BMC Geriatrics

CASE REPORT Open Access

Brain with coexistent acoustic schwannoma and ependymoma

Haofuzi Zhang[1], Peng Zou[1*], Wei Lin[1] and Xiaofan Jiang[1]

Abstract

Background This particular case is a world-first with no previous literature reports on patients presenting with both benign acoustic schwannoma and malignant ependymoma. Case presentation: A 60-year-old woman with unexplained right-sided hearing loss that had worsened progressively over 4 years, along with intermittent dizziness that had begun 3 years prior. Our preliminary diagnosis included: (1) Right acoustic neuroma; (2) Ependymoma of the fourth ventricle; and (3) Hydrocephalus. We employed the right sigmoid sinus posterior approach combined with the posterior median approach, beginning with removal of the fourth ventricle tumor and then proceeding to acoustic schwannomas resection through rotating operation positions. Conclusions: The case presented significant challenges owing to: The difficulty encountered in arriving at a diagnosis; The difficulty in choosing a suitable surgical approach; The complexity of the surgical sequence; The intricacy of the surgical process. It's rare, complex, and had excellent surgical results.

Keywords Acoustic schwannoma, Ependymoma, Diagnosis, Surgical process

Background

A 60-year-old woman presented at our outpatient clinic seeking medical attention for unexplained right-sided hearing loss that had worsened progressively over 4 years, along with intermittent dizziness that had begun 3 years prior. This case represents a world-first, as there are no previous reports in the literature of a patient presenting with both benign acoustic schwannoma and malignant ependymoma. The final pathological results confirmed our initial diagnosis. It is crucial to consider the possibility of concurrent acoustic schwannoma and ependymoma when encountering tumors with differing textures in the posterior fossa. We hope this rare case report will capture the attention of scholars.

*Correspondence:
Peng Zou
446844165@qq.com
[1]Department of Neurosurgery, Xijing Hospital, Fourth Military Medical University, Xi'an 710032, China

Case presentation

Physical examination revealed decreased hearing on the right side, while laboratory test results were negative.

Figure 1 shows the patient's enhanced MRI before and 72 h after surgery. The preoperative MRI indicated patchy mixed signals in the right cerebellopontine angle (CPA) area, leading to a consideration of acoustic schwannoma; multiple cystic long T1T2 signals were also present in the fourth ventricle, suggesting ependymoma and supratentorial ventricular system expansion.

Our preliminary diagnosis included: (1) Right acoustic neuroma; (2) Ependymoma of the fourth ventricle; and (3) Hydrocephalus.

We developed a thorough surgical plan, beginning with the patient in a supine position for external drainage of the brain ventricle to prevent brain hernia formation. The patient was then placed in a left prone position, and the head was fixed with a three-nail head brace. We employed the right sigmoid sinus posterior approach combined with the posterior median approach,

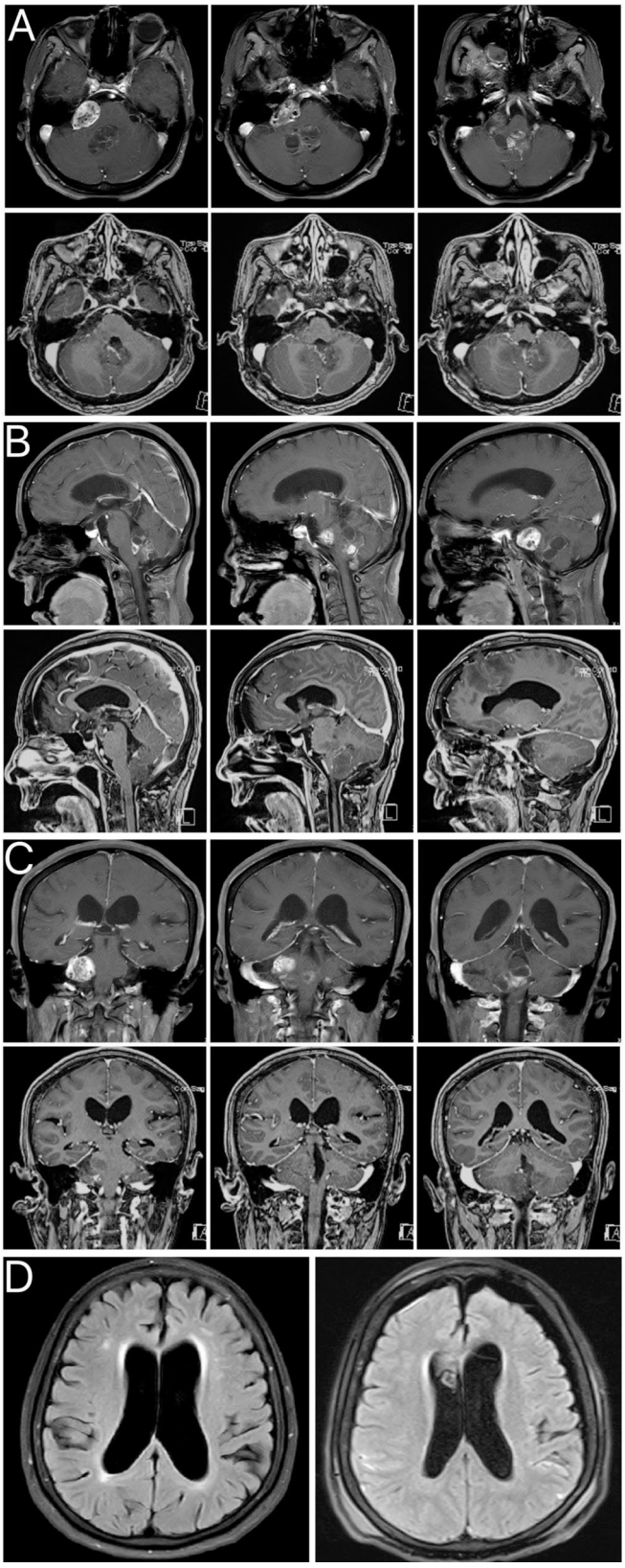

Fig. 1 Enhanced MRI before and 72 h after surgery. **A** Axial view. CPA and IV ventricle existed abnormal masses, with different signals. CPA region tumor invaded the internal auditory canal. **B** Sagittal view. IV ventricle tumor was closely related to the midbrain aqueduct. **C** Coronal view. These two tumor were completely removed. **D** Postoperative hydrocephalus was relieved compared with preoperative hydrocephalus

beginning with removal of the fourth ventricle tumor and then proceeding to acoustic neuroma resection through rotating operation positions (Supplementary Figure S1 A-B). During the intraoperative resection of the fourth ventricular tumor, we prioritized the protection of the dorsal side of the brainstem. Electrophysiological monitoring was employed to accurately identify the location of the facial nerve during the removal of the vestibular schwannoma, ensuring its preservation. Furthermore, we carefully removed the posterior wall of the internal auditory canal to thoroughly explore the tumor within. To prevent postoperative swallowing difficulties and choking on drinking water, we took extra precautions to protect the glossopharyngeal and hypoglossal nerves. Additionally, we implemented the use of a gastrointestinal tube to provide necessary nutritional support during the postoperative period. Furthermore, tracheal intubation was postponed until the patient's consciousness was fully restored.

The surgical procedure was executed smoothly (Supplementary Video), and the final pathological results confirmed our initial diagnosis (Supplementary Figure S1 C-D). There are two specimens of the brain tumor. One specimen's pathological results showed a right-sided acoustic schwannoma, with immunohistochemical results positive for S100 (+), SOX10 (+), H3 K27me3 (+), and Ki-67 (+, < 3%). The other specimen's pathological results showed a fourth ventricle ependymoma, with immunohistochemical results positive for GFAP (+), Vimentin (+), ATRX (+), and Ki-67 (+, 2%). The patient experienced significant relief from hydrocephalus and was discharged ten days after the operation (Fig. 1). And the patient consented to the publication of her image. We have included postoperative MRI images taken at 3 months (Supplementary Fig. 2) to provide additional visual information. During a telephone follow-up, the patient reported a return to a normal lifestyle. Furthermore, we have added a supplementary figure showcasing postoperative facial expressions. According to the House-Brackmann classification of facial nerve palsy, this patient was classified as level 2 (Supplementary Fig. 3).

Discussion and conclusion

This particular case is a world-first with no previous literature reports on patients presenting with both benign acoustic schwannoma and malignant ependymoma. The case presented significant challenges owing to:

The difficulty encountered in arriving at a diagnosis. While some experts believed that the lesion could be attributed to a tumor of the same nature originating from the middle cerebellar peduncle between the fourth ventricle and the CPA area, the final pathological results contradicted this notion.

The difficulty in choosing a suitable surgical approach. Although some authorities believed that the surgery should be performed in batches, we chosed to complete the two highly complex neurosurgery operations at once.

The complexity of the surgical sequence. To prevent supratentorial brain hernia, we opted to perform extraventricular drainage first and then resect the fourth ventricle tumor instead of the cerebellopontine angle tumor. This approach was because removing the fourth ventricle tumor was more conducive to the release of cerebrospinal fluid from the cistern magnum and, in turn, exposing the cerebellopontine angle tumor.

The intricacy of the surgical process. When resecting the tumor of the fourth ventricle, there was a risk of damaging the medulla oblongata, which was the center of life. Such damage could have endangered the patient's life. The removal of the acoustic schwannoma, according to Samii classification [1], was a type 4a case, and we removed the tumor within the internal auditory canal.

Several previously reported cases have emphasized the association between CP angle Schwannomas and meningiomas. In January 2024, Neupane et al. [2] documented the case of a 20-year-old female who presented with right CPA (9th/10th cranial nerve) schwannomas as well as a meningioma in the left anterior cranial fossa. Another study by Tao et al. [3] described a 17-year-old female patient with acoustic neuromas and a solid-cystic lesion in the cervical and thoracic spinal cord. Pathological examination confirmed the presence of ependymoma in the spinal cord and acoustic schwannoma. The case we are reporting is exceptionally rare, as the patient exhibited both a benign vestibular schwannoma of the CPA and a malignant ependymoma of the fourth ventricle, distinguishing it from previously documented cases.

The case we have reported does have certain limitations. One possible connection between the CPA and fourth ventricle tumors could be the invasion of the foramina of Luschka, a channel that connects these regions. However, due to the lack of sophisticated instruments available currently, it is challenging to detect and confirm the exact link between these two tumors. Pathological examination remains the most reliable method for confirmation. Additionally, an important limitation we encountered during the surgery was the slight irritation of the glossopharyngeal and vagus nerves, leading to symptoms such as coughing and hoarseness. This highlights the need for careful attention and consideration of these factors in future surgeries.

In summary, this case was rare, complex, and had excellent surgical results. It is important to consider the possibility of concurrent acoustic schwannoma and ependymoma when encountering tumors with different textures in the posterior fossa. The occurrence of these two tumors together is not well-understood, and there

are limited reported cases and unclear pathophysiological mechanisms. Further research focusing on the genetic and molecular mechanisms underlying the coexistence of these tumors is necessary to enhance our understanding and optimize patient outcomes in this rare clinical scenario.

Abbreviations
CPA Cerebellopontine angle

Supplementary Information

The online version contains supplementary material available at https://doi.org/10.1186/s12877-024-05664-0.

Supplementary Material 1: **Supplementary Fig. 1**: A The patient was in the left prone position, and the head was fixed by three nails. During the operation, the three nails and the operating table were rotated to achieve the union of the surgical approach. B We employed the right sigmoid sinus posterior approach combined with the posterior median approach. The white arrow shows the right transverse and sigmoid sinus junction after removal of the bone flap. C Right acoustic schwannoma. Under light microscopy, the tumor cells are arranged in a sparse and dense pattern. In the sparse areas, the cells form a network structure accompanied by mucinous degeneration, while in the dense areas, the tumor cells are arranged in a spindle-shaped pattern (as indicated by the arrows). D Ependymoma of posterior cranial fossa of fourth ventricle. Under light microscopy, the tumor cells are spindle-shaped or oval, arranged in a papillary pattern around fibrovascular cores. Some tumor cell nuclei are large and deeply stained. A large number of foam cells and areas of calcification are visible in certain regions.

Supplementary Material 2: **Supplementary Fig. 2**: Three months after the surgery, the patient underwent an MRI examination. The MRI images revealed a completely normal CP angle with no signs of abnormalities or obstructions in the fourth ventricle.

Supplementary Material 3: **Supplementary Fig. 3**: Ostoperative facial expressions.

Supplementary Material 4: **Supplementary Video**: The surgical procedure.

Acknowledgements
Not applicable.

Author contributions
Haofuzi Zhang was a major contributor in writing the manuscript. All authors read and approved the final manuscript.

Funding
Not applicable.

Data availability
No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate
The study was conducted in accordance with the Declaration of Helsinki, and approved by the Institutional Review Board (or Ethics Committee) of Xijing Hospital (protocol code: KY20193098, date of approval: 28 February 2019). Informed consent was obtained.

Consent for publication
Informed consent for publication was obtained from all subjects/ participants.

Competing interests
The authors declare no competing interests.

Received: 12 December 2023 / Accepted: 30 December 2024
Published online: 10 January 2025

References

1. Samii M, Gerganov V. Vestibular Schwannomas. Surgery of Cerebellopontine lesions. Berlin, Heidelberg: Springer Berlin Heidelberg; 2013. pp. 147–314.
2. Neupane S, Kashyap A, Paudel S, et al. A rare case of schwannomatosis with meningioma: a case report. Ann Med Surg (Lond). 2024;86(3):1724–8.
3. Tao XG, Hou ZG, Hao SY, Zhang JT, Liu BY. Two cases of Spinal Tanycytic Ependymoma Associated with neurofibromatosis type 2. Chin Med J (Engl). 2017;130(7):872–3.

Publisher's note