Bilateral Meningoencephalocele Repair Complicated by Superior Semicircular Canal Dehiscence: Case Report

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ABSTRACT

To describe an unusual case of bilateral meningoencephaloceles with concurrent bilateral superior semicircular canal dehiscence (SSCD) and to discuss the clinical presentation, diagnosis, and treatment of SSCD. A 34-year-old man presented with unsteadiness and bilateral conductive hearing loss. He was diagnosed with bilateral meningoencephaloceles and underwent staged middle fossa approaches for repair. Following the second (right-sided) surgery, he developed sensorineural hearing loss and severe dizziness, indicating labyrinthine insult in the operated ear. He was then referred to our institution for further management. On our evaluation, the patient was continuing to experience disequilibrium and sensitivity to loud sounds. Examination revealed a positive Hennebert's sign and nystagmus consistent with symptomatic SSCD in the left ear. Computed tomography scanning with reformatting into Poeschel and Stenvers views identified bilateral SSCD. Plugging of the left SSCD was performed via a middle cranial fossa approach and resulted in improvement of the conductive hearing loss and after a period of compensation, resolution of the vestibular symptoms. This case illustrates that tegmental defects may result in simultaneous meningoencephalocele and SSCD that may complicate their repair. The importance of having a high index of suspicion and evaluation with high resolution CT scanning with appropriate reformatting is emphasized. When present and symptomatic, SSCD can be successfully managed by plugging the canal.

KEYWORDS: Meningoencephalocele, tegmental defect, superior semicircular canal dehiscence, hearing loss

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A deficient middle fossa tegmen can result in meningoencephalocele, exposure of the geniculate ganglion, and superior semicircular canal dehiscence (SSCD). First described in 1998, SSCD is a lack of bone overlying the superior canal, resulting in exposure of the membranous labyrinth to middle fossa dura.¹ This dehiscence creates a third mobile window (in addition to the round and oval windows), resulting in decreased impedance within the inner ear, which leads to symptoms of noise- and pressure-induced nystagmus, low-frequency conductive hearing loss, and autophony.^{2,3} Treatment involves either plugging of the superior canal with bone wax or reroofting of the defect to seal the labyrinth.⁴ This report describes the presence of

a simultaneous meningoencephalocele and SSCD complicating the repair.

CASE REPORT

This 34-year-old man presented to another facility with a 3-year history of unsteadiness, similar to being drunk, and was found to have a positive fistula test. Audiometric evaluation showed bilateral low-frequency conductive hearing loss of up to 40 dB (Fig. 1A). The patient had a history of a fall 13 years earlier that had resulted in neck fracture. Preoperative computed tomography (CT) scan showed

Figure 1 Audiogram prior to any operative intervention (A), following bilateral middle fossa meningoencephalocele repair (B), and following left superior semicircular canal dehiscence (SSCD) plugging (C). Note right high-frequency sensorineural loss in (B), a result of perturbation of the labyrinth during prosthesis placement, and subsequent improvement in left low-frequency conductive hearing loss in (C) from plugging of SSCD.

questionable dehiscence of the lateral semicircular canal, and the patient underwent left exploratory tympanotomy for presumed perilymphatic fistula with concurrent canal wall up mastoidectomy to evaluate the lateral canal. At the time of operation, the bone over the lateral canal was intact and no fistula was seen, but both round and oval windows were patched. A meningoencephalocele abutted the head of the ossicles but could not be satisfactorily reduced, for which the patient subsequently underwent left middle fossa craniotomy and repair of meningoencephalocele using hydroxyapatite and temporalis fascia. Postoperatively, there was no significant change in the conductive hearing loss but a 10- to 20-dB decrease in bone conduction above 2000 Hz. Next, the patient underwent a right middle fossa craniotomy with repair of meningoencephalocele (which was contacting the malleus and incus) using a preformed hydroxyapatite canal wall prosthesis. Surgery resulted in a down-sloping sensorineural hearing loss with no improvement in the conductive loss (Fig. 1B). Immediately postoperatively, the patient was acutely vertiginous for several days, progressing to persistent disequilibrium similar to that of his initial presentation.

On our subsequent initial evaluation, he reported right-sided high-pitched tinnitus and

bilateral (right more than left) otalgia with loud noises. On examination with video nystagmography goggles, positive and negative pressure on pneumatic otoscopy as well as tragal pressure caused a shifting of his visual fields on the left only. Audiometry showed bilateral mixed hearing loss, conductive in the low frequencies and sensorineural in the high frequencies (Fig. 1B). Vestibular evoked myogenic potential (VEMP) thresholds were reduced to 65 dB bilaterally. Vestibular testing showed a right unilateral vestibular paresis. Superior semicircular canal dehiscence was suspected, and CT scans with Poeschl and Stenvers reconstructions were obtained and showed bilateral dehiscence of the superior semicircular canals. The hydroxyapatite implants were found to be intact and lying lateral to the superior semicircular canals, impinging on the left SSCD (Fig. 2). The patient underwent plugging of his left superior canal dehiscence via a revision middle fossa approach. At the time of surgery, the previously placed hydroxyapatite implant was firmly in position over the tegmental dehiscence and unable to be moved, making access to the arcuate eminence difficult. Medial to the hydroxyapatite slab were several additional areas of tegmental dehiscence, but the otic capsule was not readily apparent. Therefore, a revision canal wall up mastoidectomy

Figure 2 Computed tomography, reformatted in the plain of the superior canals (Poeschl view), illustrating the proximity of the hydroxyapatite implant to the superior semicircular canal dehiscence (arrow), especially on the right (injured) side. The implants span the area of tegmental dehiscence overlying the ossicles. A nondehiscent normal superior canal from another patient is shown for comparison.

was performed to identify the superior canal, a tegmental dehiscence was created, and the location of the superior canal was marked. The canal was plugged with bone wax with an overlying layer of temporalis fascia. Postoperatively, his symptoms of dysequilibrium were relieved but replaced by those of oscillopsia, which resolved entirely over several months. Audiometry showed an improvement of 15 dB at 500 Hz and a drop of 40 dB at 8000 Hz (Fig. 1C).

DISCUSSION

Epidemiology

Defects of the temporal bone overlying the mastoid occur in 15 to 34% of temporal bones, although multiple defects occur less than 1% of the time.⁵⁻⁸ Dehiscence of the geniculate ganglion is reported in 15% of temporal bones,⁹ and it is important to recognize to avoid injury during dissection along a deficient middle fossa floor. Although the presence of one type of temporal bone defect may be predictive of a second, the incidence of SSCD or a dehiscent geniculate ganglion with another tegmental dehiscence is unknown. The incidence of tegmental dehiscence at the time of surgery for SSCD is ~33% (unpublished data). Approximately one-third of patients with SSCD have bilateral dehiscences. Analysis of temporal bones showed an incidence of 0.7%,¹⁰ and radiological evaluation with CT scan showed an incidence of 1.5%.¹¹ A case of a 51-year-old woman with a unilateral (right-sided) encephalocele with concurrent right-sided SSCD has been reported previously; however,¹² there have been no previous case reports of bilateral meningoencephaloceles with concurrent bilateral SSCDs as reported in this article.

Etiology

Dehiscence of the superior canal appears to have a genetic etiology, although head trauma, perhaps

resulting in fracture of already thin bone overlying the canal, may be the inciting event in some cases. The thickness of bone overlying one superior canal has been found to correlate with the thickness of bone over the contralateral canal.¹⁰ The symptoms of SSCD, including pressure-induced nystagmus (Hennebert's sign), sound-induced disequilibrium (Tullio's phenomenon), nystagmus, chronic imbalance, autophony, and low-frequency conductive hearing loss are thought to be caused by the shunting of energy through the dehiscence superior canal, which results in a third mobile window and can be very variable. It remains unclear why some patients with SSCD have auditory symptoms, some have vestibular symptoms, and some have both. Radiographic and pathological case series suggest that a significant proportion of patients with SSCD are asymptomatic.^{10,11}

Treatment

The physiological goal of surgery is to seal the third mobile window created by the presence of the dehiscence. This can be accomplished by reroofing or plugging of the semicircular canal. Plugging of SSCD is technically easier to perform as demonstrated in Fig. 3 and appears to be more efficacious than the reroofing procedure.^{4,13} Plugging is highly effective in ameliorating sound and pressure-induced imbalance, autophony, and chronic disequilibrium and conductive hearing loss. However, a small percentage of patients develop a mild to moderate hearing loss, usually worse in the high frequencies.⁴ The incidence of sensorineural hearing loss following posterior canal plugging for benign positional vertigo is less than 5%, and it is expected that SSCD plugging should exhibit a similar margin of safety.¹⁴ In rare cases, plugging of the superior canal can lead to vestibular injury beyond the superior canal itself, resulting in vestibular hypofunction in the operated ear,⁴ which is generally well compensated for by the unoperated side.

The middle fossa approach provides good exposure and access to the superior semicircular

Figure 3 Illustration of operative technique of superior semicircular canal dehiscence plugging. Bone wax is insinuated into both limbs of the dehiscence canal using a neuropattie. Suction irrigation is used to avoid inadvertent aspiration of the labyrinth.

canal. The presence of multiple tegmental defects makes location of the SSCD more difficult; image guidance or concurrent mastoidectomy can be helpful. The procedure can easily be performed without the use of a middle fossa retractor, which is replaced by handheld Teflon-coated brain retractors or a fenestrated suction-irrigator (to avoid inadvertent aspiration of the labyrinth). Management of SSCD with coincident significant tegmental defect may be best served with both plugging the canal and repairing the tegmental defect with a split calvarial bone graft or equivalent.

Management of Bilateral SSCD

Patients with bilateral SSCD are a therapeutic challenge. Occasionally, the patient will identify the more symptomatic ear, indicating the appropriate side for intervention. If this is not the case, the magnitude of objective pressure- and sound-induced nystagmus in each ear can be used to determine the

more symptomatic side. The radiographic size of the dehiscence does not correlate with the severity of symptoms (unpublished data). VEMP testing in cases of bilateral dehiscence does not appear to be helpful in choosing the operative side, as thresholds tend to be reduced bilaterally.

Although plugging of the superior canal would not be expected to result in impairment in the function of the other two canals of the operated labyrinth, it is wise to obtain preoperative vestibular testing to assess the function of both ears prior to the first operative intervention and certainly prior to a second-side plugging.

CONCLUSION

This case illustrates for the first time, simultaneous presence of bilateral meningoencephalocele and SSCD. Although the true incidence of concurrent tegmental defects and SSCD is unknown, radio-

graphic examination of the integrity of the superior canal using Stenvers and Poeschl views prior to operative repair of a tegmental defect can identify a dehiscence superior canal. If present, the SSCD can be safely plugged at the time of operation for meningoencephalocele repair.

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