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Case Report

Cochlear Implantation in Patient with Arnold–Chiari Malformation

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We report a case of a 74-year-old patient with Arnold–Chiari syndrome (type 1) who, due to the bilateral profound hearing loss, was qualified for cochlear implantation. The difficult anatomy of the temporal bone necessitated special preparation for the procedure. The use of neuronavigation, in combination with facial nerve monitoring, enabled a safe surgical approach to the cochlea and ensured accurate placement of the implant.

KEYWORDS: Arnold–Chiari syndrome, cochlear implantation, neuronavigation, sensorineural hearing loss

INTRODUCTION

Arnold–Chiari syndrome is a rare congenital malformation occurring in approximately 1 in 1000 individuals, characterized by hypoplasia of the occipital bone and downward displacement of posterior cranial fossa structures, such as the cerebellar tonsils, into the spinal canal. Clinical symptoms typically arise from compression of the spinal cord or displaced brain structures. Additionally, disruption of cerebrospinal fluid (CSF) flow due to herniated brain tissue may contribute to the symptomatology.[1] Hearing loss is an uncommon manifestation of this condition and can be either unilateral or bilateral, with reported cases ranging from mild to profound hearing impairment.[2] Surgical management of the cranial bones in patients with Arnold–-Chiari syndrome presents significant challenges for the surgeon due to the anatomical complexity of the condition.

We present the case of a man with Arnold–Chiari syndrome who was qualified for cochlear implantation due to bilateral profound hearing loss.

Case Report

A 74-year-old man presented to the academic center with bilateral profound sensorineural hearing loss, bilateral impaired speech discrimination, and tinnitus in the right ear, which had gradually worsened over many years. The patient wore a hearing aid in the left ear but with limited gain. According to the patient, the cause of his hearing loss was most likely due to years of exposure to noise at work (he worked in construction and mining). The otoscopic examination didn't show any abnormalities. Pure tone audiometry showed bilateral sensorineural hearing loss: in the right ear of 100 dB, while in the left ear of 50 dB at frequencies 125-1000 Hz and 90 dB at 1500-8000 Hz. The patient was diagnosed with Arnold–Chiari syndrome type 1 based on a previous head magnetic resonance (MR) (the cerebellar tonsils were described to be intruded into the great aperture of the skull with preserved fluid reserve) and neurological consultation.

The patient was put on a qualification procedure for cochlear implantation. A computed tomography (CT) scan of the temporal bones was performed, along with consultations with a psychologist, speech therapist, and audiologist. The CT revealed hypoplasia

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of the mastoid process in the right temporal bone and aplasia of the mastoid process on the left side. Additionally, there was mild sclerotization observed in the incudomalleolar joints bilaterally. These findings contributed to the overall complexity of the case, requiring careful consideration for cochlear implantation (Figure 1). Auditory brainstem response testing ruled out a retrocochlear origin of the patient's deafness. After thorough evaluation of the imaging studies, audiological findings, and consultations with the multidisciplinary team, including a psychologist, speech therapist, and audiologist, the patient was considered a suitable candidate for cochlear implantation. The patient was qualified for cochlear implantation of the right ear.

The procedure began typically. During the antromastoidectomy, an anteriorly protruding sigmoid sinus and low descending dura of the middle cranial fossa were visualized. The mastoid process bone was solid and sclerotic. No air spaces within the mastoid process were encountered. The antrum and the lateral semicircular canal were not visualized. Following consultation with a second otosurgeon, it was decided to discontinue further stages of the procedure due to these anatomical challenges.

After re-examining the patient's imaging studies and their correlation with the anatomical situation in the surgical field, it was decided to make a second attempt to perform the procedure. The patient was advised to have another high-resolution CT adjusted for navigation.

The next surgery was performed. Navigation and a facial nerve sensor were used. The insight was significantly limited by an anteriorly protruding sigmoid sinus (Figure 2). The navigation system was useful here, making it much easier to find the antrum in solid, sclerotic, anatomically altered bone (Figure 3). The facial nerve sensor allowed safe drilling. The next steps of the procedure did not cause any problems, and the patient underwent successful implantation

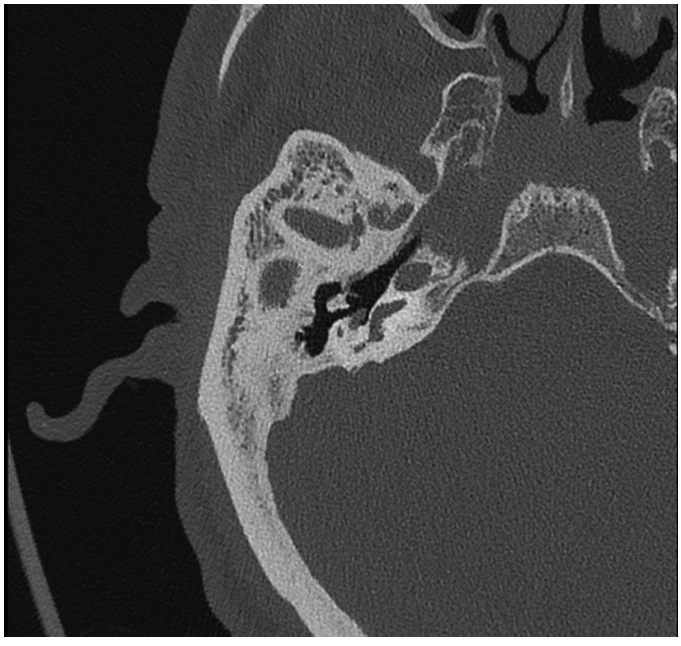

Figure 1. Right ear. Computed tomography scan of the temporal bone before first surgery.

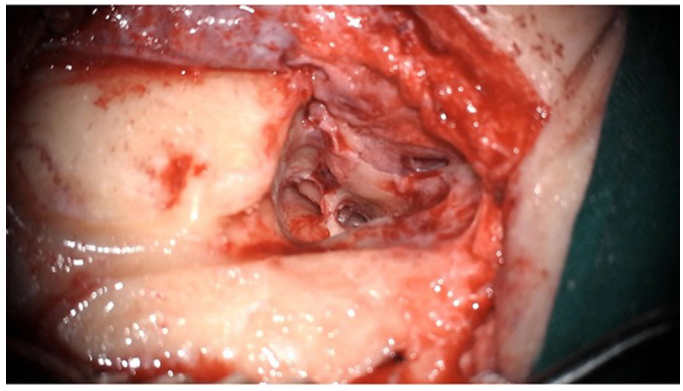

Figure 2. Right ear. Antromastoidectomy, posterior tympanotomy, cochleostomy, and bone bed are performed—surgical field is ready for the implant (protruding sigmoid sinus limits the view and approach to the cochlea).

with the nucleus 612 profile system (Figure 4). Facial nerve function remained normal. The wound healed without problems. The patient was discharged home on the fourth day after the surgery. The patient attends regular follow-up visits. He feels a great benefit associated with the use of the implant. He achieves 100% speech comprehension in tests, which occurs in a small group of patients.

The patient gave informed consent for all procedures performed during hospitalization, outpatient clinic visits, and analyzing and publishing medical data while maintaining anonymity.

DISCUSSION

Arnold–Chiari malformation is a congenital anomaly of the craniocervical junction, with an estimated prevalence of 1 in 1000 individuals. It involves underdevelopment or hypoplasia of the occipital bone, leading to herniation of the cerebellar tonsils and other posterior fossa structures through the foramen magnum into the spinal canal. Clinical manifestations arise from compression of the spinal cord, brainstem, or cerebellum, as well as from impaired CSF flow, which may result in hydrocephalus or syringomyelia due to altered CSF dynamics.[1]

Hearing loss as a symptom of Arnold–Chiari syndrome type 1 is rare. The most common sign of this condition is headaches in the occipital region that worsen when the pressure in the head increases.[3] Among the disorders related to hearing and balance, tinnitus, and dizziness are usually observed. Many theories could explain the occurrence of hearing loss in people with Arnold–Chiari syndrome. One of them is the suspicion of a stretch and damage to the eighth cranial nerve associated with the displacement of the posterior cranial structures. Another theory is pressure on this nerve or cochlear nuclei by displaced cerebellar tonsils. Other concepts also explain ischemic changes in the vestibulocochlear nerve and cochlear nuclei due to pressure on the vessels supplying blood to the brain through the spinal canal.[4] However, these theories have been challenged by Haktanir et al[4], who found no significant correlation between the extent of herniation of brain structures through the foramen magnum and the occurrence or severity of hearing loss in patients with Arnold–Chiari syndrome. Hearing loss associated with this condition can manifest as either unilateral or bilateral and typically worsens progressively over time. Sivakanthan et al[5] reviewed magnetic resonance imaging (MRI) scans of hearing-impaired patients and observed that some individuals exhibited

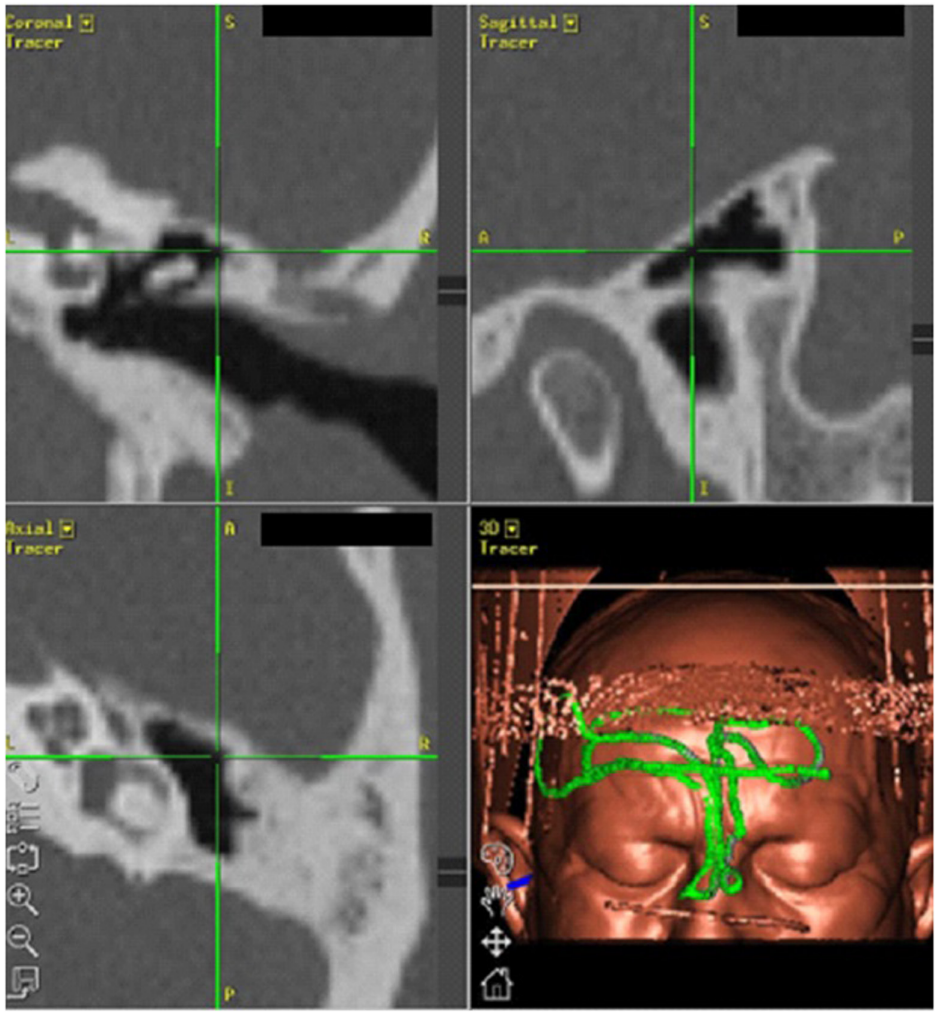

Figure 3. Antrum identification by neuronavigation.

herniation of hindbrain structures into the foramen magnum, even without a prior diagnosis of any related neurological disorder. In the present case, the bilateral sensorineural hearing loss was likely attributable to cochlear cell damage, potentially exacerbated by age-related degeneration and prolonged exposure to occupational noise.

Currently, there are 2 primary approaches for treating profound sensorineural hearing loss in patients with Arnold–Chiari malformation. One option is the decompression of the posterior cranial fossa structures that have herniated through the foramen magnum. This procedure typically involves a suboccipital craniotomy, laminectomy,

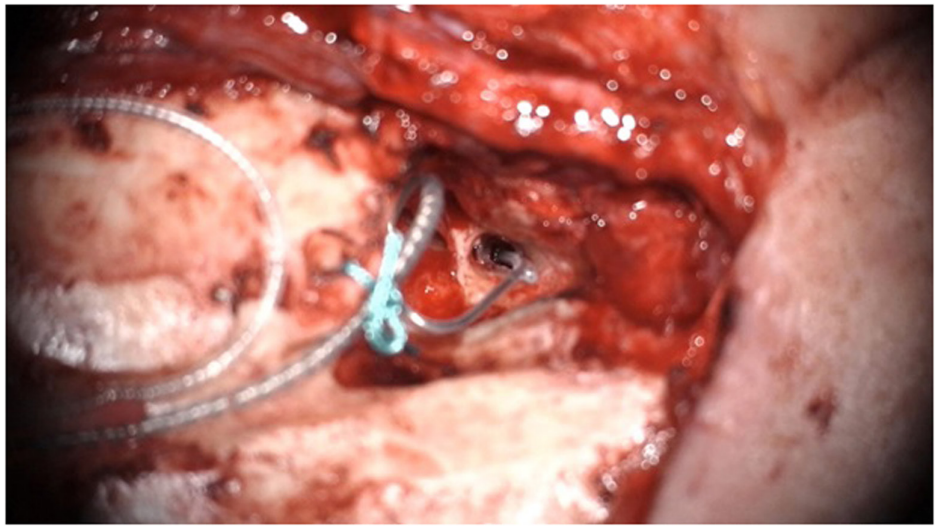

Figure 4. The intraoperative view after electrode array insertion.

and duraplasty to relieve pressure on the displaced brain tissues.[6] In some cases, this intervention leads to improvements in hearing, as well as reductions in tinnitus and vertigo. However, outcomes vary, with some patients experiencing only partial relief, such as the resolution of hearing loss or tinnitus alone.[7] Despite its potential benefits, this procedure carries the risk of several complications and remains controversial in the literature. Surgical decompression is thought to be effective primarily when brainstem or vascular compression is a significant factor in the pathophysiology of hearing loss, a determination that is challenging to confirm preoperatively.[8]

The second option for treating profound sensorineural hearing loss in patients with Arnold–Chiari syndrome is cochlear implantation. This approach is considered in individuals who have significant hearing loss but do not exhibit other severe neurological symptoms. Prior to surgery, it is crucial to confirm the integrity of the auditory pathway to ensure the potential effectiveness of the implant. To date, only 2 cases of cochlear implantation in patients with Arnold–Chiari malformation have been documented in the medical literature—1 in a patient with type 1 and another with type 2 malformation.[9,10] In both cases, no significant intraoperative complications or obstacles were reported by the surgeons.

We also opted for cochlear implantation in our patient. Given the challenging anatomy of the temporal bone, the procedure necessitated the use of neuronavigation, which, in conjunction with facial nerve monitoring, ensured safe access to the cochlea and precise placement of the implant. This approach resulted in a successful outcome, restoring significant hearing function and effectively reintegrating our patient into the auditory world.

CONCLUSION

Defects in cranial structures remain a surgical challenge, but they typically do not present a contraindication for cochlear implantation. The success of such procedures hinges on meticulous preoperative planning, which involves a thorough analysis of imaging studies to assess the patient's unique anatomy. Utilizing advanced tools like neuronavigation and facial nerve monitoring plays a crucial role in enhancing spatial orientation within the complex surgical field, ensuring the safe and accurate placement of the cochlear implant.

Availability of Data and Materials: The data that support the findings of this study are available on request from the corresponding author.

Informed Consent: Written informed consent was obtained from the patient who agreed to take part in the study.

Peer-review: Externally peer-reviewed.

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