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ORIGINAL RESEARCH

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Bifid Intratemporal facial nerves in an adult without other congenital anomalies

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Abstract

Objective: To describe a case of bifid intratemporal facial nerves without associated middle or inner ear abnormalities encountered on computed tomography (CT) imaging during preoperative preparation for unilateral cochlear implant placement in an adult male with profound sensorineural hearing loss (SNHL).

Methods: A rare case of bilateral bifid intratemporal facial nerves in an adult male is presented. The finding's impact on approach to safe cochlear implantation is discussed.

Results: Bifurcation of the intratemporal facial nerve is rarely seen and is usually associated with congenital middle or inner ear anomalies. A unique case of bilateral bifid intratemporal facial nerves without other middle or inner ear abnormalities was encountered incidentally on CT imaging during preparation for unilateral cochlear implant placement in an adult male with profound SNHL. The nerve was bifid along the mastoid segment with a nerve branch traversing through the facial recess precluding safe traditional approach to cochlear implant placement. Accessory stylomastoid foramina were noted bilaterally. Unilateral subtotal petrosectomy was performed with successful implantation and excellent hearing outcome. No additional clinical or radiographic otologic abnormalities were noted.

Conclusion: Abnormal bifurcation of the facial nerve may occur in adults without other middle or inner ear anomalies. This case highlights the importance of independent imaging review by the surgeon and vigilance to potential rare anatomic aberrations of the facial nerve during cochlear implantation.

Level of Evidence: IV

KEYWORDS
bifid, cochlear implantation, facial nerve

1 | INTRODUCTION

Bifid intratemporal facial nerve anomalies are rare and most commonly have been reported in patients with associated congenital

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hearing loss and external, middle, or inner ear abnormalities. Ossicular anomalies associated with bifid facial nerve anomalies include stapes footplate fixation[1,2] and absent stapes.[3,4] Other anomalies associated with bifid intratemporal facial nerves reported in the literature include oval window atresia with stenosis and semicircular canal dysplasia.[5] There have been few, if any, cases in the literature detailing bifid intratemporal facial nerve abnormalities in a patient without congenital hearing loss or other associated anatomic external, middle, or inner ear anomalies. This report will detail the case of a 50-year-old patient, with no known congenital otologic abnormalities, found to have bifid facial nerves during imaging workup for cochlear implantation after sudden onset sensorineural hearing loss (SNHL) resulting in profound hearing loss.

2 | MATERIALS AND METHODS

We present a unique case of an adult male found to have bilateral bifurcation of the intratemporal facial nerves without other associated external, middle, or inner ear anatomic or structural abnormalities. The patient's presentation, noteworthy imaging findings, and clinical and surgical course are described. A literature review was also performed to identify any similar cases. Report of this case was exempt from University of Tennessee Health Science Center institutional review board (IRB) approval due to meeting "not human subjects research" status by the IRB guidelines.

3 | RESULTS

A 50-year-old male with a complex medical history including human immunodeficiency virus, end-stage renal disease (ESRD) on hemodialysis, type II diabetes mellitus, and coronary artery disease initially presented with right worse than left bilateral hearing loss. His otologic history was notable for a right sudden SNHL 2 years prior to presentation and a left sudden SNHL 1 year prior to presentation. He reported multiple episodes of acute otitis media as a child that resolved with age and did not require pressure equalization tube placement. Hearing loss was associated with bilateral nonpulsatile tinnitus but no other otologic complaints.

The otologic exam was unremarkable. Audiometric evaluation showed moderate sloping to profound SNHL on the left, and severe sloping to profound SNHL across all frequencies on the right. Aided speech testing of the left ear showed 10% correct on AzBio Sentences in Quiet testing, so he was deemed a candidate for left-sided cochlear implantation. A noncontrast magnetic resonance imaging of the internal auditory canals was obtained due to the patient's history of ESRD and was unremarkable. A noncontrast fine-cut computed tomography (CT) of the temporal bones was obtained and, per radiology report, showed no abnormalities.

Upon review of imaging by the operating Otolaryngology team, the facial nerve was noted to be bifid starting at the proximal aspect of the vertical mastoid segments bilaterally. The images were further reviewed again with radiology and confirmed the nerve was bifid along the mastoid segment with a nerve branch traversing through the facial recess precluding safe traditional approach to cochlear implant placement (Figures 1–4). Accessory stylomastoid foramina

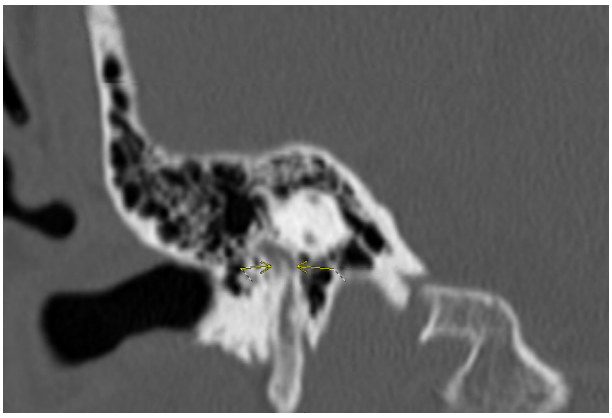

FIGURE 2 Coronal computed tomography temporal bones showing bifurcation of vertical segment of the right facial nerve through two separate bony canals.

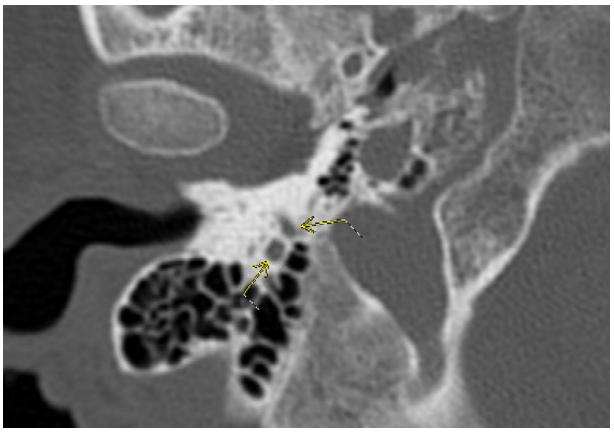

FIGURE 1 Axial computed tomography temporal bones showing bifurcation of vertical segment of the right facial nerve through two separate bony canals.

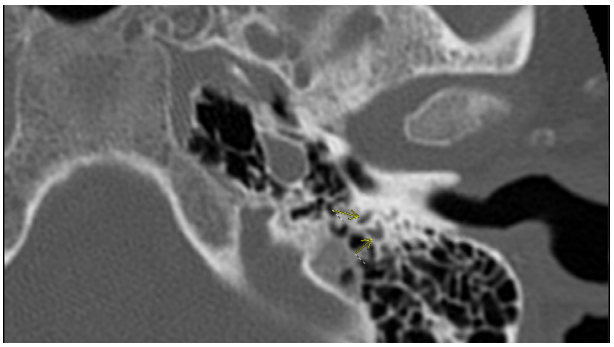

FIGURE 3 Axial computed tomography temporal bones showing bifurcation of vertical segment of the left facial nerve through two separate bony canals.

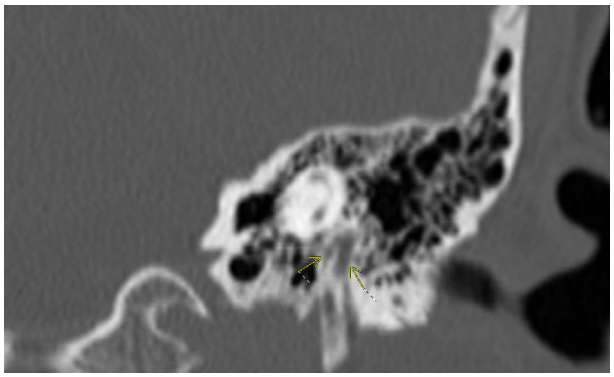

FIGURE 4 Coronal computed tomography temporal bones showing bifurcation of vertical segment of the left facial nerve with two separate bony canals.

were noted bilaterally providing further support for bifid course of the facial nerve. The labyrinthine and tympanic segments of the facial nerve appeared grossly within normal limits on imaging, and no other middle ear or inner ear abnormalities were appreciated.

The patient agreed to proceed with left cochlear implantation and ear canal closure. To avoid injury to the facial nerve on the operative side, a subtotal petrosectomy was performed to expose the round window with placement of a precurved electrode array (Advanced Bionics, Valencia, CA). A full insertion was achieved without meeting any resistance during placement. A local tissue transfer was used to rotate ear canal flaps for blind sac closure of the external auditory canal in a single stage. Intraoperative neural response imaging showed good responses in the basal, middle, and apical electrodes, and intraoperative imaging showed good electrode positioning.

The patient had an unremarkable postoperative course with implant activation ~1-month after implantation. At the 3-month postoperative appointment, the patient reported significant improvement in speech understanding in the implanted ear. Audiometric testing showed hearing thresholds of 25–30 decibel hearing level (HL) from 250 to 6000 Hertz (Hz) using warble tones and 72% using the Spanish AzBio Sentences in Quiet list number 12. He has continued regular follow ups with sustained use and benefit from his cochlear implant.

4 | DISCUSSION

The intratemporal facial nerve course is complex, having one of the longest intra-osseous courses of any cranial nerve, and is subject to occasional variations along its course that the otologist must respect.[6] Bifurcation of the facial nerve is one such rare anomaly that may be encountered. Most reports in the literature have noted bifurcation of the facial nerve within the tympanic segment of the facial nerve, often in association with middle, external, or inner ear anomalies.[1–3,5] The facial nerve develops embryologically from the second branchial arch along with the long process of the incus and the stapes suprastructure. Subsequently, aberrations in second branchial arch development can result in abnormal or absent stapes structures in addition to facial nerve anomalies including bifurcation. In a case series by Durcan et al., each of the six cases of unilateral or bilateral bifurcation of the intratympanic facial nerve encountered during middle ear operations involved concomitant abnormalities of the stapes crura or footplate ranging from hypoplastic stapes, absent stapes suprastructure, or stapes footplate fixation.[7] There has been a reported bifid tympanic facial nerve in conjunction with an abnormal long process of the incus and normal stapes as opposed to the more common association with stapes abnormalities as described above.[8] It is unclear whether this incus abnormality was acquired or congenital in the reported case, however. Accessory branching of the facial nerve can occur anywhere along its course, and there has been a report of a rare trifurcation of the mastoid segment of the facial nerve found in a 5-year-old boy, though the authors did not specifically note the status of the ossicular structures.[9]

An anatomic review of over 2000 temporal bone specimens by Marquet showed facial nerve abnormalities in 0.3% of otherwise normal temporal bone specimens with nerve ectasia, duplication, or trifurcation in 0.70% of specimens.[10] The standard approach to cochlear implantation surgery involves a posterior tympanotomy with the chorda tympani anteriorly and mastoid segment of the facial nerve posteriorly. The mastoid facial nerve is thus at particular risk of injury and can preclude optimal exposure of the round window. Historical reports have estimated the risk of facial nerve injury during cochlear implantation to be relatively low, ranging from 0.56% to 0.77%, though the risk is likely higher in patients with temporal bone and/or facial nerve anomalies.[11] A few studies have been published reporting facial nerve anomalies encountered during cochlear implantation. Raine et al. reported five cases of facial nerve anomalies precluding optimal exposure of the round window in a series of 42 cochlear implant operations, with one of the cases involving bifurcation of the facial nerve at the second genu. In that instance, the nerve was able to be decompressed and mobilized adequately to complete the cochlear implantation safely.[12] Song et al. similarly evaluated 947 adult and pediatric patients who underwent cochlear implantation and encountered major facial nerve anomalies in seven patients (0.7%). All seven of these patients were children with concurrent malformations of the cochlea and/or ossicles, and two patients had a bifurcated horizontal or vertical segment of the facial nerve.[13]

When an anomalous facial nerve course is encountered, a variety of surgical approaches may be used to keep the facial nerve safe while optimizing exposure of the round window and facilitating safe cochlear implantation. The traditional technique for safe implantation in cases with challenging anatomy, including facial nerve aberrations or congenitally malformed ears, is the subtotal petrosectomy with middle ear obliteration.[14] Such an approach was used successfully in our case. Other surgical approaches are sometimes required, however, and include a retrofacial approach, chorda tympani nerve rerouting, or a combined anterior–posterior approach.[13,15,16]

Facial nerve anomalies and associated cochlear malformations also create unique challenges for cochlear implant use and programming in the postactivation period, particularly in the pediatric population. A large retrospective review by Papsin evaluating implanted ears

with cochlear malformations found an increased incidence of facial nerve stimulation and more difficult programming requirements.[17] Cushing et al. similarly found high rates of facial nerve stimulation in their series with 78% of children with cochlear malformations showing inducible facial nerve stimulation.[18] Facial nerve stimulation and other nonauditory symptoms are also reported in the adult cochlear implant population, albeit more rarely.[19] The majority of these cases are successfully managed with cochlear implant remapping.[20] Research detailing cochlear implant outcomes and programming requirements in adults with concomitant cochlear malformations or facial nerve anomalies is much more limited, however, and our study adds to this literature.

This is the first reported case of bifurcation of the mastoid segments of the facial nerve bilaterally in an adult without any other external, middle, or inner ear abnormalities. It is important to note that this finding was discovered incidentally on preoperative imaging and was not initially noted by the reading radiologist, thus highlighting the importance of personally reviewing preoperative imaging studies. The surgical approach to cochlear implantation in this case was ultimately altered after review of imaging findings, and potential injury to the facial nerve during a standard facial recess approach to cochleostomy and cochlear implant placement was avoided by performing a subtotal petrosectomy instead.

5 | CONCLUSION

This case report highlights the importance of personally reviewing preoperative imaging studies and not depending on the radiological readings to identify potential rare otologic abnormalities that would impact surgical planning. The facial nerve course can be anomalous and exhibit bifurcation anywhere along its intratemporal course, even in patients without other associated otologic anatomic abnormalities, and it is important to note these abnormalities prior to operative intervention on the mastoid. In the case of a patient, such as ours, with bilateral hearing loss with normal external, middle, and inner ear anatomy, a facial nerve anomaly may not typically be expected. Due to concerns for unexpected anomalies impacting any planned surgical approach, the operating otolaryngologist may be better primed to recognize such abnormalities before the radiologist. Ultimately, the surgeon should utilize imaging findings and identify any anatomic variants prior to surgery to minimize injury to the facial nerve during cochlear implantation.

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