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Case Report

Diagnosis and management of bilateral vestibular schwannoma in the cerebellopontine angle: A rare case report ☆

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ABSTRACT

Vestibular Schwannoma (VS) is a benign nerve sheath tumors comprised of Schwann cells. This tumor is encapsulated, slow-growing, and originates from the internal auditory canal, extending into the cerebellopontine angle (CPA). The incidence in individuals aged 20-44 is 0.75 per 100,000 cases, with bilateral VS incidence of 0.8 per 50,000 cases. Tumors in CPA are the most common type in the posterior fossa and cause serious neurological symptoms or become life-threatening when tumors enlarge and compress the brainstem. The majority of tumors are VS (acoustic neuromas), accounting for 80%-90% of cases. Common clinical symptoms include hearing loss, tinnitus, and vertigo. Additionally, these tumors cause compression of the trigeminal and facial nerves. Advances in rapidly evolving imaging technology and surgical methods have improved diagnosis and management. A 24-year-old male complained of hearing impairment for the past 3 years alongside headaches, and dizziness leading to a feeling of imbalance, double, and blurry vision, as well as a sensation of facial thickness on the left side. Neurological examination showed cranial nerve abnormalities, including bilateral paresis of cranial nerves III, IV, VI, left cranial nerves V and VII, bilateral cranial nerve VIII, right cranial nerve XII, and cerebellar abnormalities such as intention tremor, dysmetria, dysdiadokokinesia, wide-based gait, and falling to the right during Romberg testing with both eyes open and closed. The patient underwent a contrast-enhanced MRI of the head, followed by a right CPA tumors excision through craniotomy. A detailed understanding of the medical history, physical examination, and radiological proved to be crucial in establishing an accurate diagnosis and appropriate management. This was considered essential to minimize a worse prognosis.

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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Introduction

Brain tumors are widely recognized to occur at various ages and among different populations. Tumors located in the cerebellopontine angle (CPA) are the most common type in the posterior fossa and cause serious neurological symptoms or even become life-threatening when tumors enlarge and compress the brainstem [1]. The majority of CPA tumors are vestibular schwannoma (VS) or acoustic neuromas, accounting for 80%-90% of cases [2]. VS are benign nerve sheath tumors composed of Schwann cells, encapsulated, and slow-growing, originating from the internal auditory canal and extending into CPA [3]. Other rare types include meningiomas, epidermoid cysts, arachnoid cysts, facial schwannoma, hemangiomas, choroid plexus papillomas, paragangliomas, and metastatic tumors [4]. The incidence of VS in individuals aged 20-44 is 0.75 per 100,000 cases, with bilateral VS incidence of 0.8 per 50,000 cases. According to a previous report, there is no significant difference in incidence between males and females [5]. Common clinical symptoms include hearing loss, tinnitus, and vertigo. When tumors size is significant and compresses the brainstem, it can lead to compression of the trigeminal and facial nerves [6].

Diagnosis is often challenging due to the various cell types and origins of tumors, hence, imaging examinations play a crucial role in diagnostic confirmation. Advances in rapidly evolving imaging technology and the development of microsurgical methods are expected to improve diagnosis and management of CPA tumors. This improvement is anticipated to reduce morbidity and mortality rates [4].

Case report

A 24-year-old male patient complained of hearing impairment for the past 3 years, starting in the right ear and worsening gradually. This was followed by inclusion of the left ear 1 year ago. The hearing impairment worsened to the point where hearing ability was completely lost. Around 6 months ago, the patient began experiencing intermittent headaches in the back of the head, with a pain rating of 2-3 on the Numeric Rating Scale (NRS). The headache subsided after self-medication, then 3 months ago, there was a complaint of dizziness, which led to a sense of imbalance while walking and frequent falls to the right side. Consequently, the patient adopted a wide-based gait to maintain balance, and the complaints were accompanied by blurred vision. Two months ago, double vision, and a perception of increased facial thickness on the left side compared to the right were reported, but there were no complaints of numbness or tingling in the limbs. The patient did not experience seizures, ringing in the ears, limb weakness, nausea, vomiting, weight loss, fever, persistent cough, as well as bowel or bladder disturbances. In early 2022, surgery was performed for a lump on the head, and it was diagnosed as Plexiform Schwannoma. The patient denied a history of hypertension, diabetes mellitus, stroke, tuberculosis, or other tumors.

During the examination, vital signs, and other general assessments were within normal limits. On neurological assessment, the Glasgow Coma Scale (GCS) score was 456, and there was papilledema in both eyes. Cranial nerve abnormalities included bilateral paresis of III, IV, and VI, left-sided hypoesthesia of V1, V2, and V3, left-sided lower motor neuron (LMN) type facial palsy, very severe sensory-neural hearing loss (>90 dB) in both ears, right-sided lingual palsy, and coordination disturbances in both arms. These included abnormalities in finger-pointing tests, finger-to-nose tests, intention tremors, and dysdiadokokinesia. Additionally, there was a wide-based gait, and during the Romberg test, with eyes open, the patient fell to the right side. Other neurological measurements were found to be within the normal limits.

Laboratory support examinations were within normal limits, while the contrast-enhanced MRI of the head indicated several results. These included bilateral contrast-enhancing masses in the right and left CPA, with the right-sided mass being larger. Both masses extended into the internal auditory meatus and compressed the cerebellum posteriorly, abutting against the pons, and causing narrowing of the fourth ventricle. These results suggested bilateral VS or Neurofibromatosis type 2 (Fig. 1).

During the treatment, a craniotomy was performed with suboccipital retrosigmoid approach to excise a subtotal tumors in the right CPA region, and an External Ventricular Drain was inserted. The excised tissue from the right side which is from 1 representative lesion was subsequently subjected to histopathology examination taken, and the result was showed schwannoma tumors. Postoperatively, the neurological examination tended to remain similar to the preoperative condition. A new neurological abnormality was observed namely right-sided LMN-type facial palsy. The patient underwent a postoperative contrast-enhanced head MRI, indicating bilateral vestibular schwanoma with left side appears more prominent (Fig. 2). Treatment was carried out for 2 weeks with Metamizole 3 times a day at 1 gram for pain and Dexamethasone 4 times a day at 5 mg IV, with dosage adjustments based on the clinical condition.

Discussion

VS or acoustic neuroma is the most common CPA tumors, originating from vestibular portion of cranial nerve VIII. These tumors manifest in 2 forms, sporadically or associated with neurofibromatosis (NF) type 2. Sporadic unilateral types are found in the fifth or sixth decade of life [2], while bilateral VS is a hallmark of neurofibromatosis (NF) type 2 [7]. These bilateral tumors are usually found in younger patients and are predisposed to the development of multiple tumors within the central nervous system [2].

Symptoms associated with CPA tumors vary widely depending on the size, location, and growth. These symptoms can be categorized into auditory symptoms (eg unilateral hearing loss, tinnitus) vestibular (eg vertigo) [4], cerebellar dysfunction (eg coordination disturbances, ataxia, and disequilibrium, nystagmus, tremors, and hypotonia) [1], cranial nerve-related (eg *facial hypesthesia,* facial weakness, dysphagia, and dysarthria) [4], and also experience vomiting, diplopia, altered mental status, and papilledema [8].

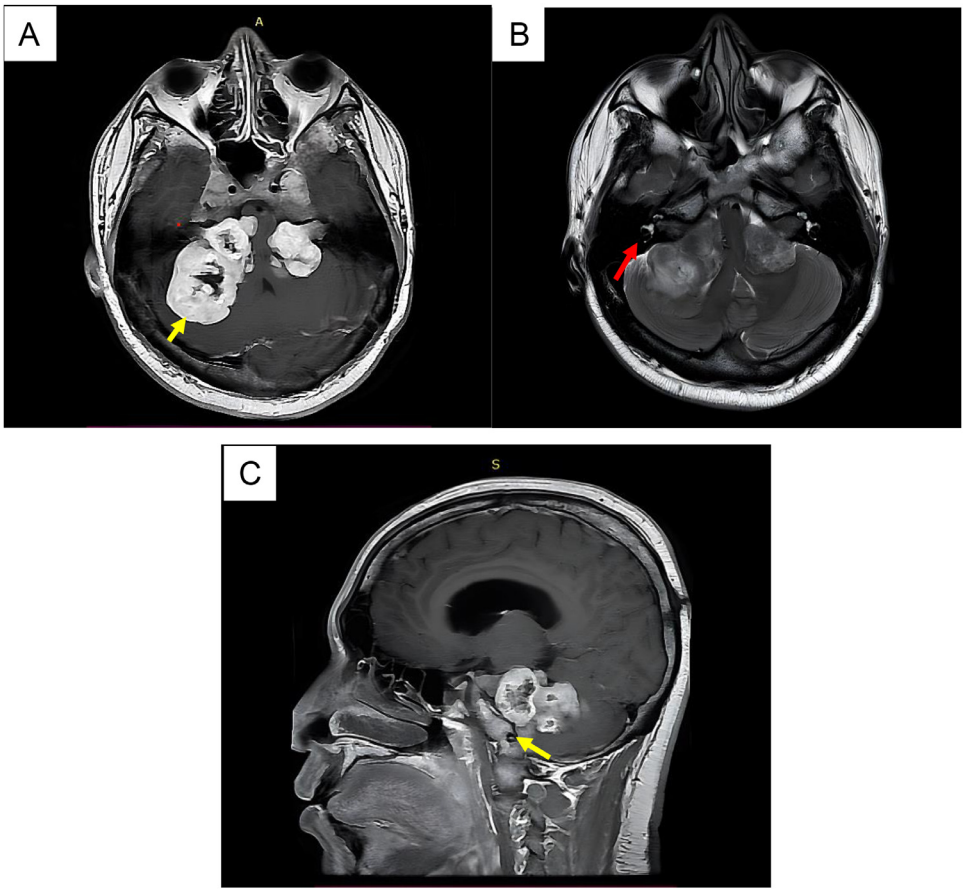

Fig. 1 – Axial T1-weighted MRI with contrast (A), Axial T2-weighted MRI (B), Sagittal T1-weighted MRI with contrast and (C) showing tumors in CPA extending into the internal auditory meatus (red arrow) and compressing the cerebellum against the pons (yellow arrow).

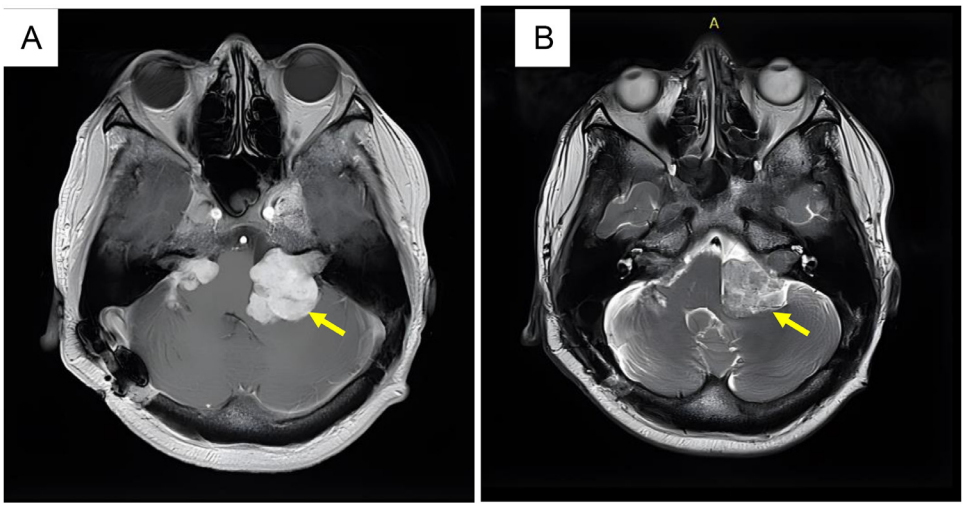

Fig. 2 – Axial T1-weighted MRI with contrast (A) Axial T2-weighted MRI (B) showing bilateral vestibular schwanomma with left side appears more prominent (yellow arrow).

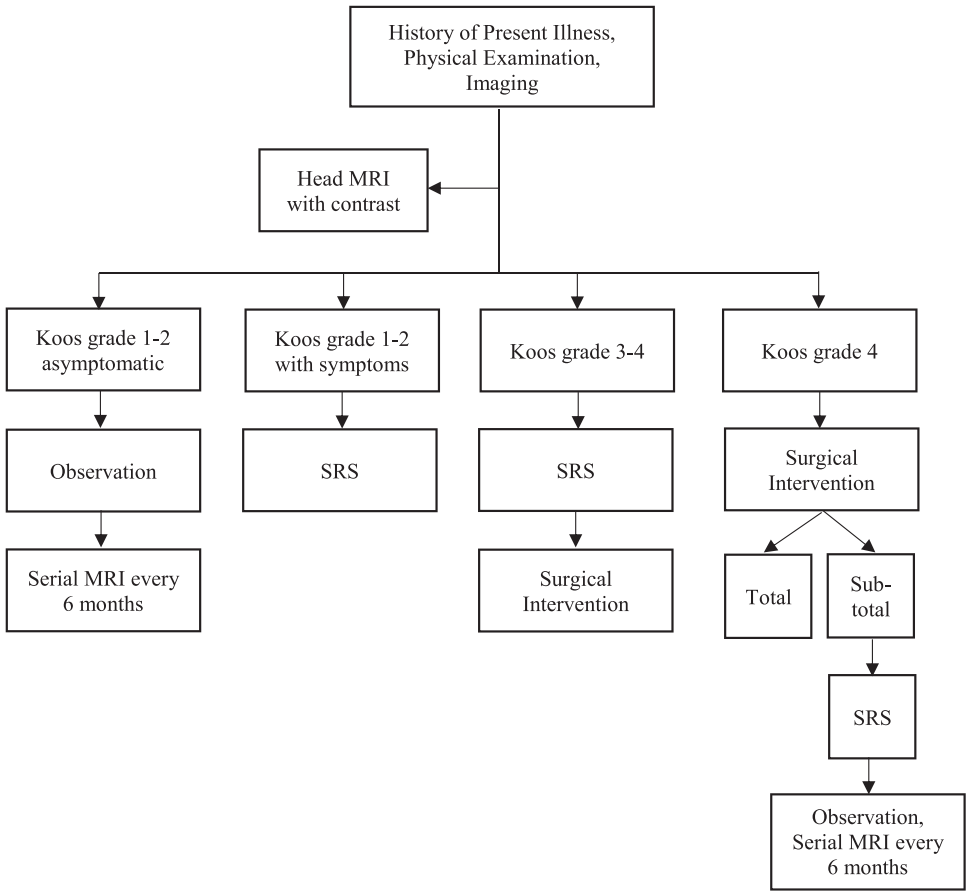

Fig. 3 – Algorithm for the management of VS.

VS appears as a solid nodular mass with intracanalicular components within the internal auditory canal. Larger lesions may extend into the cerebellopontine cistern, while smaller ones are often localized within the internal auditory canal or labyrinth. The mass typically appears isointense on T1-weighted imaging, with strong contrast enhancement following gadolinium administration. On T2-weighted imaging, the lesion is heterogeneously hyperintense, and larger lesions may show scattered cystic degenerative changes as well as hemorrhagic areas [7]. The situation of the parent nerve and enhancement pattern are predominantly peripheric.[9] T1-weighted imaging with gadolinium contrast remains the gold standard for diagnosing VS.[7] This needs to be distinguished from neurofibromatosis and meningioma. Meningiomas generally appear isointense on the Tl sequence, which is strong and homogeneous postcontrast with a characteristic dura around the lesion called “dural tail” [2]. In neurofibromatosis, the situation of the parent nerve and enhancement pattern are predominantly central [9].

The classification of tumors stage (Table 1) and the hearing function scale (Table 2) are crucial for assessing the appropriate management for patients. Tumors management is categorized into observation, radiotherapy/stereotactic radiosurgery (SRS), and microsurgery. For Koos stage I-II tumors without symptoms, observation therapy is prioritized [7], which has the advantage of avoiding complications resulting from interventional treatment. In slow-growing tumors, there is a high likelihood of long-term hearing loss. The average tumors growth rate is 1-1.2 mm per year. Serial MRI scans should be performed every 6 months [2] with observation therapy to

Table 1 – Koos grading system [7].

Grade	Tumor description
I	Small intracanalicular tumor
II	Small tumor with protrusion into the cerebellopontine angle; no contact with the brainstem
III	Tumor occupying the cerebellopontine cistern with no brainstem displacement
IV	Large tumor with brainstem and cranial nerve displacement

Table 2 – Gardner–Robertson scale for hearing function [7].

Grades	Pure tone audiogram (dB)	Speech Discrimination (%)
I	0-30	70-100
II	31-50	50-69
III	51-90	5-49
IV	91-max	1-4
V	Non testable	0

monitor tumors growth and hearing function. SRS is recommended for Koos stage I-II with hearing impairment to prevent further disturbances.

In instances of Koos stage III-IV, both SRS and surgery are considered viable options. Although the risk with SRS is lower than surgery, discussing the options with the patient is important. The primary goal of therapy in Koos stage IV is brainstem decompression, making surgery the only choice. Excision can be performed as a total or subtotal procedure, and after subtotal tumors resection, SRS may be needed, or observation with serial MRI scans every 6 months (Fig 3) [7]. Potential complications of surgical intervention include permanent hearing loss, facial paralysis, ipsilateral facial hypoesthesia, and cranial nerve deficits [2].

The VS prognosis depends on the size of tumors, severity of neurological symptoms, and surgery type [10]. When subtotal excision is performed for VS, the recurrence rate increases to 27.6% compared to total excision, which has a recurrence rate of 3.8% [7]. Nevertheless, tumors growth rate after subtotal excision is approximately 0.35 mm/year[11] which is lower than the 1 mm per year in patients who do not undergo surgery [2]. Facial nerve and hearing function after subtotal excision are significantly better compared to total excision. Subtotal excision aims to remove sufficient tumors volume to alleviate mass effect-related symptoms and preserve facial nerve and hearing function, thereby significantly impacting the quality of life [12].

In our case, diagnosis was established based on the history, physical examination, and diagnostic tests. The patient who was less than 30 years old, presented auditory, vestibular, cerebellar, cranial, neurological symptoms, and bilateral VS were detected on MRI, initially suggested a possible diagnosis of NF type 2. However, there were no other clinical symptoms suggestive of NF type 2, such as cataract, retinal hamartoma, cutaneous manifestations and also on MRI we found a solid mass mixed with a cystic component and a peripheral contrast enhancement pattern suggesting the possibility of a scwhanoma. However, a definitive diagnosis is determined by histology examination.

From the clinical symptoms in this case, it could be classified as Koos stage IV and Gardner Robertson grade IV on the hearing function scale, which indicated the need for surgery and subtotal resection becomes an option due to the tight adhesion of the tumor to the nerves making difficult to separate the tumor on the facial nerve. Postoperatively, the neurological examination tended to remain similar to the pre-operative condition due to subtotal resection. Right facial paralysis that occurred after the surgery may be caused by cerebral edema, so we decided to administer dexamethasone and then the condition improved after 2 weeks. However, we still plan to perform serial MRI every 6 months and observe clinical symptoms, considering high recurrence rate in subtotal excision.

A detailed understanding of the medical history, physical examination, and radiological results is essential for establishing an accurate diagnosis and appropriate patient management. This is crucial for minimizing a worse prognosis and educating the patient more effectively.

Patient consent

I have received consent from the patients, and their legal representatives for use their radiology images and their case to publication in journal.

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