Vestibular schwannoma coexisting with dermoid cyst: A case report

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Abstract. Multiple primary intracranial tumors, or the presence of two or more primary intracranial tumors, are a rare clinical occurrence. The current study presents the case of a 28-year-old patient with concurrent left vestibular schwannoma, left cerebellar hemisphere dermoid cyst and craniovertebral junction malformation, specifically basilar invagination and Klippel-Feil syndrome. The patient exhibited symptoms of torticollis and recurrent headaches, with no apparent hearing loss. A far lateral approach was selected for surgical resection to address these complex conditions and achieve gross total resection in a single-stage surgery while preserving both facial and auditory nerve function. Successful gross total resection was achieved and the function of both nerves was effectively preserved. Of note, the coexistence of vestibular schwannoma and dermoid cyst in the same patient has not been documented in the existing literature. The present study provided a comprehensive account of the presentation and progression of this uncommon medical scenario. Furthermore, a surgical principle for the management of multiple primary intracranial tumors was proposed.

Introduction

Multiple primary intracranial tumors are characterized by the simultaneous presence of two or more primary intracranial tumors. These cases are exceptionally rare, with limited instances documented in the medical literature and the specific incidence remains undetermined (1). Of note, the coexistence of a vestibular schwannoma and a dermoid cyst is an uncommon occurrence, and no such case has hitherto been reported to our knowledge. It is now understood that intracranial dermoid cysts constitute approximately 0.3% of all intracranial tumors and typically originate from residual mesodermal and ectodermal tissue within the neural tube during embryonic development (2,3). By contrast, vestibular schwannomas, arising from Schwann cells of the vestibular branch of the vestibulocochlear nerve, represent a relatively common type of intracranial tumors, accounting for ~8% of all intracranial tumors (4). Surgical resection remains the primary curative approach for both of these tumor types. However, the surgical approach requires meticulous consideration when they coexist within a patient. A single-stage surgical approach should be carefully considered to achieve gross total resection. The current study presented the case of a patient afflicted by both a vestibular schwannoma and a dermoid cyst who underwent one-stage surgery, resulting in the successful achievement of gross total resection. This case report aims to enhance awareness regarding the occurrence and management of multiple intracranial tumors.

Case report

A 28-year-old male patient presented at Department of Neurosurgery, Beijing Tiantan Hospital (Beijing, China) with the chief complaint of 'recurrent headaches for the past six years, which had worsened over the last year' in November 2021. Approximately six years ago, the patient began experiencing persistent occipital headaches that would occasionally subside on their own. However, the severity and frequency of these headaches had intensified over the course of the past year. Of note, there were no records of intracranial tumors or neurological diseases among the patient's family members. Upon physical examination, the patient was determined to exhibit a head tilt to the right and torticollis. The patient was alert and oriented, and neurological examination revealed no focal deficits. Magnetic resonance imaging (Fig. 1A-E) revealed the presence of two lesions. One was a 63x63x44 mm cystic-solid lesion centered in the left cerebellar hemisphere, exhibiting well-demarcated borders and causing compression of the fourth ventricle with resultant supratentorial hydrocephalus. The second was a 14x19x17 mm solid, homogenously enhancing tumor located within the left cerebellopontine angle and extending into the internal auditory canal. Head CT revealed calcifications along the cyst wall and enlargement of the left internal auditory canal (Fig 1F). Audiometry findings (Fig. 2) indicated bilateral hearing

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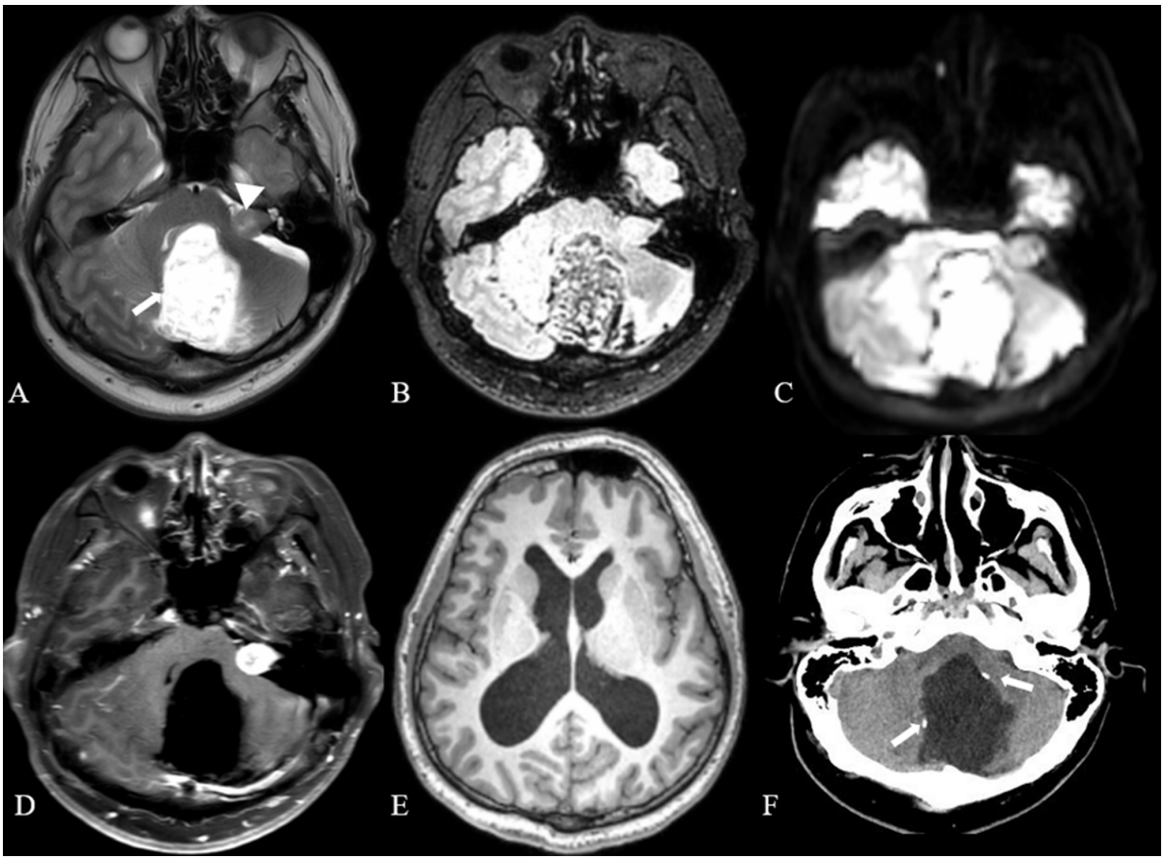

Figure 1. Preoperative images. (A) Axial T2-weighted MR image revealed a large, left cerebellar hemisphere cystic lesion (arrow), along with a solid lesion in the left CPA region (arrowhead). (B) In the axial fluid-attenuated inversion recovery MR image, the left cerebellar hemisphere cystic lesion is dark and heterogeneous. (C) On axial diffusion-weighted MR imaging, the left cerebellar hemisphere cystic lesion was displayed with a markedly high signal. (D) The axial T1-weighted contrast-enhanced MR image displayed the left cerebellar hemisphere cystic lesion without enhancement, but the solid lesion in the CPA region was homogeneously enhanced. (E) The fourth ventricle outflow was decompressed and axial T1-weighted MR imaging revealed supratentorial hydrocephalus. (F) Axial CT scan revealed the partial calcification around the cystic wall and enlargement of the left internal auditory canal compared to the right (arrows). CPA, cerebellopontine angle; MR, magnetic resonance.

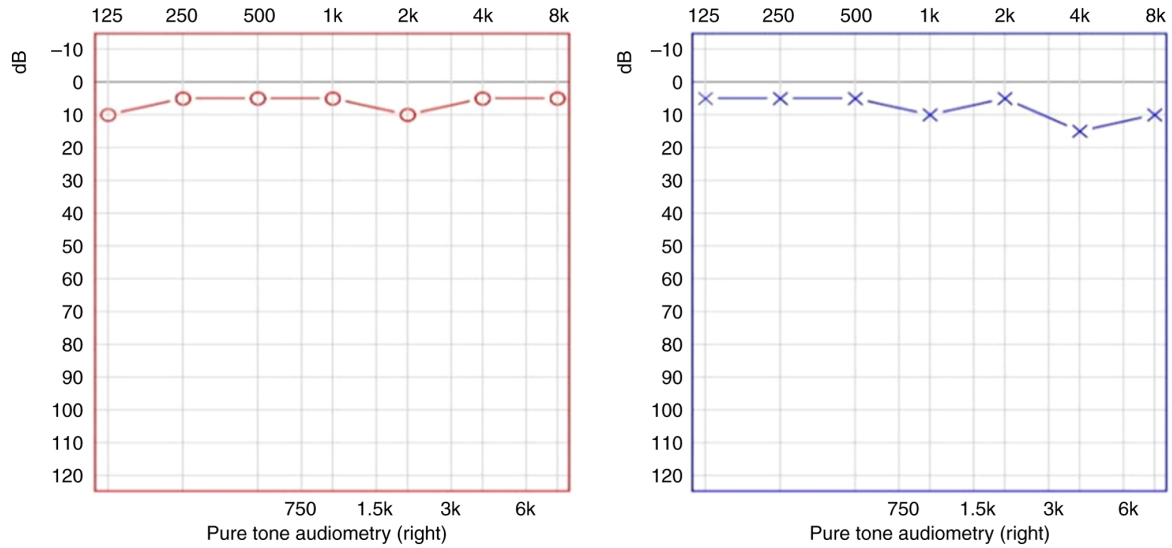

Figure 2. Preoperative audiometry. Right ear: PTA=37 dB; left ear: PTA=37 dB. PTA, pure tone audiometry.

consistent with Class B, as per the American Academy of Otolaryngology-Head and Neck Surgery (AAO-HNS) criteria. Preoperative laboratory values were within normal limits.

The chosen surgical approach was the left far lateral approach, as opposed to the retrosigmoid or posterior median approach. The upper boundary extended to the transverse

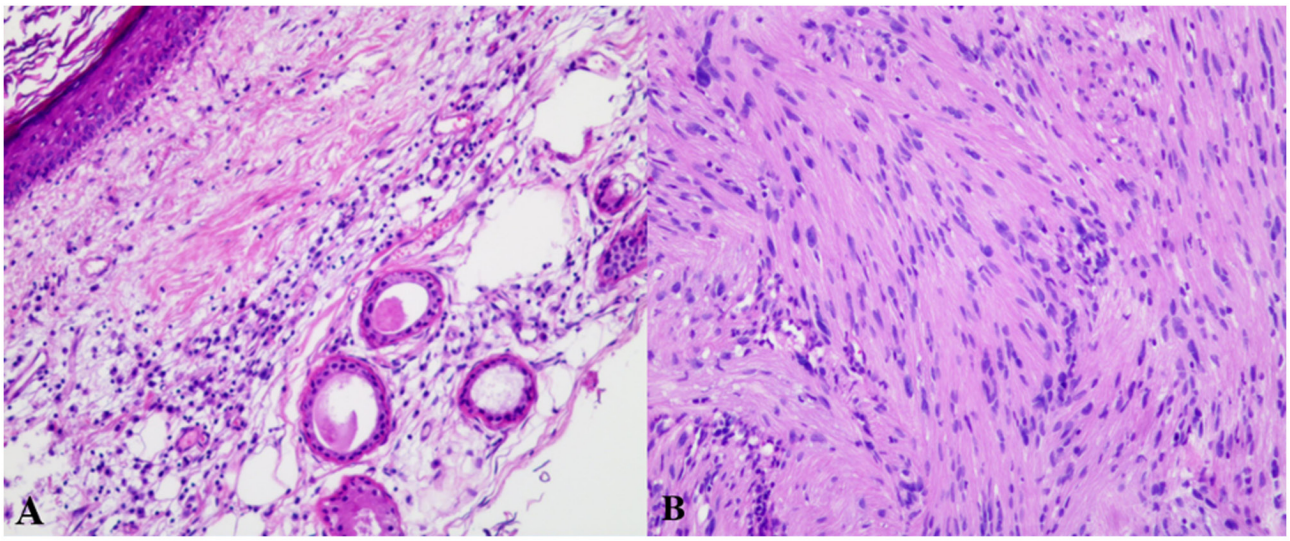

Figure 3. Histopathological examination of the resected tumor. (A) H&E-stained section of the cerebellar hemisphere tumor (magnification, x100). (B) H&E-stained section of the cerebellopontine angle tumor (magnification, x200).

sinus and the foramen magnum was opened inferiorly without grinding the occipital condyle. The cystic-solid tumor located in the left cerebellar hemisphere was found to have an irregular cystic wall. Beneath the tumor, the contents had ruptured through the cystic wall, containing grey-white material filled with hair and sebaceous gland-like substances. After decompressing the tumor, careful separation along the cystic wall ensured the integrity of the remaining cystic wall, followed by complete tumor resection. The cystic wall was found to be partially calcified. The other tumor was exposed in the left cerebellopontine angle (CPA) region by gently retracting the cerebellum medially. This tumor had a grey-yellow appearance, firm texture and rich blood supply. The arachnoid membrane overlying the tumor surface was dissected, followed by the opening of the tumor capsule in an avascular plane to achieve adequate intratumoral decompression. The posterior wall of the internal auditory canal (IAC) was then drilled stepwise to expose the IAC tumor, which was subsequently resected while preserving the anatomical integrity of the ipsilateral facial and cochlear nerves. The estimated intraoperative blood loss was 200 ml over the 7-h procedure.

Postoperative histopathological examination revealed that the cystic wall of the cerebellar hemisphere tumor (Fig. 3A) consisted of two layers: A cornified layer and a stratified squamous epithelium layer. Within the fibrous connective tissue inside the cyst, sebaceous glands and other skin appendages were visible, along with focal lymphocytic infiltration, supporting a diagnosis of dermoid cyst. The left CPA tumor (Fig. 3B) displayed spindle-shaped cells with elongated elliptical nuclei, closely and parallelly arranged in a palisade pattern, indicating a diagnosis of schwannoma (WHO I).

The patient demonstrated good postoperative recovery. Facial symmetry was maintained with normal muscle tone at rest. The motion of the upper forehead was basically normal and the patient was able to close the eyelids totally with force, with only mild asymmetry of the oral commissure (House-Brackmann grade II, as presented in Fig. 4A) (5). Audiometry conducted three months after surgery (Fig. 4B) revealed a left ear pure-tone average (PTA) of 52 dB and a speech discrimination score (SDS) of 100% (data not shown), while the right ear had a PTA of 30 dB and an SDS of 100%. Postoperative imaging (Fig. 4C-F) confirmed complete tumor resection with no residual lesions. Regular follow-up examinations at 3, 12 and 24 months post-surgery revealed resolution of the headache symptoms and stable bilateral hearing levels. However, the patient did report episodic tinnitus. Head MRI at one year post-surgery (Fig. 5) showed no evidence of residual or recurrent tumors.

Discussion

The present study reported on a rare case of multiple primary intracranial tumors, which may be classified into homologous and heterologous types based on their histological origin. The homologous type consists of multiple tumors with the same origin, such as multiple meningiomas and multiple gliomas, while the heterologous type comprises tumors with different histological origins (6). Heterologous primary tumors are infrequent and predominantly affect women aged 30 to 60 years. The most common combination includes meningioma and pituitary adenoma, with meningiomas often coexisting with gliomas, primary intracranial lymphoma, schwannoma and craniopharyngioma (7). Another intriguing phenomenon is 'collision tumors', where different pathological tumor types occur in adjacent locations (1). While sporadic vestibular schwannoma coexisting with other tumors has been reported in cases of the neurofibromatosis type 2 (8-11), there are no documented cases of concurrent intracranial dermoid cysts. Of note, dermoid cysts have a lower incidence rate and rarely coexist with other intracranial tumors. There has been one documented case of a dermoid cyst in the frontal lobe coexisting with craniopharyngioma (12).

For heterologous primary tumors, certain studies have suggested that the lesion or surrounding edema may stimulate the transformation of surrounding astrocytes or arachnoid granule cells into tumor cells (7,13). A case report documented a combination of dermoid cyst, intracranial aneurysm and glioblastoma (GBM), where chronic inflammatory stimulation

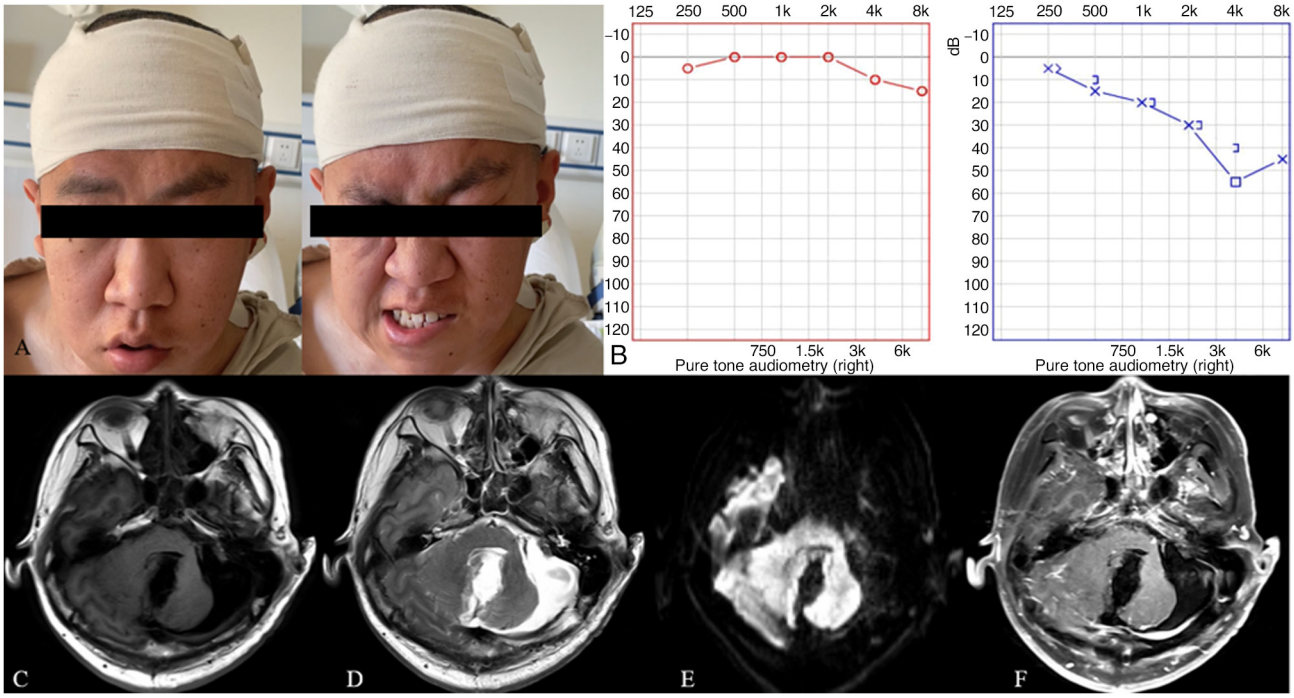

Figure 4. Postoperative condition. (A) Postoperative facial nerve function. The face is normally symmetrical at rest, the eyelids can be fully closed during moving (masked for anonymity) and the commissure is slightly asymmetrical (H-B II). (B) Postoperative hearing examination. Right ear: PTA=30 dB; left ear: PTA=52 dB. High-frequency hearing of air conduction in the left ear decreased after surgery, but there was still practical hearing. Postoperative magnetic resonance imaging, including (C) T1WI, (D) T2WI, (E) diffusion-WI and (F) contrast-enhanced T1WI indicated complete resection of the tumor, with no residual tumor. PTA, pure tone audiometry; WI, weighted imaging.

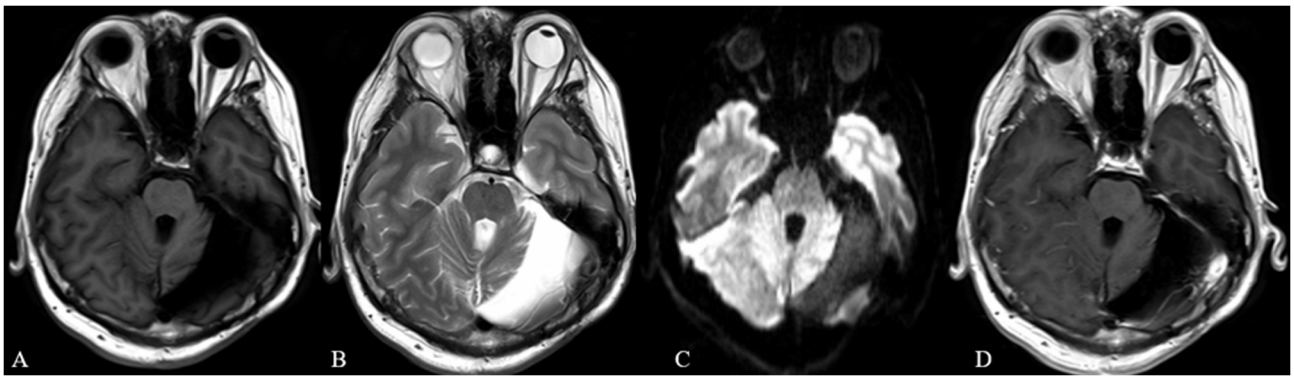

Figure 5. Head magnetic resonance images at 1 year after surgery. The images, including (A) T1WI, (B) T2WI, (C) diffusion-WI and (D) contrast-enhanced T1WI indicated no tumor residue or recurrence. WI, weighted imaging.

caused by the dermoid cyst and repetitive epilepsy may have contributed to the development of GBM (14,15). However, this theory did not explain the occurrence of another tumor in a distant site. Certain studies have proposed that both tumors may share the same genetic pathway, promoting the occurrence of both tumors, but at present, there is insufficient evidence to support this perspective (13). Dermoid cysts are widely thought to result from embryonic developmental abnormalities, whereas sporadic vestibular schwannoma appears unrelated to such developmental issues (12,16). Further investigation is warranted to understand the etiology of tumors in such cases.

Surgery for multiple intracranial tumors should be determined based on their location and clinical characteristics. If the tumors are close in proximity, a surgical approach covering both should be chosen for one-stage resection. In cases where the tumors are located far apart, there is debate regarding which tumor should be resected first. Certain clinicians suggest that the more malignant tumor should be removed initially before addressing the relatively benign tumor in the second stage (7). However, the approach pursued in the present study prioritizes the tumor with a more significant mass effect or a greater impact on function. Careful consideration was given to the selection of the surgical approach to achieve this objective.

In the case of the present study, the patient had a dermoid cyst in the left cerebellar hemisphere and a vestibular schwannoma in the left CPA region. The dermoid cyst had compressed the fourth ventricle, leading to hydrocephalus. Following our surgical principle, the dermoid cyst was resected as the primary

procedure. Despite the vestibular schwannoma appearing asymptomatic and the patient not experiencing hearing loss, it is important to note that hearing loss associated with vestibular schwannoma tends to deteriorate over time. It has been reported that the average rate of deterioration in PTA for patients with vestibular schwannoma is ~4.4 dB hearing degeneration per year. Furthermore, hearing loss is irreversible, and preserving postoperative hearing is the optimal outcome (17,18). Given the mass effect caused by the dermoid cyst, the vestibular schwannoma had contacted the brainstem, reaching Grade II according to the Koos classification criteria. Currently, there is no evidence to suggest that Grade II tumors are more suitable for stereotactic radiosurgery treatment, whereas surgery is applicable for tumors of all sizes (19). Based on our experience and the necessity of resecting the dermoid cyst, the present approach sought to achieve total tumor resection and attempt to preserve hearing through a single-stage procedure.

In the present case, the far-lateral approach was innovatively selected for tumor resection. The far-lateral approach offers extensive exposure for posterior fossa operations, providing access to the entire CPA, foramen magnum and upper cervical region. It allows for better visualization of the ventral or ventrolateral brainstem compared to the retrosigmoid approach (20). However, the far-lateral approach is more complex and time-consuming, with a risk of vascular injury, particularly when dealing with the occipital condyle (21). The far-lateral approach is rarely used for routine resection of CPA tumors, except when the tumor extends inferiorly into the foramen magnum region (22). Sanai and McDermott (21) had proposed a modified far-lateral approach for the resection of larger posterior fossa or CPA tumors. However, in this case, the vestibular schwannoma exhibited a minimal mass effect and the far-lateral approach did not provide sufficient surgical space to expose the lesion in the absence of a dermoid cyst. With the presence of a dermoid cyst, the far-lateral approach allows for exposure of the vestibular schwannoma by gently pulling the cerebellum following excision of the dermoid cyst. In addition, the far-lateral approach offers a more oblique view of the fundus of the IAC compared to the widely used retrosigmoid approach, necessitating less removal of the posterior wall of the IAC. In contrast to the conventional far-lateral approach, the primary focus in the present study was on exposing the left cerebellar hemisphere and left cerebellopontine angle region, thus preserving the occipital condyle. However, the bone window was extended superiorly to optimize cerebellar exposure. Resecting the dermoid cyst along the tumor boundary aimed to maintain the integrity of the cyst wall and reduce the risk of postoperative aseptic inflammatory response. Two key surgical techniques were employed to achieve postoperative hearing preservation in patients with vestibular schwannoma: Complete separation of the tumor capsule and exposure of the tumor within the IAC. The far-lateral approach facilitates safer and more effective tumor resection through improved exposure.

Numerous studies have suggested a potential complex relationship between intracranial dermoid cysts and craniocervical junction (CVJ) malformations (23,24). The need for surgical treatment of CVJ malformations during dermoid cyst surgery remains controversial. A case report documented a patient with a dermoid cyst accompanied by CVJ anomalies, where occipitocervical fusion was not performed during surgery. Instead, a neck collar was prescribed for one month, which yielded positive therapeutic effects (1). However, certain studies have advocated for occipitocervical fusion during the surgery for resecting dermoid or epidermoid cysts, with the preferred treatment involving surgical resection of the cysts along with posterior fixation (24). Given that the CVJ malformation was asymptomatic in this case, performing occipitocervical fusion during the resection of both intracranial tumors posed a high surgical difficulty and risk. Surgery may be postponed until symptoms arise.

In conclusion, the coexistence of a vestibular schwannoma and dermoid cyst is a rare condition within the realm of multiple primary intracranial tumors. The pathogenesis of their simultaneous occurrence remains elusive. The far-lateral approach was selected to achieve gross total resection and preserve neurological function in a one-stage surgery. The surgical principle for multiple primary intracranial tumors aims to achieve one-stage excision; if this is not feasible, the tumor with a more significant mass effect or greater functional impact should be addressed first. The surgical approach should also be adjusted to suit the specific characteristics of the lesion.

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Availability of data and materials

The data generated in the present study may be requested from the corresponding author.

Authors' contributions

WW provided medical records and designed the outline of this case report. RZ wrote the manuscript, prepared the figures and completed the follow-up. RF collected patient information and medical records, and wrote the 'case report' section of the manuscript. All authors were involved in the revision of the manuscript. All authors confirm the authenticity of all the raw data, and have read and approved the final version of the manuscript.

Ethics approval and consent to participate

The study was approved by the Ethics Committee of Beijing Tiantan Hospital (Beijing, China). Studies involving human participants followed ethical guidelines established by the institutional and/or national research committee, and complied with the 1964 Helsinki Declaration and its subsequent amendments.

Patient consent for publication

The patient provided written consent for the publication of his general information about his gender and age, case information including chief complaint and medical history, as well as clinical images.

Competing interests

The authors declare that they have no competing interests.

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