Adult aqueductal stenosis presenting with fluctuating hearing loss and vertigo

Report of two cases

ORHAN BARLAS, M.D., HÜSAMEDDIN GÖKAY, M.D., MİNA TURANTAN, M.D., AND NERMIN BAŞERER, M.D.

Departments of Neurosurgery and Otolaryngology, Istanbul Faculty of Medicine, Istanbul, Turkey

Two cases of aqueductal stenosis presenting with fluctuating hearing loss, tinnitus, and vertigo are presented. Audiovestibulometric assessment of both cases disclosed the characteristic pattern of disorder of the membranous inner ear. Non-tumoral aqueductal stenosis was demonstrated by computerized tomography in one case and by positive contrast ventriculography in the other. Shunting procedures of the cerebrospinal fluid resulted in resolution of inner ear dysfunction in both patients.

KEY WORDS □ aqueductal stenosis □ Ménière's syndrome □ endolymphatic hydrops □ hearing loss □ hydrocephalus □ vertigo

The constellation of episodic vertigo, tinnitus, and fluctuating hearing loss was described as a disorder of the inner ear in 1861 by Prosper Ménière.10 We report two patients who presented with these three classic symptoms. Audiovestibulometric assessment of these two cases were indicative of inner ear disorder. Further investigations revealed aqueductal stenosis, and shunting procedures of the cerebrospinal fluid (CSF) resulted in resolution of the symptoms of the membranous inner ear disease. Although the association of inner ear disease and hydrocephalus and increased intracranial pressure (ICP) have previously been described,4,19 a review of the literature shows that this clinical picture has not received its due attention.

Case Reports

Case 1

This 40-year-old man was transferred to the Department of Neurosurgery of Istanbul Faculty of Medicine for emergency insertion of a ventriculoatrial shunt on January 20, 1971.

History. The patient had been in good health until 1963, when the first episode of headache and right-sided hearing loss occurred. In the following years, he experienced similar episodes with increasing frequency. Four years before admission he suffered the onset of diplopia, and 2 years before admission tinnitus developed. Between the episodes he was free of all symptoms except for hearing loss, which increased with the onset of headache and diplopia. The first episode of vertigo occurred 5 months before admission. Repeated neurological examinations were negative. Psychometric tests disclosed impaired memory. Audiological tests revealed sensorineural hearing loss in the right ear. Recruitment test battery was positive. Threshold shift on the tone decay test of Corhart was negative. Vestibular testing showed bilateral horizontal rotatory spontaneous nystagmus on lateral gaze, with caloric inexcitability of the right ear. Ten days after a generalized convulsion he was admitted to a neurology clinic. Lumbar puncture performed there was followed by severe headache. Right carotid and left vertebral angiography, which was complicated by a left-sided hemiplegia, disclosed enlarged lateral ventricles and right-sided transtentorial herniation.

Admission. General examination showed the patient to be an athletic man with unremarkable physical findings. Neurological examination revealed signs of meningeal irritation, mild papilledema, right-sided hearing loss, and left-sided hemiparesis. Cranigrams showed an eroded dorsum sellae. Serological tests for venereal disease were negative. An electroencephalogram was normal. His level of consciousness progres-
sively deteriorated in the hours following admission. A ventriculoatrial shunt was inserted in an emergency procedure. Considerable regression of the clinical picture was observed after this measure. Exploration of the posterior fossa was carried out and revealed negative findings. The atrial catheter of the shunt was thought to be not functioning well, and was replaced in the cisterna magna during this procedure. Positive contrast ventriculography performed 12 days after the operation revealed occlusion of the aqueduct of Sylvius. He was discharged on March 24, 1971, and when last seen in 1975 he was symptom-free except for right-sided non-fluctuating hearing loss and increased deep tendon reflexes on the left.

Case 2

This 22-year-old man was seen in an ear, nose, and throat outpatient clinic for evaluation of headache, fluctuating hearing loss, tinnitus, and vertigo. He was referred to our neurosurgical clinic when a computerized tomography (CT) scan performed to rule out acoustic neurinoma showed triventricular dilatation.

History. Episodic tinnitus in the right ear had started in 1977. A year later, pain developed in the same ear, and tinnitus became constant with severe episodes. In 1979, left-sided tinnitus and fluctuating hearing loss developed. For the past 5 months he had bouts of mild bitemporal headache, pain in the left ear, and vertigo. Vertigo was frequent and severe, with constant disequilibrium between the attacks. On one occasion it had lasted for 30 minutes.

Admission. The patient was a high-school graduate who had to leave his job because of disabling vertigo. General physical examination was unremarkable. Neurological examination disclosed mild recent memory loss, nystagmus on left lateral gaze, and left-sided hearing loss. Cranigrams, x-ray films of the spine, and tomograms of the petrous pyramids were normal. Serology for venereal disease was negative. There were bilateral temporofrontal sharp waves in the electroencephalogram.

Audiological assessment disclosed a sensorineural hearing loss that amounted to approximately 50 dB in the speech frequencies. Recruitment test battery including short-increment sensitivity index test, alternate binaural loudness balance test, Langenbeck and Lüscher tests, and Metz test of impedance audiometry was positive, confirming the end-organ nature of the hearing loss. Threshold adaptation on the tone decay test of Corhart was negative. Tests to measure impedance of ipsilateral and contralateral acoustic reflexes were normal. Vestibulometric tests with electronystagmography showed horizontal rotatory spontaneous third-degree nystagmus on left lateral gaze and diminished caloric responses in the left ear. Hearing improvement of more than 15 dB was observed after ingestion of glycerol, confirming that hearing loss was fluctuating in nature.

Operation. A ventriculoperitoneal shunt was inserted in February 2, 1982. The CSF pressure was observed to be high during the procedure. Vertigo and headache did not recur during the 6 months after the operation. Postoperative audiograms consistently showed a 30-dB improvement over the best preoperative audiogram in the left ear, and electronystagmography disclosed marked improvement.

Discussion

These two patients presented with signs and symptoms of dysfunction of the membranous inner ear and hydrocephalus. Although histological criteria of inner ear disease in these patients are not available and no one test of inner ear dysfunction can localize the disease process within the sensorineural system, a battery of audiovestibulometric tests reliably indicates the locus of pathological change. Audiovestibulometric test batteries in our patients consistently revealed the characteristic pattern of dysfunction of the membranous labyrinth in both cases. However, decompression of hydrocephalus in Case 1 and CT scanning in Case 2 led to the diagnosis of hydrocephalus and insertion of CSF shunts. Dramatic resolution of fluctuating hearing loss, vertigo, and tinnitus after shunting procedures in both cases strongly suggested an association between aqueductal stenosis and membranous inner ear disease.

Both of the patients had slowly progressive hydrocephalus secondary to aqueductal stenosis. Non-tumoral stenosis of the aqueduct of Sylvius is a well defined cause of obstructive hydrocephalus. Congenital, developmental, inflammatory, infectious, and traumatic factors have been implicated in the etiology of aqueductal stenosis. Russell described four types of abnormality causing aqueductal stenosis: gliosis, forking, septum formation, and simple stenosis. Clinical presentation of aqueductal stenosis is markedly varied. Patients with aqueductal stenosis often have long histories of headache, gait disorders, epilepsy, endocrine dysfunction, or intellectual impairment before presenting with signs and symptoms of increased ICP.

In our patients, symptoms in periods of exacerbation consisted of headache, frequent vertigo, increased tinnitus and hearing loss, and, in Case 1, diplopia. A diagnosis was not reached in Case 1 before hydrocephalus decompensated, and had it not been for the CT scan, Case 2 would be considered to have had Ménière's disease. After shunting procedures, the patient in Case 2 made an almost complete recovery, whereas hearing loss persisted in the patient in Case 1, in whom treatment of hydrocephalus was instituted at an advanced stage of the disease.

The clinical findings of hearing loss, vertigo, and tinnitus have been observed in patients with adult aqueductal stenosis. Audiovestibulometric tests, reported in only two instances, either indicated a supranuclear lesion, or remained negative. Consequently, these symptoms were attributed to compres-
Aqueductal stenosis with auditory symptoms

sion of the brain stem and mesencephalon. On the other hand, inner ear dysfunction in patients with hydrocephalus secondary to basal arachnoiditis has been reported, although special audiovestibulometric tests were not available at the time of publication of this report.4

A pathological correlate of the inner ear disease resulting in fluctuating hearing loss, tinnitus, and vertigo is believed to be endolymphatic hydrops. First demonstrated by Hallpike and Cairns5 in 1938, endolymphatic hydrops results from inadequate reabsorption of the endolymphatic fluid. The endolymph is believed to be reabsorbed along the scala media by the stria vascularis, Reissers membrane, and the planum semilunatum, and at the saccus endolymphaticus which lies intracranially among the dural sheaths.78 Various etiological processes, such as a congenital anomaly, inflammation, vascular insufficiency, allergy, metabolic disorder, and acoustic neurinoma, result in inadequate reabsorption of endolymph.9 Wittmaack10 reported production of experimental endolymphatic hydrops in cats by repeated subarachnoidal injections of china ink, which also caused increased ICP. Endolymphatic hydrops has also been documented by Wittmaack10 in patients with increased ICP in whom he performed postmortem histological examinations of the labyrinths.

Relief of endolymphatic hydrops which followed shunting of CSF in our patients implies that lowering of ICP improved drainage of the endolymph. It may be speculated that in our patients decompensation of previously compromised endolymphatic reabsorption occurred with the onset of hydrocephalus, and resulted in endolymphatic hydrops.

References


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Address reprint requests to: Orhan Barlas, M.D., Istanbul Tip Fakültesi, Nöroşirüjji Kliniği, Çapa, Istanbul, Turkey.