuropathy, and a younger sibling of subject 9 (subject 10) had an auditory neuropathy with normal vestibular test results.

Only 2 patients responded with any symptoms when asked about dizziness. One patient (subject 3) reported an occasional dizziness that was related to medications. The other patient (subject 14) reported a past imbalance. For both patients, their symptoms were judged to be nonvestibular in origin.

Patients with and without abnormal caloric test results could not be distinguished on the bases of their auditory functions (ie, pure tone average, speech recognition, ABRs, otoacoustic emissions, and cochlear microphonics) or sex. The patients with abnormal caloric responses were on average older (35.6 years) than patients with normal caloric responses (17.8 years).

COMMENT

The vestibular functions of 14 patients with auditory neuropathy were tested using electronystagmographic measures accompanying caloric stimulation. Vestibular abnormalities occurred in 9 of the 14 patients, 7 of whom had concomitant peripheral neuropathies. These results provide evidence that certain types of degenerative peripheral neuropathies are associated with a neuropathy of the vestibular nerves. A review of the literature revealed 3 examples of this type of association. Babin et al¹¹ reported temporal bone findings in a patient with neurosarcoidosis. The patient was deaf without vestibular complaints. There was evi-

dence of inflammatory demyelinating neuropathic changes of the vestibular, auditory, and facial nerves and a concomitant degeneration of the vestibular and cochlear sensory structures. Von Brevern et al¹⁷ described a patient with sarcoidosis and a generalized neuropathy who was deaf and had vertigo. The results of the vestibular testing were consistent with an ischemic neuropathy of the vestibular nerves. There was a bilateral auditory neuropathy manifested by preserved otoacoustic emissions generated by outer hair cells in the presence of bilaterally absent ABR and deafness.⁸ Frohman et al¹² described a patient with inflammatory demyelinating polyneuropathy with fluctuating findings of oscillopsia and ataxia that varied in extent with the patient's response to therapy. Bithermal calorics were abnormally reduced.

Our patients with vestibular and auditory neuropathies had few complaints that could be associated with their vestibular neuropathy. Rather, their symptoms of unsteadiness, weakness, or sensory change, when present, seemed related to their concomitant peripheral neuropathies. Patients with peripheral neuropathies often have gait ataxia that is considered sensory and not vestibular in nature. We suggest that the lack of vestibular symptomatology in our patients reflects both the bilateral distribution of the disorder and the slow rate of vestibular nerve degeneration, which were characteristics of their accompanying hereditary and degenerative peripheral neuropathies.

Those patients with auditory neuropathy without any evidence of a vestibular neuropathy were young. The mean age of those patients with abnormal calorics was 35.6 years, which was considerably older than for those with normal test results (17.8 years). This suggests that vestibular neuropathy in patients with auditory neuropathy is a late manifestation.

CONCLUSIONS

One of the difficulties in trying to define vestibular nerve involvements in peripheral and cranial neuropathies is

that, if the onset is gradual, vestibular symptoms are likely to be minimal. The major symptom that our patients expressed (gait imbalance) was attributed to their peripheral neuropathies, but it could also be affected by their vestibular disorders. Awareness of the presence of vestibular dysfunctions in patients with chronic peripheral neuropathies may allow therapeutic approaches directed at the vestibular disorder that would improve gait. We encourage vestibular testing in patients with generalized neuropathies to reveal any concomitant asymptomatic vestibular disorders.

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