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

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ABSTRACT

Lateral and sigmoid sinus malformations are uncommon and dangerous anatomical variations that surgeons may encounter when performing a retroauricular approach. We report three cases of rare temporal bone venous sinus anomalies seen in patients who underwent cochlear implant surgery. The first patient had a diagnosis of CHARGE syndrome and presented a bilateral persistent petrosquamosal sinus with sigmoid sinus agenesis, which made mastoidectomy for cochlear implantation difficult. The second patient presented an anomalous venous lake in the occipital region, which communicated the left dural venous sinuses with a conglomerate of pericranial vessels in the left nuchal region, also consistent with left *sinus pericranii*. The third patient presented with an extracranial sigmoid sinus that produced a troublesome bleeding immediately after the muscular-periosteal flap incision was performed.

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

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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 retro-auricular 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

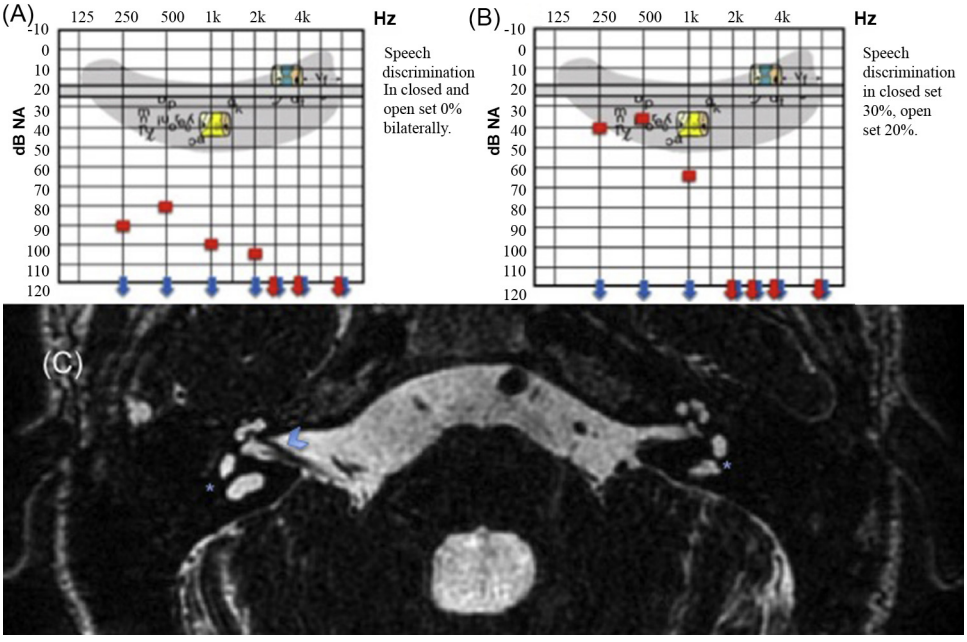

Fig. 1. Audiometric and MRI findings prior to cochlear implantation. (A) Audiometric results without hearing aid. The patient showed no speech discrimination. (B) Audiometric results with a well-adapted hearing aid in the right ear. Speech discrimination thresholds in closed and open set are below 50%. (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 hypoplasic cystic labyrinth bilaterally. dB = decibel, Hz = hertz.

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