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Successful Surgical Management of Large Cerebellar Cholesteatoma Involving Brainstem and Cranial Nerves: A Case Report

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Abstract

Background:: *Intracranial cholesteatomas are rare, non-neoplastic lesions that typically originate in the temporal bone but can extend into the posterior fossa. Their occurrence in the posterior fossa is extremely rare and presents significant surgical challenges due to their potential to compress vital structures such as cranial nerves and vascular components. These lesions can lead to neurological deficits, and their management requires careful preoperative planning and precise surgical techniques.*

Case Summary: *A 39-year-old male presented with progressive left-sided hearing loss, headaches, fever, and neck rigidity. MRI imaging revealed a large cholesteatoma compressing the cerebellum and several cranial nerves. The patient underwent a retrosigmoid approach for tumor resection. Intraoperative findings included involvement of cranial nerves V, VII, VIII, X, XI, and XII, as well as significant compression of the AICA, PICA, and basilar artery. The tumor was friable and easily dissected, allowing for complete resection without complications such as bleeding or cerebrospinal fluid leaks.*

Conclusion: *This case highlights the importance of early diagnosis, careful preoperative imaging, and meticulous surgical planning in the management of intracranial cholesteatomas. The retrosigmoid approach provided optimal access for complete tumor removal while preserving critical neural and vascular structures. Key clinical takeaways include the necessity of a multidisciplinary approach and the careful preservation of function in complex posterior fossa surgeries.*

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Introduction

Definition

"Cholesteatoma" is a well-demarcated non-neoplastic lesion in the temporal bone, which is commonly described as "skin in the wrong place." [1]

History and Etymology

Joseph-Guichard Duverney, a French anatomist, was the first to describe a temporal bone lesion in 1683, probably representing a cholesteatoma [2]. In 1838, this pathology was named "cholesteatoma" (Greek: chole = bile, stear = fat, -oma = tumor) by the German anatomist/pathologist Johannes Müller [3]. However, this term is incorrect because the lesion does not contain fat and is not of a neoplastic nature. Although other more descriptive denominations were suggested, such as "pearl tumor," "margaritoma," or "keratoma," the most commonly used expression is the misnomer "cholesteatoma." [3]

Epidemiology

The annual incidence of cholesteatoma is reported as 3 per 100,000 in children and 9.2 per 100,000 in adults with a male predominance of 1.4:1 [4]. Middle ear cholesteatomas have a higher incidence in individuals younger than 50 years of age, whereas EAC cholesteatomas present predominantly at 40–70 years of age [4]. Hereditary predisposition is probable. There is a high prevalence among white individuals, and cholesteatoma is rarely detected in the Asian, American Indian, and Alaskan Eskimo populations [4].

Acquired cholesteatomas may develop by various etiopathogenic mechanisms:

- The "migration theory" postulates relocation of squamous epithelium from the margin of a perforated or retracted tympanic membrane (TM) into the middle ear, forming a cholesteatoma [5].
- The "basal hyperplasia theory" assumes an inflammation-associated proliferation of basal cells breaking through the basement membrane, thus giving rise to a cholesteatoma [6].
- The "postsurgery/posttraumatic theory" claims iatrogenic implantation of epidermal elements into the middle ear cavity [7].
- According to the "retraction pocket theory," the main trigger for cholesteatoma formation is poor ventilation of the mastoid cavity and the middle ear as a result of eustachian tube dysfunction. This leads to an increased negative pressure in the tympanic cavity, resulting in retraction of the TM with invagination of part of it, usually the pars flaccida. Chronic retraction pockets may facilitate hyperplastic epidermal ingrowths into the middle ear with consecutive development of granulation tissue and bone erosion [8]. On the basis of this mechanism, cleft palate [9] and poorly pneumatized mastoids (underdeveloped or as a consequence of chronic inflammation) [10] are associated with a higher risk for cholesteatoma formation. The developing cholesteatoma may cause secondary perforation in the TM. It is not known why some retraction pockets are transformed into cholesteatomas and others are not; an associated inflammation leading to hyperkeratinization has been suggested to play a role [11].

- The "metaplasia theory" is based on the identification of mucous glands in the inflamed connective tissue taken from cholesteatomatous ears [12]. However, various studies [13,14].revealed that acquired cholesteatomas originate from the squamous epithelium (ectoderm) of the EAC and the bordering TM, rather than from metaplasia of the cuboidal epithelium (endoderm) of the middle ear. For the development of idiopathic EACC, rudiments of the first branchial cleft or dissemination of the germinal epithelium might be responsible. Repeated microtrauma (cotton-tipped applicators, hearing aids) and diminished microcirculation (e.g., from smoking) might be risk factors as well [15].

Secondary EACCs occur at the site of idiopathic stenosis or narrowing of the EAC due to a lesion (such as an osteoma, exostosis, nevus, or mycetoma) or following trauma, surgery, inflammation, or radiation therapy [16].

Case Presentation

Patient Information

A 39-year-old male from Minsk presented with symptoms beginning in April 2024. His initial complaints included headaches associated with fever and neck

rigidity. Notably, he denied any issues with balance, facial palsy, facial pain, voice changes, visual disturbances, limb weakness, taste deficits, facial asymmetry, or disturbances in bowel or bladder function. The only significant symptom was progressive hearing loss in the left ear.

Medical History

- No prior similar episodes.
- Recurrent sinusitis, typically associated with febrile episodes.
- No history of previous surgeries.
- No known chronic diseases.

Family History

- Father diagnosed with Alzheimer's disease in 2022.
- No known family history of tumors or malignancies.

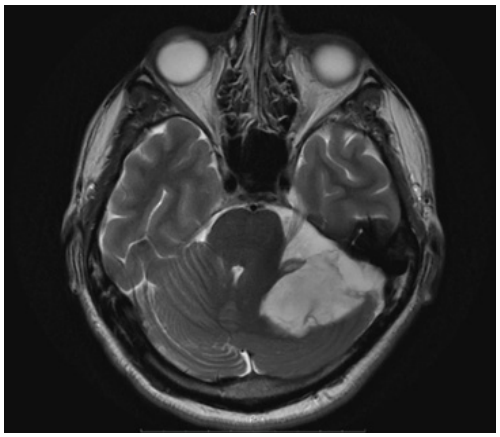
Social History

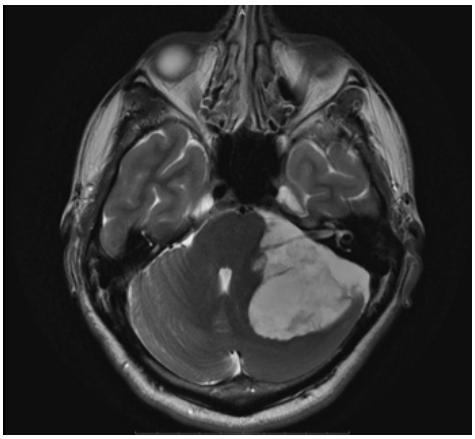
- No history of smoking or alcohol consumption.
- No occupational exposure to loud noise.

Diagnostic Workup

An MRI performed on December 18, 2024, revealed a lesion consistent with cholesteatoma.

Imaging

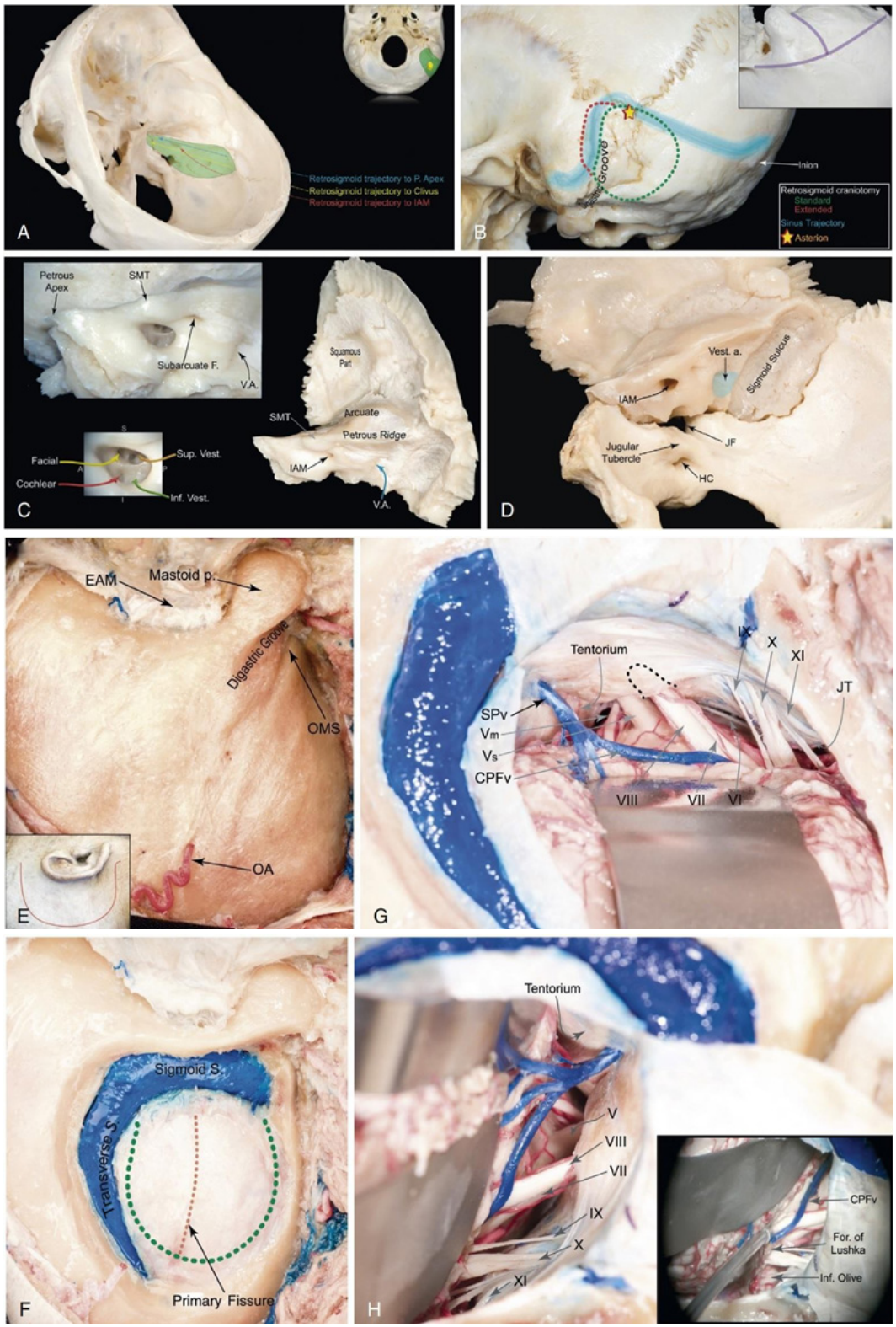




Surgical Procedure Approach

The Retrosigmoid Approach

The retrosigmoid approach [32] is a variation of the suboccipital craniotomy that is designed to provide optimal access to the cerebellopontine and cerebellomedullary cisterns and the posterior aspect of the cerebellopontine angle (Fig. 3.2A). The retrosigmoid approach uses a lateral suboccipital craniotomy combined with a partial mastoidectomy to enter the superior aspect of the posterior fossa in the dorsolateral compartment. This approach is best used to access tumors of the cerebellopontine angle, which, while having their epicenters posterior to the lower CNs, may infiltrate superiorly to the middle incisural space, laterally to the internal acoustic meatus, or medially to the lateral aspect of the pons or into the cerebellar hemisphere. This approach also provides an exposure to aneurysms of the AICA, the proximal segment of the PICA, the basilar trunk, and vascular compression of the trigeminal nerve. When planning the surgical strategy for a particular case, the retrosigmoid approach may be weighed against the far lateral, the endoscopic endonasal, and the transmastoid approaches. A useful rule to maximize extent of resection while staying in the safe zone is to design the surgical strategy around the concept of “not crossing the nerves.” Consequently, the neurosurgeon should consider all the surgical approaches that may be used for a particular lesion and their potential combinations (360-degree approach to the lesion).

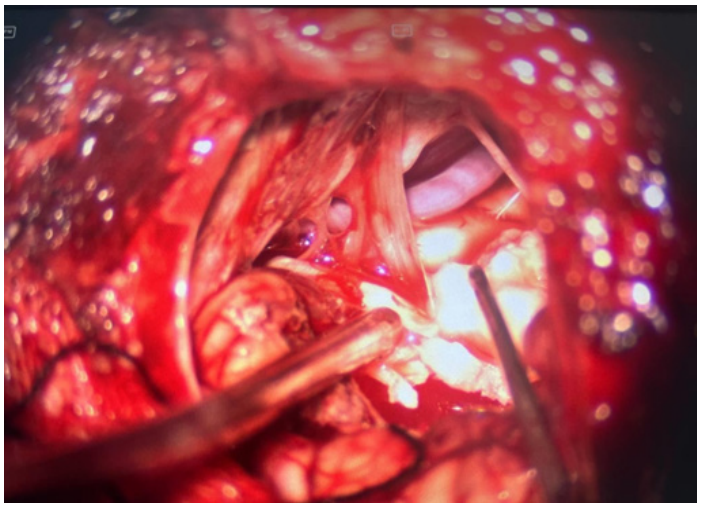


(A) Surgical corridor of (green areas) and dissection trajectories available during (lines) the retrosigmoid approach. Upper right, yellow arrow shows the line of view from the surgeon's perspective. IAM, Internal acoustic meatus; P. Apex, petrous apex. (B) Lateral suboccipital (classical) craniotomy (green dotted circle) and extended transmastoid retrosigmoid craniectomy (red dotted line); star, asterion. Venous flow through the transverse (horizontal portion) and sigmoid ("S" portion) sinuses is shown by the blue line. Upper right, Inferred location of the asterion during the skin incision (purple lines). (C) Right, the internal acoustic meatus (IAM) is located at the middle third of the petrous bone in its posterior fossa surface, between the suprameatal

tubercle (SMT) and the vestibular aqueduct (V.A.). Upper left, Close-up view showing the relationship of the IAM to the SMT and the subarcuate foramen (Subarcuate F.). Lower left, Locations of the facial, superior vestibular (Sup. Vest.), cochlear, and inferior vestibular (Inf. Vest.) nerves. (D) The jugular tubercle serves as both the roof of the hypoglossal canal (HC) and the floor of the jugular foramen (JF). IAM, Internal acoustic meatus; Vest. a., vestibular aqueduct. (E) C-shaped skin incision (red line, lower left) includes the planned craniotomy area and stops below the tip of the mastoid process (Mastoid p.). EAM, External acoustic meatus; OA, occipital artery; OMS, occipitomastoid suture. (F) For the semicircular dural incision (green dotted arc), a dural flap is raised in a semicircular fashion in the same orientation as the skin flap. Bone removal is started at the posterior edge of the transition from the transverse sinus (Transverse S.) to the sigmoid sinus (Sigmoid S.). (G) Dissection of the cerebellopontine cistern exposes the motor (Vm) and sensory (Vs) roots of cranial nerve V, as well as cranial nerves VI through XI, the cerebellopontine fissure vein (CPFv), the jugular tubercle (JT), and the superior petrosal vein (SPv, also called the Dandy vein). Dotted line shows the drilling area for accessing the internal acoustic canal. (H) The cerebellomedullary cistern provides access to cranial nerves V, VII, VIII, and IX to XI. Lower right, The inferior olive (Inf. Olive) and the foramen of Lushka (For. of Lushka) are exposed near the cerebellopontine fissure vein (CPFv). (Used with permission of the University of California San Francisco's Skull Base & Cerebrovascular Laboratory.)

Intraoperative Findings

After opening the dura and carefully retracting the tentorium, we observed a cholesteatoma compressing the cerebellum. During tumor removal, we meticulously identified cranial nerves V, VII, and VIII. The tumor was friable with a cheesy texture, allowing for easy dissection.



Due to its considerable size, we extended our exposure by following the tumor downward toward the medulla, where we identified the Anterior inferior cerebellar artery (AICA), posterior inferior cerebellar artery (PICA), and, at a greater distance, the basilar artery. Additionally, cranial nerves X, XI, and XII were visible, and the tumor was compressing and involving these structures. However, the tumor was easily dissected from the nerves without significant resistance.

Surgical Techniques

After cutting the dura, we began the procedure using a microscope for enhanced visualization. The tentorium was gently retracted, and dissection of the tumor was initiated. Due to the tumor's location, we performed meticulous layer-by-layer dissection to ensure complete removal while avoiding damage to surrounding structures. The tumor's friable texture facilitated easy dissection, and removal was achieved without significant difficulty. Throughout the surgery, there was no intraoperative bleeding or cerebrospinal fluid (CSF) leaks.

Outcome of Surgery

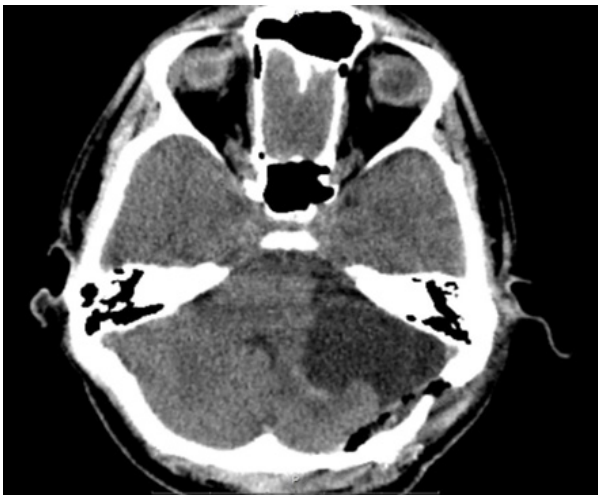
- Degree of Resection: The tumor was completely removed.
- Immediate Postoperative Complications: There were no postoperative complications.
- Postoperative Care: The patient was transferred from the ICU to the general ward (palate) one day after surgery.

Postoperative Management

- Post-Op Neurological Examination: The patient underwent a thorough neurological examination postoperatively, which revealed no deficits. All cranial nerve functions were intact, and the patient showed appropriate motor and sensory responses.
- Management in the Neuro-Intensive Care Unit (ICU): The patient was closely monitored in the neuro-ICU for 24 hours following surgery. Vital signs and neurological status were regularly assessed to ensure stability, with no immediate complications arising.
- Use of Steroids, Antibiotics, or Anticonvulsants: The patient was administered steroids for postoperative edema, antibiotics to prevent infection, and no anticonvulsants were necessary as there were no signs of seizures.

Imaging

A CT scan was performed the day after surgery to assess the surgical site and ensure there were no immediate complications, such as hematoma or edema.

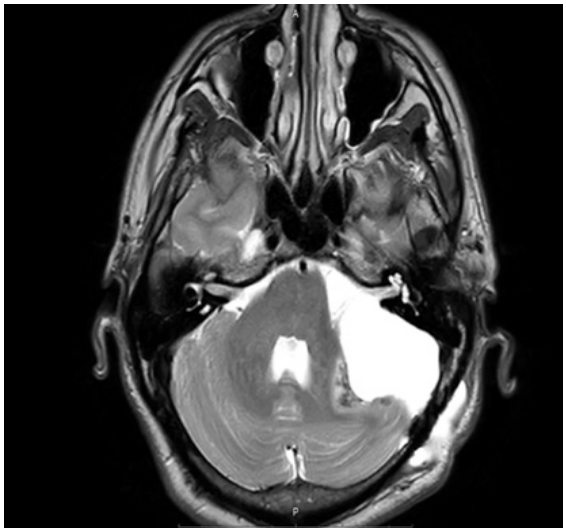


Recovery

The patient showed progressive improvement following surgery. Neurological function remained intact, and there were no neurological deficits observed. No specific rehabilitation measures were required immediately post-surgery, as the patient's recovery was uncomplicated. However, regular neurological assessments and monitoring of cranial nerve function were conducted to ensure continued recovery.

Outcome and Follow-Up

- After 1 month, the patient reported improvement in hearing, which had been the primary issue post-surgery.
- Neurological Recovery Status: The patient's neurological recovery has been progressing well, with no deficits or complications. There were no signs of weakness, sensory loss, or cranial nerve abnormalities.
- Imaging Follow-Up: An MRI was performed, showing that the compressed areas are gradually returning to their normal anatomical configuration, indicating effective healing and no signs of tumor recurrence.



Functional Outcomes

- Balance: The patient reported gradual improvement in balance, with no signs of ataxia or unsteadiness during follow-up assessments.
- Cranial Nerve Function: Cranial nerve functions remained normal, with no deficits in facial movements, taste, or sensation. There was a marked improvement in hearing, which had been the primary concern post-surgery.
- Motor Strength: Motor strength was preserved throughout recovery, and the patient showed no signs of weakness or paralysis in any limbs. Coordination and strength in both upper and lower

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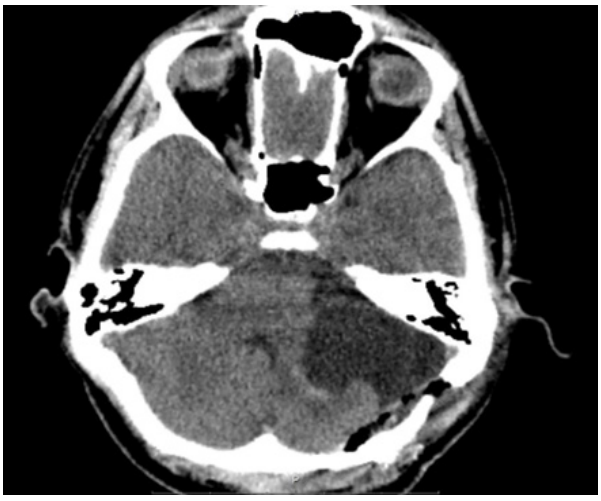

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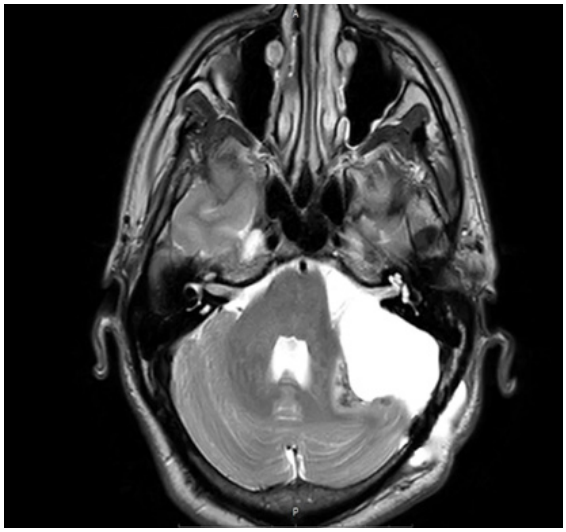

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extremities were within normal limits.

Discussion

Literature Review

Intracranial cholesteatomas, although rare, can occur in the cerebellopontine angle and involve the brainstem and cerebellum [17]. These tumors are typically characterized by a slow-growing, expansile mass that can compress adjacent structures, including cranial nerves, the brainstem, and cerebellum [17,18]. The most common symptoms include sensorineural hearing loss, cranial nerve deficits, and cerebellar signs such as ataxia [18].

The literature emphasizes that early diagnosis is crucial, as these tumors are often misdiagnosed due to their slow growth and non-specific symptoms [19]. Surgical resection is the treatment of choice, with complete tumor removal being the goal to prevent recurrence and neurological deterioration [17,19]. However, the proximity to vital structures like the brainstem and cranial nerves makes surgery challenging. Several studies have highlighted the importance of microsurgical techniques, including the use of operating microscopes and neuronavigation, to minimize damage to these critical structures [20,21].

When involving the brainstem and cerebellum, these tumors often require more complex surgical approaches with a higher risk of complications, including cranial nerve injury and neurological deficits [22]. Long-term prognosis largely depends on the extent of resection and the preservation of cranial nerve function, with studies suggesting favorable outcomes when complete resection is achieved and no significant postoperative complications arise [18,21].

Surgical Challenges and Techniques

Operating near the brainstem and cranial nerves presents significant challenges due to the delicate and vital nature of these structures [22]. The proximity of the tumor to the brainstem and cerebellum increases the risk of neurological damage, particularly to cranial nerves that control essential functions like vision, hearing, facial movement, and swallowing [22,23]. Dissecting a cholesteatoma in this region requires meticulous planning and technical precision, as nerve preservation is paramount to avoid permanent deficits [21,23].

Neuromonitoring plays a crucial role during these surgeries, allowing real-time assessment of cranial nerve function and motor pathways [24]. This technique guides the surgeon in avoiding inadvertent damage to nerves or the brainstem. Somatosensory evoked potentials (SSEPs), motor evoked potentials (MEPs), and auditory evoked potentials (BAEPs) are typically used to monitor the integrity of the nervous system throughout the procedure [25].

Careful dissection is essential, particularly in separating the tumor from cranial nerves V, VII, VIII, IX, X, XI, and XII, which may be involved in the pathology [22]. The use of high-magnification microsurgical techniques ensures that the tumor is removed layer by layer while minimizing trauma to surrounding tissues [21,23]. Fine microsurgical instruments and the operating microscope enable precision in dealing with fragile neural and vascular structures.

Hemostasis is another critical aspect of the surgery. Although bleeding was not encountered in this case, vascular control must be meticulously achieved, especially near the basilar artery, PICA, and ICA, which may lie adjacent to the tumor [26]. Proper handling of blood vessels and hemostatic agents helps minimize blood loss and prevent postoperative complications such as hematomas or CSF leaks [26].

Comparison with Other Case Reports

This case of intracranial cholesteatoma involving the brainstem and cerebellum shares similarities with other reported cases in terms of its slow-growing nature, presentation with hearing loss, and involvement of cranial nerves [17,18,22]. However, it also presents several unique aspects that distinguish it from previous reports.

Many published reports describe cholesteatomas that predominantly affect the cerebellopontine angle, often compressing cranial nerves V, VII, and VIII [27]. In contrast, this case involved multiple cranial nerves (V, VII, VIII, X, XI, and XII), with compression of both the cerebellum and brainstem, highlighting the extensive reach of the tumor and its potential for causing more severe neurological deficits [22,28].

The friable and cheesy texture of the tumor, which allowed for relatively easy dissection, is also noted as uncommon, as many cholesteatomas have a firmer consistency that complicates removal [28].

Additionally, this case stands out due to the successful complete resection without intraoperative complications such as bleeding or CSF leaks, which are often reported in similar surgeries due to the proximity to major vessels like the basilar artery and PICA [26,29]. The use of microsurgical techniques, neuromonitoring, and careful dissection contributed to the favorable outcome, with particular emphasis on nerve preservation.

In terms of post-surgical recovery, this case demonstrated rapid improvement, particularly in hearing, which is often a slower process in other reports [30]. The absence of postoperative neurological deficits further distinguishes this case as a positive outcome, compared to cases with long-term deficits or need for prolonged rehabilitation [31].

Lessons Learned

This case highlights several important lessons for managing intracranial cholesteatomas involving the brainstem and cerebellum:

- Early Diagnosis: Early recognition and timely diagnosis of cholesteatomas, particularly those affecting the cerebellopontine angle and adjacent structures, are crucial for a successful outcome [19]. Advanced imaging (MRI and CT) plays a key role in confirming the diagnosis and planning treatment.
- Multidisciplinary Management: A team approach involving neurosurgeons, otolaryngologists, radiologists, and neurologists is essential for comprehensive care [20]. In this case, interdisciplinary collaboration facilitated optimal surgical planning and cranial nerve preservation.
- Microsurgical Precision and Neuromonitoring: The success of this surgery underscores the value of microsurgical techniques and intraoperative neuromonitoring in identifying and preserving vital neural structures [21,24].
- Nerve Preservation: Careful dissection to preserve cranial nerve function is paramount. while complete resection is important, protecting nerves that control essential functions such as hearing and swallowing is critical for minimizing postoperative deficits [18,25].
- Postoperative Care and Follow-Up: Regular imaging and clinical follow-up are essential for detecting recurrence and ensuring recovery [31]. In this case, smooth postoperative recovery and favorable imaging confirmed the success of the intervention.

Conclusion

This case highlights the successful management of a large intracranial cholesteatoma involving the cerebellum, brainstem, and multiple cranial nerves. The patient underwent a complete tumor resection with no postoperative complications, demonstrating the importance of early diagnosis and precise surgical planning. The use of microsurgical techniques, neuromonitoring, and careful dissection played a crucial role in preserving cranial nerve function, particularly in a challenging surgical area near critical structures such as the brainstem and vascular supply.

The key lessons from this case include the significance of multidisciplinary collaboration, the importance of nerve preservation, and the necessity of careful postoperative monitoring. Timely intervention and personalized surgical approaches were pivotal in achieving a favorable outcome, with the patient showing significant improvement in hearing and neurological recovery.

This case underscores the importance of meticulous surgical planning, neuromonitoring, and postoperative care in achieving favorable outcomes for patients with complex cerebellar cholesteatomas. These elements are critical in minimizing risks and ensuring optimal recovery.

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