Drop Attacks and Vertigo Secondary to a Non-Meniere Otologic Cause

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Background: Tumarkin falls are sudden drop-attack falls that occur in a subset of patients with Meniere syndrome (endolymphatic hydrops), an inner ear disorder characterized by vertigo spells and hearing loss.

Objective: To describe the clinical features and quantitative audiovestibular testing results in a case series of patients with Tumarkin falls, episodic vertigo, and normal hearing.

Setting: University referral center for disorders of balance and hearing.

Methods: Case series (unselected) of all patients with Tumarkin falls and a normal audiogram at least 1 year after onset of vestibular symptoms (n=6) from a retrospective analysis of the records of all patients with Tumarkin falls presenting to Neurotology Clinic at UCLA Medical Center, Los Angeles, Calif, from October 1, 1975, to February 1, 2001 (N=55). Quantitative audiologic and vestibular function testing, neurologic history, and examination were performed.

Results: Five of 6 patients had unilateral caloric paresis, and 1 had bilateral vestibulopathy. Five of 6 had a personal and/or family history of migraine headaches meeting International Headache Society criteria. All patients had a subjective sensation of feeling pushed by an external force, and half of the patients had a subjective tilt of the environment concurrent with the fall.

Conclusions: The incidence of migraine is high in this subgroup of patients with Tumarkin falls and normal hearing. The clinical description of the falls is similar to those associated with Meniere syndrome. Further studies are needed to understand the etiology of Tumarkin falls in these patients with normal hearing.

Arch Neurol. 2003;60:71-75

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tion with vestibulopathies other than Meniere syndrome and delayed endolymphatic hydrops, a variant of Meniere syndrome. We present 6 case reports of patients with sudden falls with characteristics of Tumarkin falls, recurrent spontaneous vertigo spells, and normal hearing. These patients clearly do not meet the diagnostic criteria for Meniere syndrome as defined in 1995 by the American Academy of Otolaryngology–Head and Neck Foundation.6

METHODS

From October 1, 1975, to February 1, 2001, 55 patients were seen at the Neurotology Clinic at UCLA Medical Center, Los Angeles, Calif, who met the clinical criteria for Tumarkin falls. The criteria for inclusion as Tumarkin falls were the report of a sudden fall that occurred without warning and without loss of consciousness, with no associated neurologic symptoms, normal results of a neurologic examination, and the clinical assessment of one of us (R.W.B.). All patients had undergone quantitative electronystagmographic testing with bithermal caloric testing and audiologic testing. All 55 medical charts of these patients were analyzed retrospectively, and of these, 47 patients had otologic symptoms of tinnitus, aural fullness, or fluctuation; 4 patients had no clinical symptoms of vertigo; and 1 met IHS criteria for acephalgic migraine with visual aura. Three of 6 met the IHS criteria for migraine headaches, with an average of 4.5 years. Three of 6 patients had Tumarkin falls as the initial presentation of vestibular problems. All patients reported the subjective sensation of being pushed by an external force. Three of 6 reported a sudden tilt of the environment concurrent with the fall. In patients 1 and 5, the room tilt was 90° toward the ipsilateral damaged ear, and in case 6 the room tilt was 90° vertically. No patients had auditory symptoms of tinnitus, ear fullness, or fluctuating hearing temporally related to the vertigo or falls, or in isolation.

REPORT OF CASES

Case 1

A 43-year-old woman (patient 3) reported a 2½-year history of sudden violent falls, characterized by the subjective sensation of being pushed to the left. There was no warning, no loss of consciousness, and no concomitant neurologic signs. In the several weeks before examination, she had 1 or 2 episodes per week. She also reported episodic spells of vertigo characterized by a sensation of swaying to the left, difficulty walking, and nausea lasting several minutes. She denied tinnitus, ear fullness, or fluctuating hearing in isolation or associated with the dysequilibrium or falls.

The patient's medical history was significant for a long-standing history since adolescence of bifrontal pulsating headaches associated with nausea lasting 1 day, without visual aura. She needed to lie down and sleep off these headaches. Family history was significant for a father who had severe migraine headaches and a 19-year-old daughter who had migraine headaches with visual aura.

Workup included magnetic resonance imaging of the brain and internal auditory canals with gadolinium, and auditory brainstem evoked potentials, the results of which were normal. Serial audiograms were normal. The electronystagmogram was significant for a 43% caloric paresis on the right side.

Case 2

A 37-year-old man (patient 6) presented with a history of recurrent spontaneous vertigo spells, lasting from a few seconds to half an hour, dating back 12 years. The vertigo spells were triggered by lack of sleep or skipping a meal. At the time of examination, he had recently had spells characterized by the subjective sensation of a sudden tilt of the environment vertically, lasting only seconds. On several occasions, he felt as if pushed from behind by an external force, and experienced a sudden fall. There was no warning, no loss of consciousness, and no concomitant neurologic symptoms. He denied tinnitus, ear fullness, or fluctuating hearing in isolation or associated with the vertigo or falls.

The patient had a history of sick headaches as a teenager, associated with photophobia and phonophobia. Family history was significant for a mother with episodic spontaneous vertigo spells. There was no known family history of migraine headaches. Workup included serial audiograms, which were normal. Quantitative vestibular testing was significant for a profound bilateral peripheral vestibular dysfunction.

RESULTS

There were 6 patients: 4 women and 2 men (Table 1). All patients had a history of episodic spontaneous vertigo, recurrent Tumarkin falls, and no otologic symptoms of tinnitus, aural fullness, or fluctuations in hearing. The average age at onset of vertigo spells was 40.6 years. The interval between the onset of vertigo spells and that of the Tumarkin falls ranged from at onset to 12 years, with an average of 4.5 years. Three of 6 patients had Tumarkin falls as the initial presentation of vestibular problems. All patients reported the subjective sensation of being pushed by an external force. Three of 6 reported a sudden tilt of the environment concurrent with the fall. In patients 1 and 5, the room tilt was 90° toward the ipsilateral damaged ear, and in case 6 the room tilt was 90° vertically. No patients had auditory symptoms of tinnitus, ear fullness, or fluctuating hearing temporally related to the vertigo or falls, or in isolation.

QUANTITATIVE AUDIOVESTIBULAR TESTING RESULTS

All patients had an audiogram at least 2.5 years after onset of vertigo spells. The range in interval between onset of vertigo spells and audiologic testing was 2½ to 14 years (Table 1). In all cases, serial audiograms disclosed normal hearing. In patients 1 through 5, quantitative vestibular testing (electronystagmography) showed a unilateral caloric paresis. Patient 6 had a profound bilateral peripheral vestibulopathy, manifested by an absent response at 0.05 Hz and a symmetrically decreased gain and increased phase lead at 0.2, 0.4, 0.8, and 1.25 Hz on sinusoidal rotational testing.

MIGRAINE HISTORY

In five (83.3%) of 6 patients, there was a personal and/or family history of migraine meeting IHS criteria (Table 2). Three of 6 met the IHS criteria for migraine headaches, and 1 met IHS criteria for acephalgic migraine with visual aura. Four of 6 patients had a family history of migraine headaches. In the patient without a personal or family history of migraine (patient 1), there was a history of headaches concurrent with vertigo, but the head-
syndrome presenting with Tumarkin falls at age greater than 50 years. All 6 of our patients with Tumarkin falls and normal hearing had a history of episodic spells of vertigo ranging from several seconds to several days. The small number of patients may reflect a true low incidence or the previous lack of recognition of the association of Tumarkin falls with migraine. Although selection bias can be an inherent confounder of any retrospective study, we tried to minimize that risk by the inclusion of all patients diagnosed as having Tumarkin falls and the unselected inclusion of all of these patients who had normal hearing.

The interval between vestibular symptoms and presentation to the Neurotology Clinic at UCLA Medical Center ranged from 2½ to 14 years. By definition for inclusion to the Neurotology Clinic at UCLA Medical Center, all of these patients had normal hearing. As a result of this selection bias, the results that suggest an association of Tumarkin falls and migraine in this study, during that period there were no otologic symptoms of fluctuating hearing, tinnitus, or aural fullness, and there was documentation of normal hearing on serial audiograms. The diagnostic criteria for definite Meniere disease, as proposed by the American Academy of Otolaryngology–Head and Neck Foundation, include (1) 2 or more episodes of vertigo of at least 20 minutes to 24 hours in duration; (2) audiometrically documented hearing loss; (3) tinnitus or aural fullness; and (4) exclusion of other causes. Most patients who go on to develop Meniere disease will have audiologic symptoms of tinnitus, aural fullness, or fluctuating hearing within 1 year of onset of symptoms. The 6 patients described in this study, therefore, do not meet the diagnosis of Meniere disease, and they are unlikely to later develop Meniere disease.

The clinical description of the falls in these patients is identical to that described by patients with Meniere disease. As a result of this selection bias, the results that suggest an association of Tumarkin falls and migraine in this study, during that period there were no otologic symptoms of fluctuating hearing, tinnitus, or aural fullness, and there was documentation of normal hearing on serial audiograms. The diagnostic criteria for definite Meniere disease, as proposed by the American Academy of Otolaryngology–Head and Neck Foundation, include (1) 2 or more episodes of vertigo of at least 20 minutes to 24 hours in duration; (2) audiometrically documented hearing loss; (3) tinnitus or aural fullness; and (4) exclusion of other causes. Most patients who go on to develop Meniere disease will have audiologic symptoms of tinnitus, aural fullness, or fluctuating hearing within 1 year of onset of symptoms. The 6 patients described in this study, therefore, do not meet the diagnosis of Meniere disease, and they are unlikely to later develop Meniere disease.

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than 65 years, Ishiyama et al10 reported a similar clinical presentation of Tumarkin falls. The factor that distinguishes the patients in the present study is the absence of hearing abnormality; in all previous studies, patients with Tumarkin falls had audiometrically documented hearing loss.

In this study, 3 of 6 patients reported a subjective room tilt in association with the fall. The direction of the room tilt was stereotyped for each individual patient and was in the direction opposite to the direction of the fall. The subjective room tilt has been associated with an ocular tilt reaction that can occur with otolithic stimulation3 and has been noted in some patients with Meniere syndrome-associated Tumarkin falls.3,4 There was not a temporal relationship between the vertigo spells and the Tumarkin falls; however, patients 1, 3, and 4 reported that the Tumarkin falls occurred in the setting of a recent flurry of episodic vertigo spells, both indicative of ongoing peripheral vestibular abnormality.

In these cases of Tumarkin falls without associated auditory symptoms or signs, there was an association with migraine in 5 of 6 cases (Table 2). An association between Meniere disease and migraine had been proposed by Meniere in 1861.9 The prevalence of migraine in patients with Meniere disease is about twice as high as that in the general population.10 The prevalence of migraine in the present case series was almost 3-fold higher than the highest estimates of the prevalence of migraine (5.3%-19% in males; 11%-28% in females).11 However, since migraine is so common, the combination of migraine, vestibular loss, and Tumarkin falls in these patients may have been a chance occurrence. In two thirds of our patients, the episodes of vertigo had features in common with migraine, including precipitation by alcohol, lack of sleep, emotional stress, and hormonal changes of menses. These triggers have been noted in Meniere disease as well. Thus, it is possible that the disorder in these patients represents an atypical variant of Meniere disease. Electrocochleography will be useful as an adjunctive measure to look for evidence of endolymphatic hydrops despite normal hearing.

Neurologic symptoms are common with migraine; episodic vertigo occurs in about one fourth of unselected patients with migraine.12 However, the definition of migraine-equivalent or migraine-associated vertigo remains unclear when there is not a clear temporal association between the headache or migraine aura and the vestibular symptoms, and most often the vertigo attacks occur during the headache-free interval.13

The association of audiovestibular disorders with migraine extends back to the 19th century.14 Migraine has been associated with benign paroxysmal vertigo of childhood15 and benign recurrent vertigo in adults, ie, episodic vertigo without hearing abnormality.13,14,16,17 Rassekh and Harker17 reported that 81% of patients with “vestibular Meniere’s,” recurrent attacks of vertigo without associated auditory symptoms, had migraine.

There is evidence that migraine is associated with peripheral vestibular end-organ damage. Cutrer and Baloh13 reported that 20.9% of patients meeting the criteria for migraine-associated dizziness had unilateral caloric paresis. Cass et al18 reported that 18% of patients with migraine-associated dizziness had unilateral caloric paresis. Ishiyama et al10 presented an association between benign paroxysmal positional vertigo (BPPV) and migraine as further evidence of peripheral vestibular damage associated with migraine. Patients with idiopathic BPPV were 3 times more likely to have migraine than patients with BPPV secondary to a known cause, and almost half of the patients with the onset of BPPV before 50 years of age met the diagnostic criteria for migraine.19 Childhood BPPV has also been linked to migraine.20 Benign paroxysmal positional vertigo is a well-documented sequela to ischemic damage of the inner ear, presumable due to release of otoconia from the macular membrane.21 Migraine has been proposed to cause sudden sensorineural hearing loss22-23 and monocular blindness,24 possibly via vasospasm of the small arteries of the ear and eye. Thus, it may be possible that migraine causes damage to the peripheral vestibular system via vasospasm of the small arteries of the inner ear.

The occurrence of Tumarkin falls in this subgroup of patients with vestibulopathy and vertigo hints that migraine-associated vertigo may be a phenomenon of the peripheral vestibular system in at least this subset of patients; however, further investigation in a larger study is required. In the population of patients presented in this study, all patients with Tumarkin falls without hearing abnormality had objective evidence and symptoms of peripheral vestibulopathy, ie, recurrent spontaneous vertigo spells and caloric paresis. In contrast, Dieterich and Brandt25 reported a low incidence of caloric paresis (8%) and a high incidence of electronystagmographic findings indicative of central abnormalities, in a retrospective study of 90 patients with migraine-associated vertigo. Thus, migraine-associated vertigo probably has heterogeneous causes. In the present study, given that the results of neurologic examination were normal and there was no evidence of central abnormalities on the electronystagmogram recording, a central cause for the sudden falls is unlikely. If the mechanism of the Tumarkin falls in our patients is the same as that of Tumarkin falls in Meniere syndrome, these falls originate from the vestibular periphery. Although we must use caution in establishing an etiologic link, we propose that the mechanism of migraine-associated vertigo and Tumarkin falls in these patients is a peripheral vestibular phenomenon. Calcium channel blockers, β-blockers, selective serotonin reuptake inhibitors, and acetazolamide have been used for the prevention of migraine-associated vertigo spells.26 Whether these agents are efficacious for the prevention of Tumarkin falls in these patients is unknown. We propose that for intractable Tumarkin falls with evidence for asymmetric vestibulopathy on testing, selective vestibular ablative procedures should be considered, ie, vestibular nerve section with hearing preservation.

Accepted for publication May 15, 2002.
REFERENCES