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Postpartum Vertigo and Superior Semicircular Canal Dehiscence Syndrome

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Abstract

Background—Superior semicircular canal dehiscence is a recently described cause of imbalance, hearing loss, and tinnitus. Symptoms may begin after abrupt changes in intracranial or middle ear pressure.

Case—This woman presented with a 6-year history of imbalance, hearing loss, and pulsatile tinnitus beginning when she was pushing during labor. A temporal bone computed tomography showed a dehiscence of the superior semicircular canal. Surgical repair of the dehiscence through the middle cranial fossa resulted in immediate resolution of the patient's symptoms, and she returned to full activity within 3 weeks.

Conclusion—Superior semicircular canal dehiscence is increasingly recognized as a cause of multiple otologic symptoms. Obstetricians and gynecologists with patients complaining about postpartum vertigo should inquire about symptom onset and focus their questions around events during the second stage of labor. Patients with symptoms of dehiscence should be referred to a neurotologist for management, including possible surgical repair.

INTRODUCTION

Imbalance is a common complaint, including among obstetrics and gynecology patients (1). Dehiscence of the superior semicircular canal has increasingly been recognized as a cause for episodic or chronic imbalance, hearing loss, autophony (an abnormally loud perception of one's own voice), or a combination of these symptoms. We present a case of superior semicircular canal dehiscence (SSCD) syndrome presenting with acute vertigo during labor.

CASE

A 43-year-old G5P2 patient was referred for neurotologic care with a 6-year history of imbalance, tinnitus, and hearing loss on the right side. Her symptoms began with an “explosion” in her right ear as she was pushing during labor with her eldest child. This was accompanied by a sudden onset of pain, pulsatile tinnitus, and autophony in the right ear. After delivery, she noted new symptoms of dizziness that she described as feeling like she was “being pushed or knocked over by a wave” which worsened with exposure to loud sounds. She also reported a newly heightened sensitivity to motion which made it impossible for her to enjoy amusement

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park rides and forced her to walk next to walls in order to steady her gait. Her previous history was remarkable only for a slight high-frequency sensorineural hearing loss on the left side related to noise exposure at work as a firefighter and paramedic and occasional headaches which had been evaluated previously with normal MRI studies.

On physical exam, she appeared well with normal findings on otoscopy and an unremarkable general neurologic exam. Valsalva with glottis open blowing against a pinched nose elicited combined vertical-torsional eye movements and caused subjective imbalance. She had a positive Hennebert sign (abnormal eye movements with pressure in the external auditory canal) on the right side. She heard a tuning fork placed on her forehead louder in her right than left ear, indicating a right conductive or left sensorineural hearing loss.

Pure-tone audiometry of the right ear confirmed a mild conductive hearing loss and demonstrated an abnormal hypersensitivity to bone-conducted sounds transmitted directly through the skull to the inner ear. Pure tones of 500, 1000, and 2000 Hz presented at 120 dB HL provoked symptoms of imbalance when presented in the right ear but not the left. Vestibular testing with calorics (warm and cool water irrigations of the external auditory canals) and with rotation in a chair about an earth-vertical axis were unremarkable. Vestibular-evoked myogenic potential (VEMP) testing indicated hypersensitivity of the vestibular system to sound in the right ear but not the left. A high-resolution temporal bone CT showed a dehiscence of the bone normally separating the lumen of the superior semicircular canal and the middle cranial fossa on the right (Figure 1). Surgical repair of the dehiscence through the middle cranial fossa resulted in immediate resolution of the patient's symptoms, and she returned to full activity within 3 weeks.

COMMENT

Superior semicircular canal dehiscence syndrome (SSCD) relates a dehiscence of the bone normally overlying the superior semicircular canal to symptoms of vertigo and oscillopsia induced by loud sounds (Tullio's phenomenon), changes in middle ear pressure, or changes in intracranial pressure (2). The syndrome is increasingly recognized as a cause of chronic disequilibrium and dizziness, tinnitus, and conductive hearing loss sometimes accompanied by enhanced bone-conducted hearing (3).

Dense labyrinthine bone surrounding the superior semicircular canal normally bulges into the middle cranial fossa, forming the arcuate eminence. This bone covering the canal is extremely thin in about 1.3% of all individuals and frankly dehiscent in approximately 0.7% (4). It is believed that a dehiscence may result from congenital underdevelopment of the temporal bone followed by an event later in life that breaks open the weak area. Precipitating events may involve high intracranial or middle ear pressures such as during weightlifting or pushing during labor as reported here, but most patients do not remember a specific time of onset of symptoms (3). The differential diagnosis includes perilymphatic fistula, which can also be caused by a rapid pressure change and manifest with symptoms of imbalance and hearing loss. This condition results from leakage of fluid from the inner ear into the middle ear, usually through the oval or round windows.

High-resolution non-contrast temporal bone CT is used to confirm the clinical diagnosis of SSCD. Using patients with symptoms of SSCD as a gold standard, the positive predictive value of axial and coronal CT scans is only about 50%. However, fine-cut (0.5 mm or finer) scans reformatted parallel to and orthogonal to the semicircular canals can achieve a positive predictive value for dehiscence of 93% (5). A convincing clinical history and a positive CT scan may be sufficient to make the diagnosis, but audiometry and vestibular assessment are usually used as confirmatory tests. Patients often demonstrate a conductive hearing loss and

supranormal bone conduction manifesting as an abnormal sensitivity to sounds normally generated by the body. An audiogram with these findings can be an important indicator of canal dehiscence in a patient with an appropriate clinical history and merits a referral to a neurotologist. This is particularly important in health care settings where other tests are not available.

Standard vestibular tests including calorics and rotational exam are rarely informative, but recently developed techniques to quantify vestibular-evoked myogenic potentials (VEMPs) can help determine the presence of a dehiscence. A VEMP is a normal reflex relaxation of the sternocleidomastoid muscle in response to loud sounds in the ipsilateral ear. In patients with SSCD, the threshold for evoking a potential from the affected ear is often lower than normal, as in the patient reported here (6).

Several conditions other than superior canal dehiscence and perilymphatic fistula are important to consider when evaluating gravid or post-partum patients with symptoms of imbalance. Meningiomas are known to be progesterone sensitive and may grow rapidly during pregnancy. These can cause imbalance if they affect the vestibular nerve directly or central areas of the brainstem or cerebellum that govern balance function. Otosclerosis is also hormone sensitive and is known to worsen during pregnancy. This disorder of bone homeostasis affecting the inner ear often presents with hearing loss and tinnitus, but imbalance is not uncommon.

The management of SSCD ranges from reassurance for patients with mild symptoms to surgical management for more severely affected patients. There is no medical therapy for SSCD. The superior canal can be approached either through the mastoid or the middle cranial fossa, allowing the dehiscent area to be plugged with bone wax. In a series of 20 patients undergoing surgical repair, results were excellent with a complete resolution in symptoms in 15 (7). Hearing is theoretically at risk for any ear procedure, but no loss was reported in a large series of patients undergoing primary repair of a canal dehiscence (8).

Disequilibrium is notoriously difficult to diagnose and patients may go untreated for years. Obstetricians and gynecologists with patients complaining about new onset postpartum vertigo should inquire about symptom onset and focus their questions around events during the second stage of labor. Patients with symptoms of SSCD should be referred to a neurotologist.

BOX

Symptoms and signs of superior semicircular canal dehiscence

Vertigo in response to loud sounds

Vertigo in response to abrupt pressure changes (such as sneezing or performing a Valsalva maneuver)

Conductive hearing loss

Hypersensitivity to bone-conducted sounds (hearing one's own heartbeat, breathing, eyeballs moving, or feet striking the ground while walking)

Autophony (hearing one's own voice)

Chronic imbalance

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Figure 1. Oblique computed tomography scan through the right mastoid cavity and middle cranial fossa. Superior semicircular canal indicated with asterisk. Dehiscence between the superior semicircular canal and the middle cranial fossa indicated with arrow.