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Tinnitus: Report of Ten Cases of Perilymphatic Fistula and/or Endolymphatic Hydrops Improved by Surgery

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Abstract: Presented are ten cases of patients with perilymphatic fistula and/or endolymphatic hydrops who had tinnitus as a major complaint. Tinnitus and its degree of severity often correlate closely with the state of health or hydrodynamic integrity of the inner ear, as these cases illustrate.

INTRODUCTION

When treating patients with perilymphatic fistula (PLF) and/or endolymphatic hydrops, vertigo and hearing loss (or fluctuation) are often the patient's main symptoms. The additional symptom of tinnitus is often a primary concern of the patient, but because it is less easily quantified the physician ranks it, along with the symptom of pressure or fullness, as lower in importance.

A review of early[1–3] and recent cases of PLF and/or endolymphatic hydrops encountered in my career suggests that tinnitus, and its various qualities of loudness and pitch, may indicate the state of health of the inner ear. The onset of tinnitus and its increase in intensity, frequency or character may precede a serious escalation in symptoms of vertigo, hearing loss and possibly a perception of aural pressure or pain. Alternatively, hearing improvement and resolution of vertigo often coincide with diminution or the disappearance of tinnitus. This may happen as the result of medical and/or surgical treatment. Medical therapy includes head rest, salt restriction and vestibular suppression. Surgical modalities include simple oval and/or round window reinforcement, endolymphatic shunt, cochlear aqueduct blockade and vestibular nerve section. When these means are effective tinnitus will often diminish, as illustrated by the following ten case reports.

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METHOD

Ten patients with Meniere's disease and/or PLF treated with surgery to control symptoms of disabling vertigo and hearing deterioration or fluctuation were selected from cases brought to surgery during the past 22 years. Each case was chosen because the symptom of tinnitus was a prominent complaint. Tinnitus varied in intensity according to the patient's primary symptoms.

The surgical modalities employed included simple exploratory tympanotomy with oval and/or round window reinforcement, endolymphatic shunt, cochlear aqueduct blockade and vestibular nerve section either by the posterior fossa or transmastoid route.

Each case is presented along with the patient's medical history, physical findings and appropriate laboratory, audiological and vestibular studies. The author's comments follow each case report. A general discussion and conclusion are provided.

CASE REPORTS

Case 1

This 45-year-old man was referred to the Dartmouth-Hitchcock Medical Center (DHMC) in July 1976 with a 7-year history and auditory findings of Meniere's disease. The patient had frequently occurring attacks of disabling vertigo, progressive deterioration in hearing (speech reception threshold [SRT] = 60 dB, speech discrimination score [SDS] = 68%), near-constant pressure and loud, low-pitched roaring

tinnitus. Medical management (low-salt diet and diuretics) failed to control his vertigo. He had no allergies and all blood chemistries were normal. An endolymphatic sac-subarachnoid shunt was performed in August 1976. For a short time the patient's vertigo stopped and pressure and tinnitus lessened significantly, but his vertiginous attacks resumed within 5 weeks. In October 1976 his ear was re-explored and the shunt revised. The patient was then asymptomatic for about one month before he experienced one of his most severe and disabling attacks of vertigo, nausea and vomiting. Because of the chronic severity of his problem and his relatively poor hearing, a translabyrinthine vestibular nerve section was performed in December 1976. This resulted in complete relief of this patient's vertigo, pressure and loud roaring tinnitus.

Comment

This operation was performed primarily to eliminate the patient's disabling vertigo, but his greatest sense of relief postoperatively was the absence of tinnitus. Pulec[4] performed 151 cochlear nerve sections for intractable tinnitus, which resulted in complete relief in 101 cases, significant improvement in 43 cases, and no improvement in 7 cases. In Pulec's series the following conditions were present: Meniere's disease (N = 84), chronic otitis media (N = 24), post-stapedectomy with inner ear injury (N = 11), trauma (N = 18), and a variety of other conditions (N = 14). All patients were believed to have tinnitus as the result of damage to the cochlea or cochlear nerve. Pulec[4] stresses the need of a thorough preoperative assessment, including hematological, audiological and radiological testing. A 5-mm segment of the nerve is removed medial to the spiral ganglion. The translabyrinthine approach was used in all but two patients who had a middle fossa approach. Pulec states that this approach is usually effective when the origin of the tinnitus is in the damaged cochlea.[4] Shulman[5] gives an excellent review of cochlear and/or vestibular nerve section for control of tinnitus.

Case 2

This 39-year-old woman presented to the DHMC in October 1974 with a 14-year history of Meniere's disease which had become bilateral. Her right ear was affected coincident with the birth of her first child, when she developed severe nausea, vomiting, vertigo and hearing loss. The symptoms subsided but returned coincident with the birth of her second child (12 years prior to her first visit to the DHMC), and again coincident with the birth of her third child 2 years later. She continued to have near-constant symptoms in the right ear. Her hearing deteriorated to where her pure tone average (PTA) was 75 dB (range 250 Hz to 2000 Hz) (SRT = 65 dB, SDS = 76%). She described the tinnitus as a fairly loud roar of "sea shell" quality.

Approximately four years prior to her first visit to DHMC, the patient began having similar symptoms involving her left ear. Hearing in that ear had deteriorated over four years to a level similar to that in the right ear (PTA = 70 dB, SRT = 60 dB, SDS = 88%). Because of a slightly improved PTA and SDS she regarded the left ear as her functional ear, and was the ear in which she wore a hearing aid. At the time of her presentation to the DHMC she was experiencing approximately three minor attacks of vertigo per week, preceded by an increase in tinnitus and a decrease in hearing in her left ear. Medical treatments had little success. Her allergic history was negative.

A left endolymphatic-subarachnoid shunt was performed in February 1975. Postoperatively, the patient's vertigo and roaring tinnitus in the left ear disappeared completely. Surprisingly, the tinnitus in her right ear greatly diminished for about 18 months. Her SRT improved to 42 dB and her SDS to 92%. Her greatest audiometric improvements were at 500 Hz, 1 kHz and 2 kHz, where thresholds went from 75 Hz, 60 dB and 50 dB to 60 Hz and 30 dB and 35 dB, respectively. With some mild fluctuations her improved thresholds were maintained for 8 years. After about 18 months, loud roaring tinnitus increased in the right ear and eventually became nearly constant. The decision to perform an endolymphatic shunt on the right ear was delayed because she had no vertigo, only a progressively increasing and roaring tinnitus.

In August 1983 a right endolymphatic-subarachnoid shunt was performed. Prior to performing the shunt the oval and round windows were observed for the presence of a possible PLF. A very obvious anterior oval window PLF was found. Before performing the shunt the oval window region was scarified and reinforced with areolar tissue. Postoperatively, the patient's tinnitus was nearly eliminated, and the occasional momentary positional vertigo disappeared. During the next two years the patient's SDS, which had never been above 68% during the previous nine years, improved to 92%.

In February 1985 the patient experienced renewed roaring tinnitus and diminished hearing in her left ear (the ear that had been operated on originally). A PLF was strongly suspected due to an obvious PLF found in the recently explored right ear.

The left ear was explored in March 1985. Upon opening the left middle ear, fluid was found within the oval window niche; the fluid accumulated repeatedly after aspiration. During a minor stapes manipulation the patient reported a sudden drop in her hearing. Both windows were reinforced and the case terminated. Postoperatively, the patient's hearing thresholds were greatly elevated and her SDS fell to 0%. Vertigo was eliminated and tinnitus stopped. Interestingly, at this point discrimination in the right ear rose to 92%.

Subsequently, debilitating vestibular symptomology from the right labyrinth recurred and was treated with streptomycin in December 1985 (24 grams over 14 days) and in October 1986 (20 grams over 10 days). On both occasions an increase in loud high-pitched tinnitus preceded the onset of vertigo, and on both occasions attacks of vertigo and the high-pitched tinnitus were controlled by courses of streptomycin. The patient did well until tragically killed in an automobile accident in August 1987.

Comment

The initial good surgical result in the patient's left ear most likely occurred because the cochlear hydrodynamic state was altered toward the normal by endolymphatic sac decompression, thereby permitting better overall cochlear physiology in the affected ear (and possibly allowed an unrecognized PLF to heal). The improvement of tinnitus in the nonoperated ear is interesting. The same finding has been reported recently by Ito and Sakakihara[6] in a study of tinnitus suppression in candidates for cochlear implants. They report a contralateral tinnitus suppression in 60% of their cases. Pulec[4] makes this same observation. This suggests that auditory pathways cross the brainstem to connect both ears, and that there is a pathologic connection of the efferent and afferent system of the auditory and vestibular nerves. These observations support a mechanism of centrally mediated tinnitus suppression. Efferent pathways traveling via the medial olivary-cochlear stria, and then to the outer hair cells by way of the vestibular nerve, most likely mediate some central control over the opposite cochlea. Likewise, discrimination eventually improved in our patient's right ear when the left-ear PLF was repaired and the left ear became a nonhearing ear.

The tinnitus in endolymphatic hydrops is likely due to the detachment of the tectorial membrane from the stereocilia to produce a low-pitched tinnitus.[7] As the overlying gelatinous tectorial membrane becomes decoupled from the hair cell, the Brownian Motion of the air particles in front of the tympanic membrane is detected. Harris[8] found that the noise level at the hair cell increased to +33 dB (in reference to auditory threshold) for loose coupling. Tondorf[7] concluded that the increased noise level at the hair cell input, an increment of 55 dB that is caused by the partial decoupling of the stereocilia from the tectorial membrane, ought to be perceived as tinnitus. In endolymphatic hydrops the decoupling is maximal in the region where the basilar membrane is the largest (i.e., the apical region). Blebs on the stereocilia and development of "giant" stereocilia have been demonstrated by Bredberg et al.[9] with the aid of scanning electron microscopy. A successful endolymphatic shunt most likely affects a partial return to the more normal anatomical and physiologic state, thus affecting tinnitus suppression. This seems to be the case when performing a shunt for both Meniere's disease or for endolymphatic hydrops secondary to the presence of PLF.[10] Interestingly, in our patient the tinnitus was also suppressed by streptomycin ablation.

This patient's Meniere's disease-like symptoms were most likely due to a PLF with secondary cochlear hydrops.[10] The strain of transmitting spinal fluid pressure through a patent cochlear aqueduct may have lead to her symptoms and her progressive cochlear-vestibular deterioration. This is one of three cases of PLF this author has seen that has been initiated or exacerbated by vaginal delivery (see Case 4).

Case 3

This 42-year-old women first presented to the DHMC in January 1984 with a 4-year history of dizzy spells and tinnitus in her left ear (Figure 1). The tinnitus varied in loudness and pitch. Her hearing began to fluctuate daily in early 1983. She had vertiginous episodes lasting for minutes to 1.5 hours, which were sometimes accompanied by nausea and vomiting. Initially her vertiginous episodes were not brought on by exercise, but were eventually exercise induced. On three occasions she was awakened by vertiginous episodes. Occasionally a loud noise could induce vertigo. She was allergic to some inhalants. Her cholesterol was 289. A computed tomography (CT) scan of her internal auditory meati was normal. An ENG showed bilateral hypofunction with a left spontaneous nystagmus. Fistula tests were negative. Her left ear PTA was 31.5 dB in January 1984 and 16.5 dB in January 1985. This improvement was attributed to a low-salt, low-cholesterol diet. Her tinnitus diminished along with the gain in hearing. She continued, however, to have some persistent unsteadiness which increased with any significant physical exercise. Her unsteadiness did not diminish with the administra-

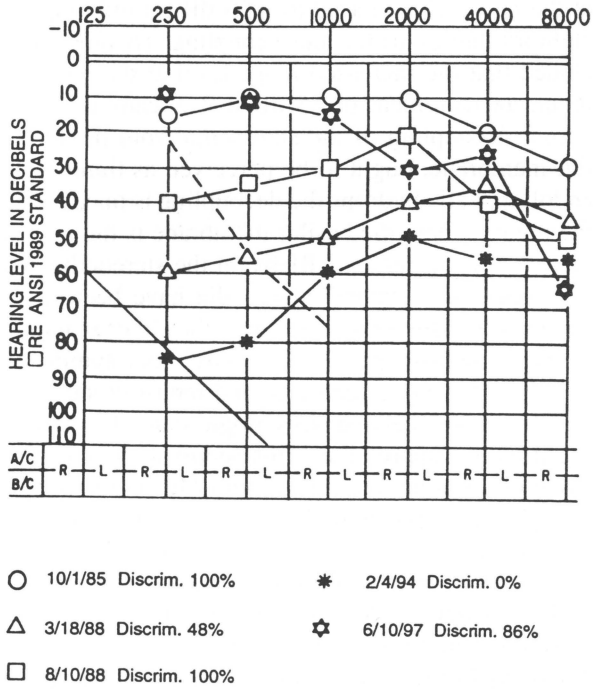

Figure 1. Serial audiograms for the patient in Case 3.

tion of 40 mg of intramuscular (IM) Lasix[1,2] or with glycerin.[11] Her balance was better after administration of 2 mg of Valium TID. As of July 1985 she felt normal about 3 to 4 days per week. About 1 year later (March 1986) she reported a slight increase in hearing loss and continued mild instability but no episodes of vertigo. In May 1986 a submucous resection and turbinate trim was performed to improve her airway. This permitted her to sleep better, and her daily periods of instability lasted only a few seconds and were less severe. Tinnitus was mild and high pitched. During the eyes-closed turning test of Singleton[12] she veered to the left. Any lifting or straining easily induced vertigo. A left PLF was suspected and repair suggested, but she declined.

She next presented to the DHMC in late February 1988, stating that 3 weeks prior she had experienced a sudden significant increase in tinnitus and decreased hearing in the left ear. This was accompanied with vertigo, nausea and vomiting lasting about one hour. Her SDS had decreased to 48% from 100% a year earlier. The PTA was now 55.5 dB. She described the tinnitus as white noise and sounding like water running from a faucet. Her instability had become constant and severe; the early morning hours were the most tolerable. In April 1988 an endolymphatic-mastoid shunt was performed (Arenberg type III sac) and both oval and round windows were reinforced with perichondrium. The stapes was felt to be hypermobile; the annular ligament was seemingly swollen, soft and almost gelatinous. A definite PLF could not be identified. After an initial difficult postoperative course she became nearly asymptomatic by the end of June (SDS = 100%, PTA = 32.5, tinnitus barely perceptible). Interestingly, this sudden improvement in all symptoms occurred within about 48 hours.

With occasional periods of mild stress-related instability, the patient did well for approximately one year. In early February 1989 her tinnitus (which became a constant mid-frequency "buzz") suddenly increased. Her hearing began to deteriorate and fluctuate, and tinnitus and imbalance recurred. In March 1989 her PTA was 60 dB with 0% discrimination. An exploratory tympanotomy was performed in April 1989. A recurrent oval window PLF was discovered (perilymph was clearly seen after repeated aspirations) after the debridement of adhesions with an argon laser. The stapes appeared hypermobile, with a thickened soft annular ligament. Repair resulted in dramatic relief of imbalance. Four weeks postoperatively the patient's hearing and discrimination again returned to near-normal levels within three days, coincident with a marked decline in tinnitus.

In May 1990 the patient's PTA was 28.5 dB with a 100% SDS. By February 1991 her PTA was 60 dB with 0% SDS. Vertigo was frequent and disabling, usually associated with physical stress. Tinnitus was loud and "sea shell" in quality. Due to a suspected patent cochlear aqueduct, a lumbar puncture was performed and approximately 10 cc of fluid was drained. Opening pressure was 180 mm of water. The patient had immediate relief of tinnitus and imbalance which lasted approximately 36 hours. Interestingly, all her symptoms subsided and remained quiescent. By 1994, her vertigo had become less disabling. A quieter mid-frequency buzzing sound persisted.

In April 1997 the patient's tinnitus became much louder and higher in pitch, and her hearing and balance deteriorated; this persisted through the end of May. In early June 1997 her tinnitus diminished suddenly to 10% of what it had been. Her balance normalized. Her hearing improved to near-normal levels (Figure 1), and SDS increased from 0% to 86%. In mid-June 1997, an audiogram and transtympanic electrocochleogram (EcoG) were performed. The patient's EcoG demonstrated a normal summating potential (SP)-action potential (AP) ratio of 30%. On several occasions she could raise her SP to 45% to 50% of the AP by doing a Valsalva maneuver (similar to the patient in Case 6).

Comment

This patient's tinnitus was more severe when cochlear vestibular disorganization was greatest—during periods of imbalance or diminished hearing. Her tinnitus was least severe after her second PLF repair, when her hearing was nearly normal and imbalance episodes nearly nonexistent. The same is true now that she has suddenly normalized (June 1997). Distortion of cochlear membranes due to diminished perilymph pressure and secondary endolymphatic hydrops most likely leads to hair cell dysfunction (secondary to hair cell-tectorial membrane decoupling[7]) and tinnitus.

This patient likely possesses a patent cochlear aqueduct, as her symptoms of instability, tinnitus and hearing loss were eased by drawing off 10 cc of spinal fluid, which could, in turn, lower perilymph pressure and thus diminish any inner-ear fluid flow responsible for causing vertigo.[13] The fact that she can increase her SP by a Valsalva maneuver may also signify an open cochlear aqueduct. An abnormally patent cochlear aqueduct is the probable cause for ease and frequency of relapse in such cases.

Case 4

This 32-year-old woman presented to the DHMC in February 1983. Five years previously, at age 27, she began to experience episodic vertigo, fluctuating hearing loss, loud sea-shell-quality tinnitus and fullness in her left ear. Symptoms subsided for 4 months during her third pregnancy at age 31, but resumed immediately coincident with her delivery. Initially she was diagnosed with Meniere's disease, but later was thought to have a PLF with secondary endolymphatic hydrops. Her PTA was 46 dB and her SDS 92% in her left ear. The left ear was explored in May 1983 and an oval window PLF was found and repaired. The patient had immediate resolution of imbalance, pressure and tinnitus. Ten days later her symptoms returned coincident with a shopping trip. An endolymphatic-mastoid shunt was performed in May 1983 with repeat PLF repair. Again she was free of imbalance and her tinnitus diminished greatly. Three months later (August 1983) most of her symptoms recurred coincident with heavy lifting. A third PLF repair was performed in November 1983, with resolution of vertigo and tinnitus. Vertigo and tinnitus, described as a mid- to high-frequency buzzing, recurred in 1989. A repeat PLF repair in April 1989 resulted in a temporary improvement in hearing and diminished tinnitus; however, three weeks later minor lifting brought about a dead ear and severe imbalance. In May 1990 a posterior vestibular nerve section, including some of the cochlear fibers, has given her long-lasting relief from vertigo and tinnitus.

Comment

As with the patient in Case 3, tinnitus volume was worse in this patient when cochlear-vestibular dysfunction was greatest. As in Case 1, the vestibular nerve section, which included about 10% of the cochlear division of the eighth-nerve, has given permanent relief from tinnitus and vertigo. Brackman and Barrs[14] report on vestibular nerve section alone resulting in resolution of tinnitus in 49% of their patients. They had a 61% success rate in controlling tinnitus when both the cochlear and vestibular nerves were sectioned (see Shulman[2] for a review of this subject).

Case 5

This 48-year-old man presented to the DHMC in March 1984. Ten years previously he suffered a closed head injury when he was hit on head with a suspended swinging bucket, which resulted in 5 minutes of unconsciousness. He had three months of rest at home due to dizziness, which eventually resolved, though his balance was never quite normal. Seven years later, while dancing, the patient became warm, clammy and nauseated. Vertigo soon followed and lasted for five hours. Upon awakening the following morning, the patient experienced loud tinnitus (medium-pitched ringing) and hearing loss in his right ear. From this point, the hearing in his right ear deteriorated, although he had some periods of quite good hearing (his last symmetrical hearing test was in 1981). The patient's tinnitus became loud, pulsatile and constant. He experienced frequent attacks of vertigo brought on by lifting or straining, which lasted for seconds to hours. He had an occasional episode of vertigo without apparent cause, and occasionally could perform heavy lifting without provoking vertigo. His audiogram was up-sloping through 2000 Hz (SRT = 56, SDS = 66%), with 4 kHz and 8 kHz depressed secondary to noise exposure. A glycerin test resulted in a 5-dB improvement in PTA and SDS improved to 86%. Carotid pressure slightly diminished the intensity of the tinnitus. An observer could not hear the pulsation or record them by tympanometry. Presenting symptoms allowed for the possibility of both hydrops and PLF. He underwent an exploratory tympanotomy in May 1984, and clear fluid seepage from the anterior end of the oval window was found. The window was reinforced with areolar tissue and an endolymphatic-mastoid shunt was performed. Postoperatively, the patient's hear-

ing improved dramatically (PTA = 37.5, SRT = 30, SDS = 88%) and tinnitus diminished to nearly non-detectable levels. A 6-year follow-up visit revealed an SRT of 40 dB and an SDS of 84%. Tinnitus remained at a very low level.

Comment

In cases of endolymphatic hydrops, whether or not combined with a PLF, a significant improvement in hearing and decline in vertigo and tinnitus most likely indicates a partial restoration of cochlear-vestibular integrity. Tinnitus intensity seems to be a marker for declining or improving cochlear-vestibular health. In this case the pulsatile character of the tinnitus may reflect vascular pulsation being transferred from the cerebrospinal fluid (CSF) through a patent cochlear aqueduct. Sismanis[15] reports pulsatile tinnitus in patients with benign intracranial hypertension which improves with treatment. Marchbanks[16] reports similar findings but only when the cochlear aqueduct is open.

Case 6

This 56-year-old man presented to the DHMC nine years after a stapedectomy for otosclerosis, resulting in a dead ear. His SRT was 65 and his SDS was 92% in his left and only-hearing ear. He was placed on fluoride to delay further hearing deterioration in this ear. During the next four years his hearing remained stable and the arthritis in his left hip resolved dramatically. However, the patient complained of constant, loud tinnitus and an occasional sensation of hearing in the dead ear. Because of this and our difficulty performing accurate audiology with appropriate masking at such levels, the middle ear was explored in February 1980. The oval window was partly occluded by a small granuloma resting in what appeared to be gelatinous material. This was easily removed, and revealed a wide open oval window with an intact, subluxated footplate in the depths of the vestibule. A slow but constant flow of perilymph came from the window. The oval window was covered with a piece of perichondrium and held in position with a House wire attached to the incus. The patient's hearing did not change, but his tinnitus resolved completely. Interestingly, his balance was never a problem.

Comment

Such positive results are rare. Perhaps stopping the constant flow of fluid (CSF or perilymph) through the cochlea permitted partial restoration of cochlear integrity with a secondary decline of the tinnitus. The tinnitus in this case must have originated in the cochlea.

Case 7

This 22-year-old man, who was a U.S. Olympic team cross-country skier, presented to the DHMC in September of 1985 with several months history of exercise-induced decreased and distorted hearing (Figure 2). He described his hearing distortion as sounding tinny, with a background metallic hum. The hearing distortion and increase in loudness of his tinnitus was brought on specifically during exertion while roller skiing. He experienced no vertigo. He was able to reproduce his hearing distortion in the clinic by doing a series of pushups and running up and down the stairs several times. After about 15 minutes of rest his tinnitus would subside and the low-frequency hearing loss would return to near-normal levels. Only 125 Hz, 250 Hz, 500 Hz and 1000 Hz were affected at this time, dropping to 30 dB, 25 dB, 20 dB and 15 dB, respectively. In November 1985 a large, free-flowing oval window PLF was found and repaired, which temporarily improved the patient's hearing and reduced his tinnitus. Postoperatively the patient rested only two weeks and returned to his skiing in attempt to make the Olympic team. After he stopped training his low frequencies im-

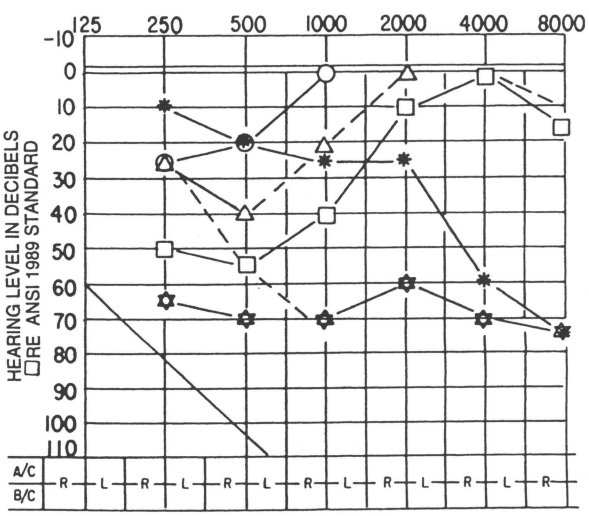

Figure 2. Serial audiograms for the patient in Case 7.

proved and the high frequencies diminished, resulting in the sensation of a mild hearing depression with good discrimination. In July 1991 his PTA was 20 dB with an SDS of 100%. Tinnitus during this time was low intensity and described as a mid- to high-frequency pure tone. He routinely used the ear for speaking on the telephone.

During the summer of 1996 (11 years after his initial problem began) the patient experienced a sudden noticeable hearing loss in the left ear, followed closely by an increase in intensity and frequency of tinnitus (still a pure tone). His PTA dropped to 70 dB with 0% SDS. A repeat middle-ear exploration was performed in August 1996. After lysis of adhesions in the oval window region an obvious PLF could not be identified with certainty. The round window remained completely sealed from the previous surgery. The oval window was reinforced, but this did not improve the patient's hearing or reduce tinnitus. His SDS rose to 68% without improvement in pure tones. His tinnitus now varies in intensity and becomes louder with physical exercise or when around loud noise. Occasionally it will stop completely for a few seconds.

Comment

This patient falls into the small percentage of patients with PLFs who do not exhibit vertigo, but rather downward fluctuations of hearing with increased tinnitus secondary to physical stress.[1,2] Proper postoperative counseling for these patients is an ongoing problem, since it is difficult to determine how much physical activity they can tolerate. In such patients the perilymph flow through the inner ear occurs in such a way as to be detrimental to hearing but not balance. A widely patent cochlear aqueduct is likely present in such patients.

Case 8

This 34-year-old woman presented to the DHMC in May 1996 with a 2-month history of the following: increased vertigo with lifting, bending or straining; intermittent high-pitched tinnitus; and intermittent right-ear pain lasting about 10 minutes and recurring five to six times per day. Seven years prior, after lifting a patient from the floor to a wheel chair, she began experiencing almost constant vertigo, which was exacerbated by exercise. Her vertigo continued until she was seen by Fitzgerald[17] who diagnosed and repaired a right oval window PLF. Following this treatment her symptoms of vertigo diminished by over 50%, and she was able to function well in her job. Her tinnitus diminished in intensity and was noticed only occasionally.

In 1996, her first symptom was increased right-ear pain,[1,2,11] followed by a marked increase in vertigo and return of tinnitus. The tinnitus was still intermittent but often present. On physical exam at the DHMC, the patient was found to have a positive subjective fistula test bilaterally that was stronger on the left. She veered to the left during her standing Quix test, high-stepping Quix,[11] ECTT,[12] and eyes-closed walking test. Her audiogram looked grossly normal with 100% discrimination bilaterally, but she had a mild symmetrical low-tone sensorineural hearing loss and a mild asymmetrical high-tone loss on the right. The SRT was 5 dB on the left and 15 dB on the right. Nearly three weeks later neurologic testing was repeated and she was given 40 gm of IM Lasix.[1,2] One hour later she was nearly asymptomatic and was completely normal on her Quix tests and eyes-closed walking test. Her tinnitus also diminished with the Lasix.[1,2] Because of her continuing tendency to drift to the left, and a mildly positive fistula test on the left but with symptoms of pressure, fullness, pain and tinnitus on the right, we suggested bilateral middle-ear explorations to repair suspected PLFs.

In August 1996 bilateral middle-ear explorations with reinforcement of oval and round window areas were performed. The left ear was explored first. A very obvious oval window fistula was seen; fluid welled up repeatedly in the oval window niche after suctioning. Both windows were reinforced with areolar tissue. Exploration of the right ear revealed many adhesions from the previous repair. Despite using the KTP laser to lyse the adhesions, a fistula could not be identified clearly.

Postoperatively the patient experienced nearly complete resolution of vertigo, pressure and tinnitus. Relief lasted almost four weeks, when, a half hour after awakening one morning, she felt unstable and experienced pressure in the right ear. A fistula test was subjectively negative on the left side but positive on the right. She still veered to the left on the eyes-closed walking test. Again, 40 mg of Lasix caused her symptoms to resolve within one hour (neurological exam results reverted to normal and right-ear tinnitus diminished). She was placed on Lasix (20 mg BID) and a low-salt diet, which did not improve her symptoms.

In September 1996 her symptoms were unchanged. She veered left on a Quix test[11] and eyes-closed walking test.[13] She complained of intermittent pain in both ears and fullness in the right ear. Hearing remained normal (unchanged). High-pitched tinnitus was often present.

In November 1996 platform posturography was performed using a Smart Balance-Master. The complete set of tests were performed before and 45 minutes after the administration of Lasix (40 mg IM). All tasks improved after Lasix. Before receiving the Lasix she could hold a tandem Romberg for only three seconds; after the Lasix was given she could hold it for 20 seconds. When asked to print in a vertical line from top to the bottom of a page with her eyes closed, the line of text curved to the right; after the Lasix she could print in a straight vertical line.

An EcoG was normal on the left side with an SP-AP ratio of 36%. On the right side the SP-AP ratio was 40% (Figure 3). Upon doing a Valsalva maneuver when supine the patient immediately raised her SP-AP ratio to 80%. Upon stopping the Valsalva maneuver the ratio returned immediately to 40%. This was repeated four times with identical results. A patent cochlear aqueduct (or some other open connection between CSF and perilymph) was felt to be necessary to permit this repeatable phenomenon.

A right endolymphatic shunt was performed in December 1996 using an Arenberg valve. Additional areolar tissue was placed around the stapes footplate, as the previously placed tissue had come away from the anterior end of the stapes. Postoperatively the patient has done remarkably well. She is completely free of vertigo for the first time in seven years. Hearing has remained unchanged and her tinnitus is nearly gone.

Comment

Performing an endolymphatic shunt can improve or eliminate the symptoms of a PLF.[1,2,18,19] The fact that doing a Valsalva during an EcoG could immediately and repeatedly change a summating potential from 40% to 80% seems to indicate a wide open CSF-perilymph connection, most likely the cochlear aqueduct. The wide open cochlear aqueduct most likely made our patient prone to developing a PLF and vulnerable to recurrence after the initial repair. The fact that the tinnitus disappeared completely after the endolymphatic shunt may indicate the restoration of a more normal cochlear physiology and anatomy, perhaps adding necessary compliance to the system.

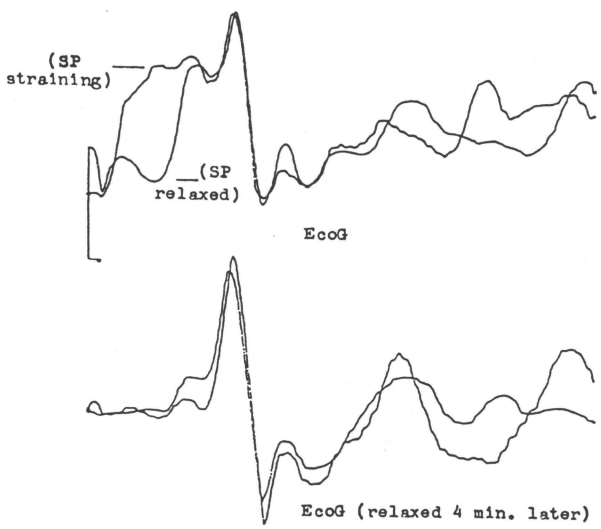

Figure 3. Transtympanic electrocochleography for the patient in Case 8, demonstrating the increase in the summating potential that occurred repeatedly with Valsalva maneuvers.

Case 9

This 7-year-old girl first presented to the DHMC in August 1987 with a 2-year history of recurring stomach aches and nausea (often causing a panic attack), and a left-ear low-tone hearing loss (threshold of 45 dB, 45 dB and 25 dB at 250 Hz, 500 Hz and 1000 Hz, respectively) (Figure 4). The otologic exam was normal. A tonsillectomy and adenoidectomy was performed in December 1987 to improve her airway and decrease the frequency of recurring otitis media with effusion. Over the next four years she experienced episodes of true vertigo which caused her to fall occasionally. Hearing in the left ear continued to fluctuate as much as 20 dB between 250 Hz and 2000 Hz, but she always had an excellent SDS.

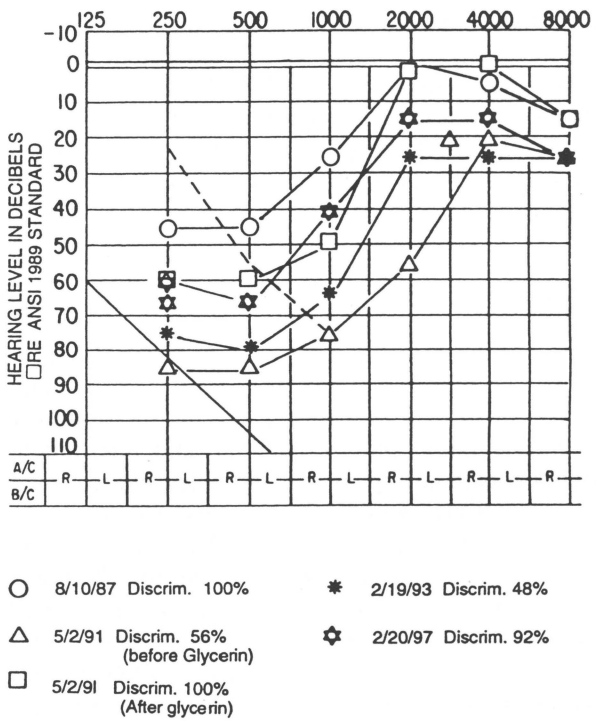

Figure 4. Serial audiograms for the patient in Case 9.

In May 1991 she presented with greatly elevated thresholds and a SDS of 56% which returned to the previous baseline after receiving glycerin. She described her tinnitus as a high-pitched ring. She experienced vertigo with significant exercise; several bouts of severe vertigo caused her to fall to the floor. In September 1991 an oval window PLF was found and repaired. Postoperatively the patient's hearing returned to its previous best level (PTA = 42.5, SDS = 100%). At 2 Hz, 4 Hz and 8000 Hz she was at 0 dB, 10 dB and 20 dB, respectively (SRT = 25 dB). She maintained good hearing and balance with occasional fluctuations in both entities for about four years, during which time she participated in sports and horseback riding. In January 1994 she reported near absence of tinnitus. In January 1995 she experienced return of high-pitched tinnitus and vertigo with exercise. Her school performance dropped markedly. She stated that in retrospect during the 1994 soccer season she had been a "little dizzy" about half of the time when playing.

In October 1995 a lumbar puncture revealed a mildly elevated spinal fluid pressure of 210 cc of water. In November 1995, following the placement of a lumbar drain, the left ear was again explored. Lysis of oval window adhesions was carried out using a KTP laser. No definite PLF could be identified. Both windows were reinforced with areolar tissue. The lumbar drain was left in place for five days. Her hearing again improved, tinnitus diminished and balance normalized. Three months later when cleaning out her horse's stall her symptoms returned in full force. In July 1996 a left endolymphatic-mastoid shunt was performed and the oval window and round window were again reinforced. A lumbar drain was not used, as her preoperative spinal fluid pressure was normal (165 cc of water). Within four weeks she was feeling 70% better than she had before her surgery, and has remained so as of her last follow-up visit in March 1997. She was permitted to go to boarding school with a limited physical education program. She has done well academically thus far. Vertigo is usually absent but occurs occasionally in mild form, almost always in relationship to exercise. In February 1997 tinnitus was intermittent and occasionally loud enough to prevent concentration. Diuretics and salt restriction have not modified this symptom. Her hearing is surprisingly good (only 15 dB below the 1987 level, SDS = 100%).

Comment

This is a case in progress. Eventually a widely patent cochlear aqueduct may need to be plugged,[3] and/or the vestibular nerve will have to be cut to allow the patient to live a normal life that includes sports and normal physical activities. Her problems probably began in 1985 as a 5-year-old when she presented with sudden onset stomach aches and nausea and a panic-like state. The author has observed this phenomena in other patients[13] who eventually developed PLF but as young children were too young to articulate the problem clearly. We adults did not ask the right questions or perform the right tests (i.e., audiogram and routine Romberg type tests). Accurate tests for cochlear aqueduct patency are needed for cases such as this.

Case 10

In January 1995 a 64-year-old farmer and general maintenance man experienced sudden increased tinnitus in his left ear. He described the tinnitus as having a machinery-like hum. Commensurate with the onset of his tinnitus he felt mildly off balance, with a definite increase in vertigo with physical activity, especially lifting. He also felt that the hearing in his left ear was slightly diminished. The following month his tinnitus increased and hearing decreased as he struck a nail with a hammer. This was followed by nausea and vertigo within minutes. He feels that his hearing never improved, and fluctuated noticeably, since this event. When first seen by an otolaryngologist his left ear SRT and PTA were 30 dB with a SDS of 84%.

He was explored for a PLF in June of 1995. An oval window PLF was thought to be present, but its reinforcement did not improve his symptoms. The patient was referred to the DHMC in August 1995. Audiology had not changed. He veered left on his eyes-closed walking test. One hour after taking 40 mg of IM Lasix the patient's vestibular symptoms disappeared and his eyes-closed walking test was normal. This good effect of the Lasix lasted about 16 hours. Interestingly, the motor-like tinnitus in his left ear also diminished but did not disappear.

In November 1995 the patient underwent an exploratory tympanotomy with revision reinforcement of the oval window and round window. Just prior to administering general anesthesia a lumbar puncture was performed to see if pseudotumor cerebri might be present. Spinal fluid pressure was 270 mm of water (more than 200 mm of water is considered abnormally high). Therefore, a lumbar drain was inserted and left in place for a total of four days after the surgery in order to facilitate healing. Postoperatively, the patient's vertigo stopped completely and his tinnitus diminished. This resolution of symptoms lasted six weeks. Feeling fine, the patient returned to work.

After lifting an air conditioner both his tinnitus and vertigo returned to preoperative levels. As before, any exertion worsened the vertigo and brought on nausea. Despite his problems he continued to work, though at a suboptimal level.

He continued to experience CSF hypertension with absence of ophthalmic signs. In May 1996 his pure tone audiogram demonstrated some random changes (generally decreasing in threshold by 5 to 10 dB) between 125 and 2000 Hz when he assumed a Trendelenburg position. Tinnitus also diminished in this position. The author felt these entities indicated cochlear aqueduct patency and suggested cochlear aqueduct blockade to the patient. This procedure had been performed three times previously[3] with no negative affects to hearing and with diminution of vertigo. This procedure was performed in July 1996. Following the surgery the patient's hearing remained stable between 125 Hz and 1000 HZ with a 15-dB to 20-dB decline above 1000 Hz. Discrimination was 64% preoperatively and declined to 44% postoperatively. His tinnitus declined immediately to nearly negligible levels and has remained so to May 1997.

Interestingly, coincident with the surgery the patient experienced a temporary hearing decline of 20 dB in all frequencies in his right (opposite) ear and some ataxia. The right ear hearing returned to baseline within three weeks and his ataxia diminished (he veered right on the eyes-closed walking test, whereas before surgery he veered left). As before the initial PLF repair, IM Lasix resulted in complete resolution of his vertigo. This leads me to suspect a hydrodynamic problem (perhaps a PLF) in the right ear. A ventriculo-peritoneal shunt was performed in September 1996 to relieve the patient's CSF hypertension and to hopefully resolve the ataxia. The patient's CSF pressure normalized, but mild ataxia remained. In March 1997, diagnosis and treatment of hypothyroidism lead to complete resolution of the residual ataxia.

Comment

Sismanis[15] has discussed the symptoms of tinnitus, vertigo and fluctuating hearing loss in patients with idiopathic CSF hypertension. Recently, Marchbanks[21] studied a group of patients with CSF hypertension using the tympanic membrane displacement technique to determine cochlear aqueduct patency. He found that patients with CSF over pressure who did not have patent cochlear aqueducts also did not develop the symptoms of tinnitus, vertigo, fluctuating hearing and pressure, while the patients who had patent cochlear aqueducts did develop these symptoms. Effecting closure in this patient stopped the tinnitus.

DISCUSSION

All ten cases presented unique diagnostic and therapeutic challenges. The patients in seven of the ten cases (Cases 2-4 and 7–10) had recurrent PLFs requiring multiple middle-ear explorations for repair. In five of these seven patients an endolymphatic shunt provided significant long-term or permanent relief of symptoms, including tinnitus. One of these seven patients (Case 4) chose vestibular nerve section for a final solution to control tinnitus and vertigo. In all seven cases tinnitus was most severe when vertigo and hearing were most problematic. Also in these seven cases when vertigo was controlled or absent and hearing improved, tinnitus diminished or became absent. The patients in Cases 7 and 9 still experience difficulty and their tinnitus continues to be troublesome and high pitched.

These seven patients, and the patient in Case 8, most likely had or have patent cochlear aqueducts.[21,22] The pulsatile nature of tinnitus in Case 5 points strongly to this possibility, as does the ability of the patients in Cases 3 and 8 to repeatedly increase their SPs during transtympanic EcoG by performing successive Valsalva maneuvers.

The patient in Case 1 is the only patient who seemed to present with a "pure" Meniere's disease picture. Even in this case an endolymphatic shunt gave temporary relief of symptoms, including tinnitus. A translabyrinthine total 8th-nerve section provided immediate and total relief from tinnitus as well as vertigo.[4] Similarly, a suboccipital vestibular nerve section provided the patient in Case 4 with immediate and complete relief from both tinnitus and vertigo.[14] Therefore, the tinnitus must have emanated from cochlea as opposed to central pathology, and the tinnitus-mediating fibers from the cochlea must often travel with the vestibular nerves.

Case 6 is presented because it is unique. Preoperatively, tinnitus was not discussed in depth. The wide open oval window was unexpected. What was probably a slow free-flow of CSF through the scala tympani, vestibule and out into the middle ear was stopped by the perichondrial graft held in position by the House wire. As the flow of fluid through the inner ear ceased, the tinnitus ceased as well.

In patients previously reported with PLF, tinnitus[1,2] seemed most common in patients with Meniere's disease-like symptoms (11 of 18). In the "miscellaneous end-organ" group tinnitus occurred in 4 of 12 patients. No patient complained of tinnitus in the

"hearing loss without vertigo" group, although the patient in Case 7 is an exception. Tinnitus was present in two of ten patients in the "vertigo without hearing loss" group. Two of six patients with post-stapedectomy PLF symptoms and findings presented with tinnitus. In many of these patients the tinnitus improved with the PLF repair.

CONCLUSION

In these ten selected cases of patients presenting with PLF, Meniere's disease or elements of both conditions, tinnitus severity seemed to correlate closely with the state of health of the inner ear. In this group of patients tinnitus severity (intensity, constancy, and pitch) served as a marker. A higher pitch or frequency of tinnitus (cases 3, 7 and 9) seemed less amenable to medical or surgical control. In dealing with the entities of PLF and Meniere's disease, the symptom of tinnitus and its various descriptors can indicate the relative health of the cochlea.

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