Journal of Clinical Neuroscience 43 (2017) 103–107

Contents lists available at ScienceDirect

Journal of Clinical Neuroscience

journal homepage: www.elsevier.com/locate/jocn

Clinical commentary

Outcomes of middle fossa craniotomy for the repair of superior semicircular canal dehiscence

Nolan Ung [a], Lawrance K. Chung [a], Carlito Lagman [a], Nikhilesh S. Bhatt [a], Natalie E. Barnette [a], Vera Ong [a], Quinton Gopen [a,b], Isaac Yang [a,b,c,d,*]

[a] Department of Neurosurgery, University of California, Los Angeles, 300 Stein Plaza, 5th Floor Wasserman Bldg., Los Angeles, CA 90095-6901, United States
[b] Department of Head and Neck Surgery, University of California, Los Angeles, 200 UCLA Medical Plaza, Suite 550, Los Angeles, CA 90095-6901, United States
[c] Jonsson Comprehensive Cancer Center, University of California, Los Angeles, 200 UCLA Medical Plaza, Suite B265, Los Angeles, CA 90095-6951, United States
[d] Department of Radiation Oncology, University of California, Los Angeles, 200 UCLA Medical Plaza, Suite B265, Los Angeles, CA 90095-6951, United States

ARTICLE INFO

Article history:
Received 24 October 2016
Accepted 21 May 2017

Keywords:
Dehiscence
Middle cranial fossa
Outcomes
Semicircular canal

ABSTRACT

Superior semicircular canal dehiscence (SSCD) is a rare defect of the arcuate eminence that causes an abnormal connection between the superior semicircular canal and middle cranial fossa. Patients often present with a variety of auditory and vestibular symptoms. Trigger avoidance is the initial strategy, but surgery may be necessary in debilitating cases. We retrospectively reviewed SSCD patients undergoing repair via a middle fossa craniotomy between March 2011 and September 2015. Forty-nine patients undergoing 58 surgeries were identified. Autophony was the most common symptom at presentation ($n = 44$; 90%). Mean follow-up was 10.9 months, with 100% of patients reporting resolution of at least one symptom. Aural fullness was the most commonly resolved symptom following surgical repair ($n = 19/22$; 86%). Hearing loss ($n = 11/25$; 44%) and tinnitus ($n = 11/38$; 29%) were the most common symptoms to persist following surgery. The most common symptom to develop after surgery was disequilibrium ($n = 4/18$; 22%). Upon comparing the overall pre-operative and post-operative groups, the number of patients with autophony ($p < 0.0001$), aural fullness ($p = 0.0006$), hearing loss ($p = 0.0119$), disequilibrium ($p = 0.0002$), sound- and pressure-induced vertigo ($p < 0.0001$), and tinnitus ($p < 0.0001$) were significantly different. Improved clinical outcomes were demonstrated in patients undergoing SSCD repair through a middle cranial fossa approach. The most common presenting symptom (autophony) was also most likely to resolve after surgery. Hearing loss is less amenable to surgical correction. Disequilibrium developed in a small number of patients after repair.

1. Introduction

Superior semicircular canal dehiscence (SSCD) describes a rare cranial defect in the arcuate eminence, which is the bone directly overlying the super semicircular canal of the inner ear. At the interface between the fluid-filled inner ear and the air-filled middle ear are two mobile windows, the round and oval windows. Sound waves are translated into mechanical energy at these air-fluid interfaces, ultimately resulting in stimulation of hair cells and normal audition. In SSCD, a connection between the superior semicircular canal and middle cranial fossa generates a third mobile window, most commonly in the superior wall of the canal. This alters fluid flow from the cochlea and amplifies impedance variation between the scala vestibule and tympani [1]. This increases the air conduction hearing threshold (especially for low frequencies) and decreases the bone conduction hearing threshold, which results in auditory and vestibular symptoms [2-4].

The clinical manifestations of SSCD include oscillopsia, sound-induced vertigo (Tullio phenomenon), pressure-induced vertigo (Hennenbert sign), pulsatile tinnitus, aural fullness, hearing loss, and autophony [5-10]. Patients with larger dehiscence tend to have more pronounced symptoms [11]. In fact, the width of the defect has been correlated with hearing loss [12]. Patients who present with autophony often have cerebrospinal fluid involvement within the dehiscent area, which amplifies sounds within the body [13-15]. Some patients report hearing their own heartbeat, eye movements, or footsteps. The pathogeneses of other symptoms associated with SSCD are not fully elucidated.

* Corresponding author at: Department of Neurosurgery, University of California, Los Angeles, 300 Stein Plaza, Ste. 562, 5th Floor Wasserman Bldg., Los Angeles, CA 90095-1761, United States. Fax: +1 (310) 825 9384.
E-mail address: iyang@mednet.ucla.edu (I. Yang).

http://dx.doi.org/10.1016/j.jocn.2017.05.003
0967-5868/

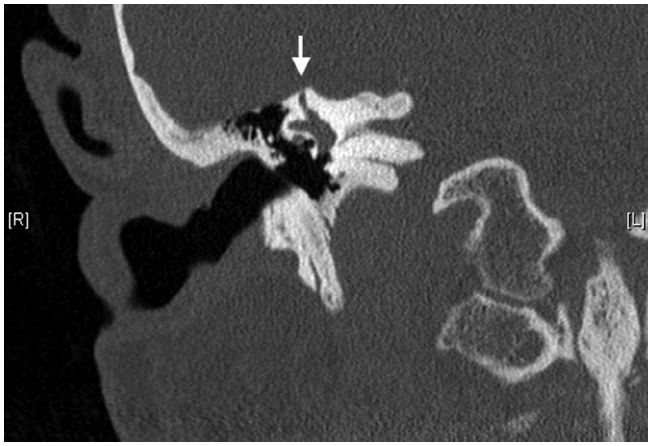

Fig. 1. High-resolution computed tomography of the temporal bone demonstrating a superior semicircular canal dehiscence (arrow).

Diagnosis of SSCD is aided by clinical presentation and audiometric and vestibular evaluation, such as the vestibular-evoked myogenic potential (VEMP) test. VEMP testing may demonstrate low threshold responses for cervical (saccular) responses in the affected ear or elevated amplitude responses for ocular (utricular) responses in the contralateral eye [16,17]. However, the diagnostic gold standard is still high-resolution computerized tomography (CT) of the temporal bone (Fig. 1) [18-20]. Occasionally, SSCD is found incidentally on imaging or during unrelated surgical procedures in otherwise asymptomatic patients [21]. Non-surgical treatment includes avoidance of triggers and may be appropriate in cases where symptoms do not significantly impact daily activities [22]. Surgical management may be indicated in patients presenting with debilitating symptoms attributable to SSCD and refractory to conservative measures. Several surgical approaches for repair of SSCD have been reported in the literature, including transmastoid approach and middle fossa craniotomy; the latter is the focus of this study [23-33].

Surgical management of SSCD has been shown to be effective in the resolution of sound- and pressure-induced vertigo, autophony, hearing loss, aural fullness, and dizziness [7,19,34-37]. The current literature is limited to small case series [2,19,34,38-42]. Our group recently reported on clinical outcomes of middle fossa craniotomy for SSCD in a prior study of 18 patients [43]. This paper provides a substantial update to that study with nearly triple the patient size and analyzes symptom resolution from the largest single-institution patient cohort to date.

2. Methods

We conducted a retrospective review of electronic medical records (EMRs) for patients undergoing surgical repair of SSCD via a middle cranial fossa approach performed by the senior authors (QG and IY) at our institution between April 2011 and September 2015. Of note, this patient cohort includes those identified in a previous study [43]. Patients with unilateral and bilateral SSCD repaired through a middle fossa craniotomy were included. Patients with SSCD undergoing non-surgical management or surgical repair through a different approach were excluded. Symptom outcomes were assessed through a review of EMRs. For patients with bilateral SSCD, those who underwent unilateral repair were evaluated at latest follow-up and those who underwent a two-stage bilateral repair were evaluated after the second surgery. This study was reviewed and approved by the institutional review board (Protocol ID: 13-001820).

2.1. Data analysis

Data on patient demographics, pre-operative symptoms, audiometry results, and post-operative outcomes (based on the most recent follow-up visit) were extracted from the EMR. When available, post-operative audiometry results were used to determine hearing outcomes, and subjective patient report was used only when necessary. Patient health information was de-identified, and statistical analysis was performed using Fisher's exact test with statistical significance set at P value less than 0.05.

2.2. Surgical technique

Middle fossa craniotomy is the most common approach to SSCD repair. The advantage of this approach is direct visualization of the dehiscence, which allows for optimal repair of the cranial defect. For all procedures, the temporal lobe was elevated under the operative microscope to expose the floor of the temporal fossa. The arcuate eminence and area of dehiscence were identified through a combination of microscopic visualization and intraoperative image-guidance. Temporalis fascia and bone wax were used to plug the dehiscence and then sealed with fibrin glue to secure the plug.

3. Results

A total of 49 patients with SSCD undergoing 58 middle fossa craniotomies for repair of SSCD, from March 2011 to September 2015, were identified. Females comprised 63.3% (n = 31) of the patients and males comprised 36.7% (n = 18), with an approximate female to male ratio of 1.7:1. Mean age for all patients was 50.3 years (range 21–84 years). SSCD repair was most often performed on the left (n = 23; 47%), then right (n = 17; 35%), and bilaterally (n = 9; 18%). Mean follow-up was 10.9 months (range 0.2–50.0 months), with 100% of patients (n = 49) reporting resolution of at least one symptom. Baseline patient characteristics are summarized in Table 1.

Autophony was the most common symptom at presentation (n = 44; 90%), followed by tinnitus (n = 38; 78%), vertigo (n = 33; 67%), disequilibrium (n = 31; 63%), hearing loss (n = 25; 51%), aural fullness (n = 22; 45%), and headache (n = 14; 29%). Aural fullness was the most common symptom to resolve following surgical repair (n = 19/22; 86%), followed by autophony (n = 35/44; 80%), vertigo (n = 25/33; 76%), disequilibrium (n = 23/31; 74%), tinnitus (n = 27/38; 71%), headache (n = 10/14; 71%), and hearing loss (n = 14/25; 56%). Patients who experienced improvement in symptoms did not report recurrence of symptoms at last follow-up.

Hearing loss (n = 11/25; 44%) and tinnitus (n = 11/38; 29%) were the most common symptoms that persisted after surgery, followed by headache (n = 4/14; 29%), disequilibrium (n = 8/31; 26%), sound- and pressure-induced vertigo (n = 8/33; 24%), autophony (n = 9/44; 20%), and aural fullness (n = 3/22; 14%). A minority of patients developed new (post-operative) symptoms, including dis-

Table 1
Summary of patient characteristics.

Variable	n (%)
Patients (Total)	49
Male	18 (37)
Female	31 (63)
Laterality	
Right	17 (35)
Left	23 (47)
Bilateral	9 (18)
Mean follow-up (mos, range)	10.9, 0.2–50.0