Sinus pericranii, petrosquamosal sinus and extracranial sigmoid sinus: Anatomical variations to consider during a retroauricular approach

Juan Carlos Cisneros *, Paula Tardim Lopes, Ricardo Ferreira Bento, Robinson Koji Tsuji
Otorhinolaryngology Department, Hospital das Clínicas – University of São Paulo, São Paulo, Brazil

1. Introduction

The retroauricular surgical approach is the most commonly practiced approach for exploration of the middle ear. It serves well to perform all kinds of middle ear related surgeries as well as inner ear surgeries, such as cochlear implantation. In some cases, rare anatomic variations of the temporal bone related sinuses, such as the sinus pericranii, might transform this common surgical pathway into a real life-threatening problem. The term sinus pericranii refers to a cranial venous anomaly, which presents as a fluctuating, compressible venous scalp mass that connects directly to the intracranial dural sinuses through dilated diploic and emissary veins, which may or may not become varicous. This anomaly provides a direct, abnormal communication between the intracranial dural sinuses and the extracranial veins [1–3]. The petrosquamosal sinus is one of these rare emissary veins, and in humans it typically regresses during fetal and early post-natal life. As noted by Mortazavi et al. in 2012, it serves as the primary cerebral drainage site in many lesser primates and quadrupeds, contrary to humans, in whom the internal jugular veins and vertebral veins represent the major outflow pathways. When present, its diameter ranges from 2 to 4 mm, and it originates at the junction of the transverse and sigmoid sinuses, coursing laterally above the superior border of the temporal bone [4]. The petrosquamosal sinus may be confused with a large mastoid emissary vein, since in many described cases, it presents as an extracranial vein varix in the vicinity of the mastoid region, which may be covered by a thin bone layer or only by subcutaneous tissue. This vascular anomaly demands a correct preoperative diagnosis since its damage during surgery may be life threatening, leading to troublesome bleeding or even fatal ischemic consequences, especially because it sometimes

* Corresponding author at: Hospital das Clínicas, University of São Paulo – Faculty of Medicine, Av. Dr. Eneas de Carvalho Aguiar, 255, 6th Floor, Room 6167, São Paulo 05403 000, Brazil. Phone number: +55(11) 974850206.
E-mail address: juancarloscisneros@me.com (J.C. Cisneros).

http://dx.doi.org/10.1016/j.anl.2016.07.003
0385-8146 © 2016 Elsevier Ireland Ltd. All rights reserved.
represents the major or only drainage route of the transverse sinus, which connects the dural sinuses with the external jugular venous system [5].

Another venous anomaly of the temporal bone that surgeons should have in mind is the extratemporal course of the sigmoid sinus. When encountered, the sinus is anteriorly and laterally displaced with no cortical bone cover. This uncommon course of the sigmoid sinus may increase the risk of damage and troublesome bleeding immediately after a common retroauricular skin incision is performed [6,7].

It is important to consider that venous malformations of the temporal bone are a more common feature when associated with CHARGE syndrome. Besides inner and middle ear anomalies associated with this syndrome, patients with CHARGE may present a number of collateral emissary veins in the temporal bone including posterior condylar veins, mastoid emissary veins connecting occipital or post-auricular veins with the sigmoid sinus, and the aforementioned petrosquamosal sinus [8].

Even though most ear surgeons often have temporal bone CT images available prior to surgery, these vascular anomalies may not be easily detected. A thorough analysis of CT scans, as well as contrast-enhanced MRI images, is useful to better diagnose these anomalies and to avoid surgical complications [9]. We report three cases of temporal bone venous sinus anomalies seen in patients who underwent cochlear implant surgery.

2. Case reports

2.1. Case 1

A 17-year-old male, with previous diagnosis of CHARGE syndrome, was evaluated for cochlear implant surgery. At the age of 3 years, moderate to severe sensorineural hearing loss in the right ear and severe to profound sensorineural hearing loss in the left ear were diagnosed, together with growth retardation and a delay in psychomotor and language development. Since childhood, he used a hearing aid in the right ear with good benefit, but at 16 years of age, hearing progressively worsened bilaterally with a profound hearing loss in the right ear and anacusis in the left (average hearing threshold of 100 dB in the right ear taking into account the 0.5, 1, 2 and 3 kHz frequencies). At this point, he became a candidate for cochlear implantation.

During physical examination, hypertelorism and a surgically closed cleft palate with an absent uvula were identified; otoscopy revealed a grade 1 bilateral microtia (according to the classification by Altmann et al.) with narrow external auditory canals [10]. Tympanic membranes were normal bilaterally. The patient also presented other features typical of CHARGE syndrome, such as genitourinary tract anomalies. Audiological and imaging findings may be seen in Figs. 1 and 2.

In this case, the association of CHARGE syndrome and sinus pericranii is readily seen [8]. Since cochlear implantation in the left ear was contraindicated because of vestibulocochlear nerve agenesis, only the right ear was implanted. After considering the vascular malformation seen in the image studies, a traditional mastoidectomy was avoided and a suprameatal approach was preferred. A tunnel was drilled just above the Henle’s spine in the posterior–superior region of the external auditory canal, passing just inferior to the dura of the middle fossa in an anterior–inferior direction, finishing in the attic. A tympanomeatal flap was elevated to properly see the round window region and insert the electrode array. A cochleostomy was performed in the estimated topography of the round window, which was absent, and the electrode array was completely introduced in the basal turn of the cochlea. The

![Fig. 1. Audiometric and MRI findings prior to cochlear implantation. (A) Audiometric results without hearing aid. The patient showed no speech discrimination. (B) Audiometric results with a well-adapted hearing aid in the right ear. Speech discrimination thresholds in closed and open set are below 50%. (C) Axial MRI image of the right and left internal auditory canals showing a cochlear nerve in the right side (arrowhead) and absence of the cochlear nerve in the left, which correlates with auditory findings. The ★ shows the hypoplastic cystic labyrinth bilaterally. dB = decibel, Hz = hertz.](image-url)
A tympanomeatal flap was replaced and the receiver-stimulator was positioned in a niche previously drilled in the squamous portion of the temporal bone, in a more vertical fashion than usual to avoid the sinus pericranii. After 3 months of activation, the cochlear implant allowed an improvement in average pure tone thresholds to 42 dB, with field discrimination in closed set of 100% and open set of 55%. The postoperative temporal bone CT scans with the cochlear implant in place are presented in Fig. 3.

2.2. Case 2

A 66-year-old male presented with a progressive bilateral hearing loss that started at the age of 2. No direct causes were identified and there was no related family history of hearing loss. He started using a hearing aid on the right ear when he was 26 years old and on the left at 55 years of age when he became a candidate for cochlear implantation. Throughout his life, he developed excellent lip-reading skills and he underwent speech therapy once a week.

During cochlear implant evaluation, ear CT scans and MR imaging demonstrated an abnormally high jugular bulb on the left side and an abnormal venous lake in the occipital region, which communicated the left dural intracranial venous sinuses with a conglomerate of pericranial vessels in the nuchal region deep to the muscle and adipose planes, consistent with sinus pericranii. Temporal bone CT scans and MRI images may be seen in Fig. 4.

During audiometric evaluation, both sides presented a restricted access to speech perception, and despite the longer auditory deprivation time for the right ear, this ear was selected for cochlear implantation to avoid any of the vascular abnormalities in the left side. A good functional result was obtained after cochlear implant activation.

2.3. Case 3

A 4-year-old male presented with a history of bilateral profound hearing loss secondary to adverse factors at birth and aminoglycoside use. Brain stem evoked response...
audiometry showed absent responses bilaterally. The patient presented with an MRI and the radiologist’s report of a CT scan performed in an outside setting. The MRI showed normal inner ear anatomy with present seventh and eighth cranial nerves bilaterally and the report of a CT scan suggested a normal anatomy of the external, middle and inner ear bilaterally. At the time of surgery, only the MRI images were available.

Fig. 3. Postoperative temporal bone CT images and cranial general scan. The axial (A–C) and coronal (D, E) images demonstrate that electrodes are properly inserted in the cochlea through a suprameatal approach. The general scan (F) shows the position selected for the receptor-stimulator to avoid the sinus pericranii.

Fig. 4. Preoperative temporal bone coronal CT scans and MRI sagittal images. (A–C) Preoperative temporal bone CT images showing a high jugular bulb and a well-pneumatized mastoid. (D, E) T1-weighted sagittal MRI images after contrast that demonstrate an anomalous venous lake in the occipital region, which communicates the left dural venous sinuses with a conglomerate of pericranial vessels in the nuchal region (arrows) deep to the muscle and adipose planes, consistent with sinus pericranii.
During surgery, anesthetic infiltration was performed in the retroauricular region in a normal fashion. A retroauricular C-shaped skin incision was performed at a distance of 0.5 cm from the conchal cartilage and the temporalis fascia plane was identified. A muscular-periosteal anteriorly based flap was performed in a regular fashion with a Bovie electrocautery, and immediately after incising the muscle, a profuse and troublesome bleeding started. The mastoid cortex was properly exposed while compression was performed to stop the bleeding. Bipolar cautery and bone wax were unsuccessful to control the bleeding. After an initial drilling of the mastoid cortex, it was identified that bleeding came from a laterally and anteriorly displaced sigmoid sinus, which had no cortical bone cover (extracranial sigmoid sinus). Bleeding was controlled by carefully placing layers of oxidized regenerated cellulose (Surgicel®, Johnson & Johnson Wound Management, a Division of Ethicon Inc., Somerville, NJ) on top of the exposed sinus without introducing them in the sinus lumen.

After achieving bleeding control, a simple mastoidectomy was performed in the regular fashion. A posterior tympanotomy was done and the cochlear implant electrode array was inserted through the round window membrane without other complications. No retroauricular hematoma or bleeding was observed during the postoperative period. The preoperative CT scan images were retrieved after surgery to evaluate if complications could have been avoided; they are shown in Fig. 5.

3. Discussion

A great number of ear surgeries starts with a retroauricular incision and a simple mastoidectomy. If a sinus pericranii, a petrosquamosal sinus or an extratemporal sigmoid sinus are present, they may be damaged during the most common initial steps of surgery, such as subcutaneous retroauricular anesthetic infiltration, the skin incision or at the beginning of the mastoid cortical bone drilling.

The first author to report the presence of a persistent petrosquamosal sinus was Moreau in 1929 [11] and its first radiological description was given by Marsot-Dupuch in 2001 [12]. In 2005, Koesling et al. retrospectively analyzed 223 high-resolution CT scans of the temporal bone to identify the occurrence of vascular anomalies. In this study, they noted a persistent petrosquamosal sinus in 1% of the analyzed temporal bones and an anterior and dehiscent sigmoid sinus in less than 1% [13]. Since apparently 1 in every 100 temporal bones may present with a patent petrosquamosal sinus or some form of sinus pericranii, ear surgeons should always perform a thorough revision of imaging studies with these vascular anomalies in mind. Attention should be paid to the fact that even though an extracranial sigmoid sinus is even more rare, in our series it was the most troublesome malformation encountered, causing a profuse bleeding in a 4-year-old patient.

Every ear surgeon must know how to deal with complications, such as the bleeding from emissary veins, that may result in the formation of a postoperative epidural hematoma [14,15]. Bone wax is a good alternative to stop the bleeding from a normal mastoid emissary vein or from an exposed area of dura, but it is not a good option when dealing with bleeding from large emissaries such as a persistent petrosquamosal sinus. Bone wax is also not recommended to stop bleeding from a sigmoid sinus tear. It is important to note that bone wax applied into bleeding emissary foramina has been known to enter the deeply placed venous sinuses and results in extensive thrombosis. The use of a muscle plug that does not obstruct the whole lumen of the sinus has also been recommended. Monopolar cautery coagulation of these venous channels may result in dural venous sinus thrombosis and consequent cerebral infarction. If coagulation is to be done in a small sinus tear or an
emissary vein, low current bipolar coagulation is recommended [16,17]. It is the authors’ opinion that placing a sufficient amount of oxidized regenerated cellulose (Surgicel®) layers on top of the bleeding vessel, slight compression and patience give the best results with minimal possibilities of complication. The use of absorbable gelatin sponge is also a good option, always remembering not to introduce it in the vascular lumen to avoid thrombosis.

The senior author in this article has performed over 1000 cochlear implantations together with a seemingly large number of mastoid surgeries for the treatment of other pathologies. Until now, he encountered the aforementioned vascular anomalies in only these three patients.

4. Conclusion

The persistent petrosquamosal sinus, the sinus pericranii in its different anatomical presentations and the extracranial sigmoid sinus are rare anatomical variations that the surgeon must have in consideration to avoid complications, which may be life threatening, during a retroauricular approach.

Conflicts of interest

The authors disclose no conflicts of interest.

References


