Superior semicircular canal dehiscence: a new indication for middle fossa craniotomy

Case report

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Superior semicircular canal dehiscence is a recently described condition resulting in pressure-induced vertigo in affected patients. The diagnosis is established with the appearance of characteristic electronystagmographic and neuroimaging findings. This condition is amenable to surgical treatment by resurfacing of the dehiscence in the defect in the middle cranial fossa floor with preservation of superior semicircular canal function. The authors report on the treatment of a 35-year-old man with superior semicircular canal dehiscence by a joint neurosurgical and otolaryngological team.

KEY WORDS • superior semicircular canal dehiscence • vertigo

Superior semicircular canal dehiscence is a newly described syndrome characterized by pressure-induced vertigo. Initially described by Minor, et al., superior semicircular canal dehiscence is an analog to perilymphatic fistula and results from idiopathic thinning or absence of bone overlying the superior semicircular canal. This thinning is thought to create an additional “mobile” window within the bone labyrinth that allows transmission of vibrations to the vestibular apparatus and leads to the perception of vertigo. The dehiscence is amenable to surgical ablation or resurfacing, and presents an opportunity for neurosurgeons and neurootologists to collaborate on a surgically treatable form of vertigo. We report on a patient at our institution who recently was treated via a middle fossa approach with resurfacing of his superior semicircular canal dehiscence.

Case Report

History. This 35-year-old man presented with a 1-year history of progressive vertigo. He reported marked worsening of his symptoms with activities that produced changes in barometric pressure. A frequent air traveler, he noted vertigo during the descent phase of flights. Symptoms could be readily induced by bearing down; release of the Valsalva maneuver resulted in vertigo. He reported complete relief of symptoms when recreationally diving to depths of greater than 20 m, with recurrence of symptoms during his ascent to the surface. Additionally, he reported worsening of his vertigo in response to loud noise. The patient could recall no event that initially precipitated his symptoms. He denied any history of head trauma.

Examination. Attempts to manage the patient’s symptoms with medications and vestibular exercises were unsuccessful. Therefore, he underwent an extensive evaluation by an otolaryngologist. Results of noncontrast magnetic resonance imaging of the head were unremarkable. An audiogram showed no evidence of decay, and normal auditory reflexes were present. Results of ENG were remarkable for torsional upbeat nystagmus induced both with the Hennebert test (in which both positive and negative pressure is applied to the intact tympanic membrane with a pneumatic otoscope) and with loud, low-frequency tones (500 Hz and 1 kHz) delivered to the right ear (Fig. 1). A temporal bone CT scan led us to suspect right-sided superior semicircular canal dehiscence (Fig. 2).

Operation. The patient underwent surgical treatment of his dehiscence. Exploration via a right middle fossa approach revealed the superior semicircular canal presenting through the cranial floor (Fig. 3). The canal was resurfaced by placement of split calvarial bone graft, temporalis fascial graft, and fibrin glue over the defect. The patient tolerated the procedure well, and was discharged home on postoperative Day 3.

Postoperative Course. The patient reported marked improvement postoperatively, but suffered recurrent symptoms after riding in an unpressurized military transport flight. He required repeated surgery and resurfacing with hydroxyapatite paste, with excellent clinical results. Postoperative audiological evaluation demonstrated no decrease in cochlear function. Postoperative vestibular evaluation dem-
onstrated mild right vestibular hypofunction. The patient reported a single episode of positional vertigo postoperatively, which resolved after 1 week of vestibular rehabilitation therapy. He reports significant improvement in his preoperative symptoms through 1 year of follow up.

Discussion
Superior semicircular canal dehiscence is a treatable disorder marked by vestibular disturbance and is characterized by specific symptoms, signs, and neuroimaging findings. Patients report vertigo that is exacerbated by changes in barometric pressure in the middle ear. Symptoms may be reproduced with insufflation of the tympanic membrane or with a Valsalva maneuver in an office setting. Additionally, some patients report the Tullio phenomenon, or worsening or vertigo in response to loud noise. The ENG modality can be used to document the physical signs associated with superior semicircular canal dehiscence. Performance of the Hennebert or fistula test results in the production of torsional upbeat nystagmus. Nystagmus is also produced in response to loud sounds delivered to the involved ear. Fine-cut CT scans of the temporal bone are confirmatory in the diagnosis of superior semicircular canal dehiscence, demonstrating thinning or absence of bone overlying the superior semicircular canal at the arcuate eminence.

Experience with the treatment of superior semicircular canal dehiscence by canal resurfacing is limited to case reports at this time. Unlike canal ablation via a transmastoid approach, resurfacing allows for preservation of residual superior canal function. Minor reported on five patients who were treated with canal resurfacing to seal the fistula. Brantberg, et al., reported on two patients who were treated with transmastoid occlusion of the superior canal. Although this method results in ablation of the superior canal, it obviates the need for craniotomy, potentially decreasing the patient’s perioperative risk. Both of the aforementioned studies documented improvement or resolution of symptoms in treated patients, without significant morbidity. We used a standard middle fossa approach for canal resurfacing, with an excellent clinical result.

Conclusions
Superior semicircular canal dehiscence is a newly char-
Superior semicircular canal dehiscence treated with middle fossa craniotomy

acterized vertiginous syndrome with a characteristic constellation of clinical features. The patient’s history and results of ENG and temporal bone CT scanning are essential in establishing this diagnosis. Surgical treatment via a middle fossa approach by a joint neurosurgical/otolaryngological team is safe and effective; it can result in marked symptomatic improvement in these patients.

References

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