Introduction

The jugular bulb (JB) is a venous structure in the posterior cranial fossa created as the sigmoid sinus transitions to the internal jugular vein at the jugular foramen. It is usually located below the hypotympanum and is separated from middle ear structures by a bony encasement. Rarely, the JB can enter into the middle ear, an anatomically abnormality known as a high-riding jugular bulb (HJB). If the bony encasement is incomplete, it is called a dehiscent jugular bulb (DJB). Patients with these abnormalities have a variety of presentations, from being asymptomatic to reporting hearing loss, pulsatile tinnitus (PT), and vertigo.

There are no known medical treatments for PT due to HJB or DJB, though both obliterator surgical and endovascular stenting treatments have been described. However, variable efficacy in controlling PT coupled with potentially serious complications have prompted a search for alternative management strategies for this patient population. One such strategy involves reinforcing, or “soundproofing,” the DJB. In 2010, El-Begemy et al. described a multilayer reconstruction of the middle ear floor using an endaural approach with canaloplasty and tragal cartilage graft. PT disappeared in 57% of patients in this series.

One alternative to the use of autologous tissues is hydroxyapatite (HA) cement, a product familiar to most otologists. In this series we describe a novel technique for the treatment of PT associated with HRJB by reinforcing the bony separation between the JB and middle and inner ear structures.

Case Series

Case 1
A healthy 50 year old female presented with sudden onset of severe right-sided pulsatile tinnitus by a bruit described as very loud and persistent, sometimes “whooshing and whooshing,” worse in quiet situations and when supine. Otoscopic examination was normal and skull and neck auscultation revealed no bruits. An audiogram was normal. HRCT and CT confirmed the presence of an ipsilateral DJB. She underwent interventional therapy. At her one week post-operative visit, she noted complete resolution of the PT. Post-operative audiogram was normal. Telephone interview at two years postoperatively confirmed absence of tinnitus recurrence.

Case 2
A healthy 70 year old male presented with a 4 year history of severe right-sided pulsatile tinnitus. It began suddenly after forcefully popping his ears open during an upper respiratory infection. He described it as constant, very loud, pulsatile and whooshing that had not changed in intensity since onset. By turning his head to the left he was able to decrease the intensity of the pulsations. Otoscopic examination was normal and skull and neck auscultation revealed no bruits. An audiogram demonstrated bilateral normal low frequency hearing sloping to asymmetric, right greater than left, severe high frequency sensorineural hearing loss. MRI was negative for retrocochlear pathology. HRCT of the temporal bone revealed a DJB. He underwent repair and intraoperatively a thin layer of cracked eggshell-like bone was noted over the JB, so that the buld could be gently compressed. He also had a small adhesion band from the jugular bulb to the round window membrane. He noted immediate resolution of the PT in the recovery room. He remained asymptomatic 17 months postoperatively with no change on audiogram.

Case 3
A healthy 64 year old female presented with a 2 year history of worsening right-sided pulsatile tinnitus. It began rapidly after a fall down 10 stairs. She noted mild “whishing” in her right ear only, which came louder over the course of the following year. She described it as constant, very loud, pulsatile and whooshing that worsened if she lay on her right side. Otoscopic examination was normal and skull and neck auscultation revealed no bruits. An audiogram was normal (Figure 1). A right DJB was identified on HRCT of the temporal bone (Figure 2, 3). She was noted to have thin, but intact bone overlying both superior semicircular canals. She underwent repair and intraoperatively a very thin layer of bone was noted over the JB, so that the buld could not be compressed. Also noted was a small adhesion band from the jugular bulb to the round window membrane. She noted near complete resolution of her PT at her 1 week post-operative visit and remained asymptomatic through follow up 5 months later, with little change on follow-up audiogram (Figure 4). HRCT of the temporal bone post-operatively showed a well-covered JB within the middle ear (Figure 5, 6).

Technique

Preoperatively, the risks and benefits of operative intervention are discussed with patients, including failure to improve symptoms. General anesthesia is induced and patients are intubated, prepped, and positioned for a trans-canal approach.

A posterior-inferior based tympanomeatal flap is raised. The length of the flap should be longer than what is typically utilized in case removal of the inferior medial bony tympanic annulus is necessary to adequately visualize the JB and round window niche. Use caution during flap elevation since the jugular bulb is likely higher and dehiscent, so more vulnerable to injury during this stage. Examine the middle ear cavity for abnormal soft tissue such as adhesions or jugular bulb diverticula, which should be addressed. Gently palpate the jugular bulb to identify sites of dehiscence or exposure.

Protect the round window niche with a small piece of saline soaked gel foam before mixing and applying HA over the contour of the jugular bulb. Less than 1 cc of OtoFill (Olympus America, PA) was used in each case. Wait until the HA sets before removing this gel foam to prevent occluding the round window, which would likely cause hearing loss. No middle ear packing is used. The tympanomeatal flap is then laid back.

Post-operative care consists of avoiding straining for 2 weeks after surgery.

Discussion

The differential diagnoses for venous pulsatile tinnitus is long, with most causes optimally managed medically. However, in rare cases, surgery can provide meaningful long-term relief without substantial risk. Such is the case with PT secondary to HJB or DJB. It is important to have a high suspicion that the JB abnormality presents the only cause of a patient’s symptoms. Ruling out other potential sources of venous PT, including sigmoid sinus dehiscence, sigmoid sinus diverticulum, and superior semicircular canal dehiscence is paramount.

The described transcranial JB resurfacing technique using HA is proposed as a safe and effective option alternative treatment for pulsatile tinnitus due to high or dehiscent JB. There are several advantages to this method of repair. The transcranial approach avoids a retroauricular incision and does not have the donor site morbidity associated with a tragal cartilage graft. HA is commonly used in otologic surgery and is familiar to surgeons. It sets quickly within minutes and does not shift over time, thus lowering the risk of recurrent symptoms.

Reconstruction of the middle ear floor with a sound proof construct of layers of autologous tissue grafts has been reported in a small series of patients (El-Begemy 2010). The jugular bulb encasement was rebuilt using a tragal cartilage graft with attached perichondrium and bone dust. Of seven patients included in this series, 5 (71%) had improvement of their pulsatile tinnitus and two (28%) patients reported a change in the quality of the tinnitus but did not have symptomatic improvement. One patient developed thought was to be due to sigmoid sinus obstruction leading to increased venous pressure. She eventually improved with acetazolamide and therapeutic warfarin treatment for 6 months.

As with all otologic surgery, delicate technique is critical to successful outcomes. Palpation of the JB can confirm the presence of an intact bony covering. However this series suggests that the presence or thickness of bone overlying the JB may have no bearing on the success of resurfacing. Rather, “soundproofing” of the middle ear from the altered flow dynamics within the JB, rather than by directly addressing underlying intracranial pathology, is responsible.

Two cases were noted to have adhesions from the jugular bulb to the round window niche. While theoretically pulsations from the JB may have been transmitted to the round window membrane may account the subjective tinnitus, this may be an incidental finding. Mucosal adhesions within the middle ear are commonplace do account for the pulsatile tinnitus in the patient without adhesions noted.

This case series describes three healthy adult patients, all with similar age and BMI, with right-sided pulsatile tinnitus. Two patients have a history suspicious for a post-traumatic etiology of their pulsatile tinnitus. The JB abnormalities presented represent a range of severity, from obvious dehiscence to baseline fracture to intact but thin bone. Irrespective, in carefully chosen patients with high suspicion for PT due to HUB or DJUB, cement resurfacing appears to be an effective long-term treatment with low risk of complications.

Conclusions

Jugular bulb resurfacing with HA is a safe and effective method of surgical treatment for pulsatile tinnitus due to high riding or dehiscent JBs. It avoids the risk of endovascular intervention and the donor morbidity of a layered tragal cartilage graft. Although this series is limited in its retrospective, case series design and by the small number of patients included, no significant complications of hearing loss, sinus thrombosis, or recurrent symptoms were encountered. Short term surgical outcomes are promising.

References


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