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Case Report

Intracranial ancient schwannoma originating from vestibular nerve: A case report and review of the literature

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ABSTRACT

Background: Ancient schwannoma (AS) is a subtype of schwannoma with degenerative features, which often progresses slowly over a long period of time. Intracranial AS is a rare benign tumor and there are no detailed reports of AS originating from the vestibular nerve.

Case Description: Herein, we present the case of a patient with the right vestibular schwannoma with multiple meningiomas and review three previous cases of intracranial AS. Near-total resection was performed for vestibular schwannoma and the pathological findings were AS (World Health Organization Grade I). Five months postoperatively, gamma knife radiosurgery was performed for a recurrent lesion of the right vestibular schwannoma in the internal auditory meatus. Although AS is known to be a benign pathology, there are cases of rapid growth and early recurrence, as the one presented here. The high Ki-67 index (up to 5%) and the presence of cysts may be related to the rapid progression of intracranial AS.

Conclusion: Therefore, careful follow-up is necessary even if adequate removal is achieved. In addition to pathological studies, the genetic background of intracranial AS warrants future investigations. Further accumulation of cases is necessary to clarify the clinical features of intracranial AS.

Keywords: Cyst, Intracranial ancient schwannoma, Pathology, Vestibular schwannoma

INTRODUCTION

Ancient schwannoma (AS) is a pathological subtype of schwannoma, first reported by Ackerman and Taylor.[1] It is a benign schwannoma characterized by pathological nuclear atypia or degenerative changes such as hemorrhage with hemosiderin deposition, lymphocytic infiltration, and cyst formation, and paucity of mitotic figures.[12] Reports on the percentage of AS among all schwannomas vary widely, ranging from 3.2% to 78.9%,[1,3,4,8,20] and are limited to reports of extracranial AS. Extracranial AS is known to progress slowly, with an average elapsed interval between symptom onset and surgery of 8.3 years,[5,10] and the pathological findings of degenerative changes are thought to support this clinical feature. AS typically occurs extracranially, such as in the retroperitoneum; however, intracranial AS has rarely been reported.[15,16] To date, only three cases of intracranial AS have been reported, and all of them originated from the trigeminal nerve.[2,3,19] Here,

we describe a rare case of an ancient vestibular schwannoma with rapid progression and early postoperative recurrence.

CASE REPORT

A 53-year-old woman was referred to our hospital with a history of gradually worsening symptoms of headache, hearing loss, dizziness, and tinnitus for half a year. MRI revealed a right cerebellopontine angle (CPA) tumor suspected to be a vestibular schwannoma, a right sphenoid ridge extra-axial tumor suspected as meningioma, and a left temporal extra-axial tumor also suspected to be a meningioma [Figures 1a and 1b]. The right sphenoid ridge tumor was relatively larger and compressed the optic chiasm; hence, we removed it first to preserve visual acuity. The pathological diagnosis was meningothelial meningioma (World Health Organization [WHO] Grade I). Meanwhile, the right CPA tumor grew rapidly with the maximum diameter expanding from 23 mm to 35 mm in 6 months, and the brainstem compression increased. Magnetic resonance imaging (MRI) showed a marked increase in the size of the cyst [Figures 2a and 2b]. Within 6 months, facial nerve palsy and trigeminal neuralgia newly appeared, and the previously recognized right hearing loss also progressed.

We resected the right CPA tumor through a lateral suboccipital retrosigmoid approach. The tumor extended into the internal auditory meatus, the cochlear nerve was running on the caudal side of the tumor, and the facial nerve was located on its ventral side, as confirmed by electrophysiological monitoring. Intraoperative findings suggested that the tumor was a vestibular schwannoma. To avoid persistent facial nerve palsy, we left a tiny tumor on the facial nerve while performing near-total resection (resection rate: 99%; [Figure 2c]). Postoperative improvement of facial nerve palsy (House-Brackmann Grade II)[9] was observed. Five months after surgery, the right residual lesion in the internal auditory meatus had grown, so stereotactic radiosurgery using Gamma Knife Icon (Elekta AB, Stockholm, Sweden) was performed for the recurrent lesion [Figure 2d].

Intraoperative images of tumor resection are shown in [Figures 3a-b]. The tumor was soft and had a cystic component. Histopathologically, atypical Schwann cells with nuclear pleomorphism were observed; however, mitotic figures were rarely observed. Degenerative changes such as hyalinized blood vessels, hemorrhage with hemosiderin

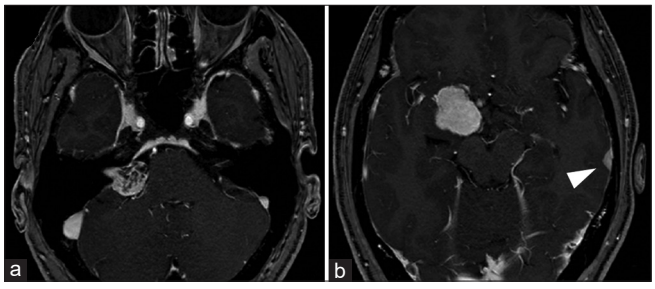

Figure 1: Preoperative imaging of the patient. MRI revealed a right CPA tumor suspected to be a vestibular schwannoma (a), a right sphenoid ridge extra-axial tumor suspected to be meningioma (b), and a left temporal extra-axial tumor also suspected to be a meningioma (b, arrow head). MRI: magnetic resonance imaging, CPA: cerebellopontine angle.

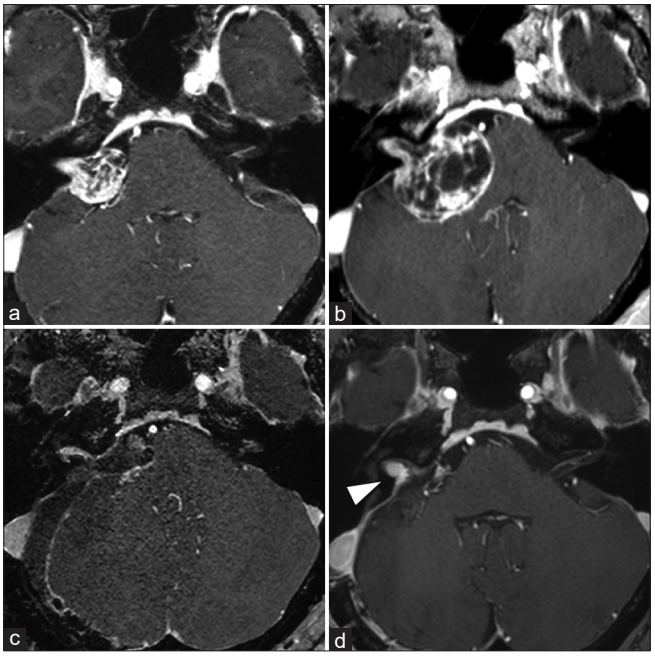

Figure 2: Imaging follow-up of the right CPA tumor. The right CPA tumor grew rapidly, with its maximum diameter expanding from 23 mm (a) to 35 mm (b) in 6 months, and the brainstem compression worsened. MRI also showed a marked increase in the size of the cyst. MRI: magnetic resonance imaging, CPA: cerebellopontine angle. To avoid persistent facial nerve palsy, we left a tiny tumor on the facial nerve while performing near total resection (resection rate: 99%) (c). Five months postoperatively, the right residual vestibular schwannoma in the internal auditory meatus had grown, so stereotactic radio surgery was performed for the recurrent lesion (d, arrow).

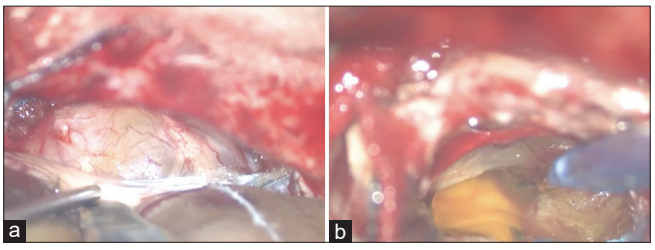

Figure 3: Intraoperative images of tumor resection. The tumor extended into the internal auditory meatus, the cochlear nerve was running on the caudal side of the tumor, and the facial nerve was located on its ventral side, which was confirmed by electrophysiological monitoring (a). The tumor was soft and had a cystic component (b).

deposition, lymphocytic infiltration, and cyst formation were observed. Immunostaining showed diffuse S100 positivity and the Ki-67 proliferation index was up to 5% [Figures 4a-d]. The tumor was diagnosed as an AS (WHO Grade I).

A detailed timeline on the treatment and the time course is presented in [Figure 5].

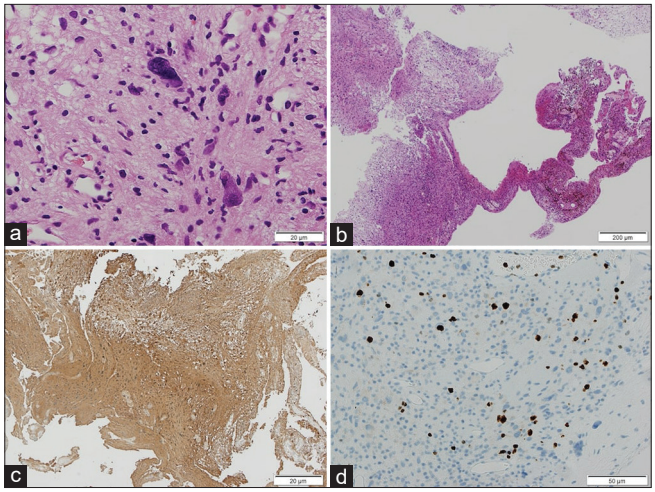

Figure 4: Histopathological findings of the tumor. Atypical Schwann cells with nuclear pleomorphism were observed, but mitotic figures were rarely seen (a). Degenerative changes such as hyalinized blood vessels, hemorrhage with hemosiderin deposition, lymphocytic infiltration, and cyst formation (b) were observed. Immunostaining showed diffuse S100 positivity (c) and the Ki-67 proliferation index was up to 5% (d). The diagnosis was ancient schwannoma (WHO Grade I). WHO: World Health Organization.

DISCUSSION

We report a case of a rapidly progressing ancient vestibular schwannoma with multiple meningiomas.[15] Ugokwe *et al.* reported the first case of intracranial AS and only three cases of intracranial AS have been previously reported, all of which were trigeminal in origin.[2,3,19] This case is the first report of AS originating from the vestibular nerve. One of the reasons for the low number of reports of AS is that they can only be diagnosed with pathological confirmation; therefore, they might be overlooked in cases in which conservative treatment or radiotherapy is used. It is also possible that cases of schwannomas were not diagnosed because little is known about their characteristics beyond pathological findings, or that cases of AS have been diagnosed but not reported.

We reviewed four cases of intracranial AS, including our case [Table 1]. The patients' ages varied from 23 to 70-years-old. Data regarding the size of the tumor were available only in two cases and they were relatively large, 35 mm and 70 mm. Cysts were present in three patients (75%). The mean interval time between symptoms and surgery was 8.3 months.

Extracranial AS is known to be characterized by a long duration from symptom onset to surgery, while intracranial AS may cause nerve compression at an early stage as the tumor progresses. It has been reported that the average time from symptoms to surgery in patients with extracranial AS is 8.3 years;[10] however, our literature review of four cases showed a much shorter average time of 8.3 months for intracranial AS. The mechanism of symptom onset is

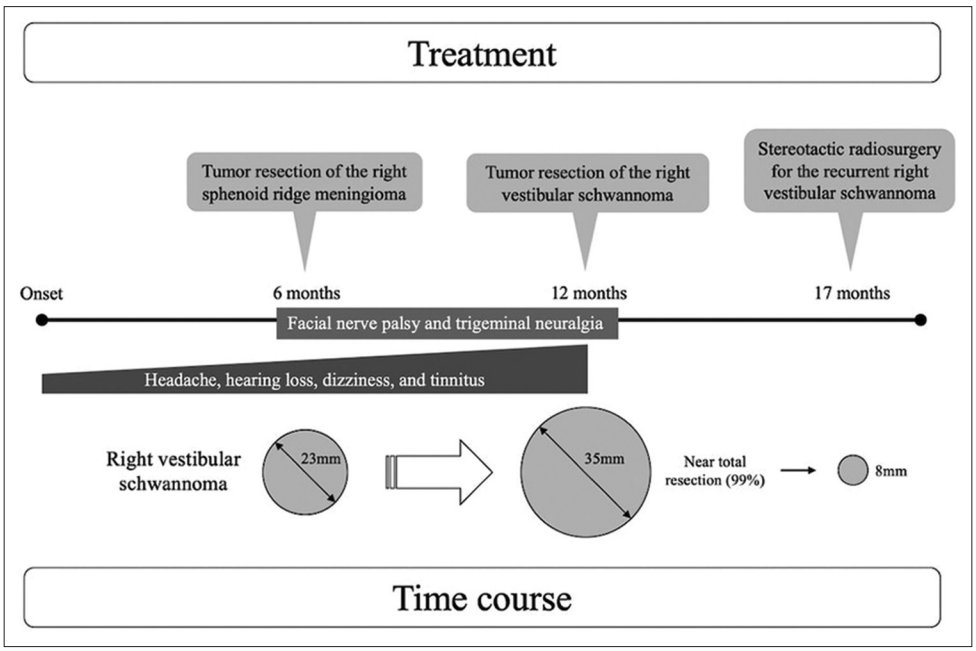

Figure 5: Detailed timeline on the treatment and the time course. The upper row describes the detailed course of the treatment and the lower row describes the detailed clinical time course.

Table 1: Four cases of intracranial ancient schwannoma.

Author[Ref]	Age	Sex	Location	Origin	Size (mm)	Cyst	Bleeding	GTR	Symptom to surgery (months)	Follow-up (month)
Ugokwe *et al.*[19]	23	M	Right	Trigeminal nerve	NA	Yes	No	Yes	10	6
Agrawal *et al.*[2]	70	M	Left	Trigeminal nerve	NA	No	No	Yes	3	NA
Al-Shudifat *et al.*[3]	35	F	Right	Trigeminal nerve	70	Yes	No	Yes	NA	24
Present case	53	F	Right	Vestibular nerve	35	Yes	No	No	12	6

F: Female, GTR: Gross total resection, M: Male, NA: Not applicable

anatomically different between intracranial and extracranial AS, and it may be necessary to consider them separately in terms of clinical pathogenesis. In this case, the time from the onset of symptoms to surgical resection for vestibular schwannoma was relatively short (1 year), and the tumor diameter rapidly increased from 23 to 35 mm within 6 months. At the same time, MRI also showed marked enlargement of the cyst, suggesting that it may have been related to the rapid progression of the vestibular schwannoma.

Cystic vestibular schwannoma is known to have larger tumor size, more rapid growth, and shorter duration of symptoms compared to solid vestibular schwannoma.[7,11,13,17] In this case, the relatively high Ki-67 index of 5% seems to have contributed to rapid progression and early recurrence; however, cyst formation might also contribute. Although cysts are seen as a degenerative finding in AS histology, the relationship between cystic vestibular schwannoma and AS is not known. However, cystic vestibular schwannoma is known to show cysts on microscopy[7] and is often characterized by hemorrhagic features such as hemosiderin deposition,[7,14] suggesting a relationship between cystic vestibular schwannoma and AS. Furthermore, in our literature review, three patients (75%) had cyst components, supporting a relationship between cysts and AS. However, the number of cases was small, and the related discussion was limited.

It is well known that the central cystic remodeling can be observed in large schwannomas. While differentiating AS from other subtypes with cystic remodeling, AS is characterized by nuclear atypia or degenerative changes on microscopy. Cysts are a degenerative finding but only one feature of the AS. On the other hand, MRI features are not well defined for AS. Hence, currently, diagnosing AS from MRI findings is difficult in the preoperative phase when pathology specimens have not been obtained, but the findings of cysts may increase the possibility of an AS diagnosis.

The pathological diagnosis is crucial; however, pathologists must be careful in their diagnosis because the nuclear atypia and degenerative features may lead AS to be misdiagnosed as malignant.[12] There may be an overlap between the pathological findings of cystic vestibular schwannoma and AS, and thorough follow-up is necessary, because early recurrence may occur with rapid growth, as in the case presented here.

Since the present patient had unilateral VS and multiple meningiomas, she was diagnosed with neurofibromatosis Type 2 (NF2) according to the latest NF2 diagnostic criteria (Manchester criteria).[6] Recent studies have shown that patients with unilateral vestibular schwannoma and multiple meningiomas are often diagnosed with mosaic NF2.[18] There have been no reports of AS accompanied by NF2. Furthermore, there are no reports on the molecular genetic background of AS. Genetic analyzes of AS might provide new insights into the genetic background of various pathological subtypes of schwannoma.

CONCLUSION

Intracranial AS is rare, so further accumulation and review of cases are necessary to clarify its clinical features. Although AS is known to be a benign pathology, there are cases of rapid growth and early recurrence, as the one presented here. The high Ki-67 index and the presence of cysts may be related to the rapid progression of intracranial AS. Therefore, careful follow-up is necessary even if adequate removal is achieved. In addition to pathological studies, the genetic background of intracranial AS warrants future investigations.

Statements and declarations

This manuscript is original and has not been published or presented elsewhere in part or in whole.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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