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Surgical Neurology International

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SNI: Neuropathology

Editor
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Case Report

Accessory nerve ancient schwannoma: A case report

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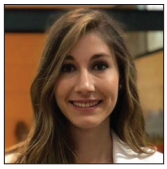

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Received : 29 July 2021
Accepted : 20 July 2022
Published : 05 August 2022

DOI
10.25259/SNI_747_2021

Quick Response Code:

ABSTRACT

Background: Lower cranial nerve schwannomas are rare and only 63 cases originating from the accessory nerve have been documented.

Case Description: We report a 61-year-old man who presented with a 3-month history of dysmetria, ataxic gait, and frequent falls. Magnetic resonance imaging revealed a giant rim-enhancing cystic lesion at the right cerebellomedullary cistern, which markedly displaced the brainstem and caused a critical compression on surrounding structures and mild hydrocephalus. Even though the nature of this lesion was not clear, it received a radiological diagnosis of meningioma as first option. Surgery was performed through an extended far lateral retrosigmoid approach with C1 hemilaminectomy, with intraoperative neurophysiological monitoring. A near-total resection was achieved due to the adhesion of the lesion to the brainstem and to the cranial nerves VII, VIII, IX, X, XI, and XII. Intraoperatively, the tumor was found to arise from the accessory nerve. The histopathological analysis concluded with a final diagnosis of ancient schwannoma, a rare histological subtype characterized by degenerative changes, typical from long-standing tumors.

Conclusion: Very few cases of intracranial ancient schwannomas have been described. To the best of our knowledge, this is the first report of this extremely rare histological variant arising from the intracisternal component of the XI nerve. The rarity of this disease at this location may lead to preoperative misdiagnosis.

Keywords: Accessory nerve, Ancient schwannoma, Case report, Schwannoma, Surgical treatment

INTRODUCTION

Schwannomas are slow-growing and benign tumors that arise from Schwann cells, which cover the axons from the peripheral nervous system. They are the most common tumor of the nerves but represent only 8% of primary intracranial tumors, and 90% of those originate from the vestibular nerve. Only 2% of intracranial schwannomas arise from the lower cranial nerves (IX, X, and XI).[2] To date, 63 cases of accessory nerve schwannomas have been reported in the literature.[10]

Ancient schwannomas are an uncommon type of schwannoma with degenerative changes that represent a longstanding growth. To the best of our knowledge, only one case of this ancient variant in the spinal accessory nerve has been documented.[4] Herein, we present the second case of this rare disease, the first affecting the intracisternal component of the XI nerve.

CASE DESCRIPTION

A 61-year-old man was referred to our neurosurgical department due to radiologic findings of two intracranial lesions. He presented with a 3-month history of gait imbalance, dysmetria, and frequent falls. During this time, he reported three isolated episodes of vomiting, but no headaches. Personal history included diagnosis of hypertension, dyslipidemia, and Type 2 diabetes mellitus. He had also been a heavy smoker for 40 years. Physical examination demonstrated a bidirectional persistent horizontal gaze-evoked nystagmus, ataxic gait, dysmetria, and positive Hoffmann and Babinski signs.

Magnetic resonance imaging (MRI) showed the two lesions previously seen. In the first mass, a radiologic diagnosis of suprasellar meningioma was made. The second was an extra-axial rim-enhancing cystic lesion of 35 × 38 × 45 mm. It was located on the inferior region of the right cerebellopontine angle, extending to the cerebellomedullary cistern, and causing marked compression on adjacent structures and a mild secondary hydrocephalus. The differential diagnosis at this time included cystic meningioma as first option or atypical schwannoma [Figure 1a].

The patient underwent surgical resection of the cisternal cystic lesion, with concomitant intraoperative neuromonitoring (IONM). With the patient in sitting position, an extended far lateral retrosigmoid craniotomy and cervical one vertebral right hemilaminectomy were performed. After opening the duramater, a heterogeneous highly vascularized cystic lesion was visualized, strongly fused to the brainstem, as well as to the VII, VIII, IX, X, XI, and XII right cranial nerves. The tumor was seen to arise from the right accessory nerve, confirmed with IONM. A near-total resection was performed to preserve these structures [Figure 1b].

On histological analysis, the pathological sample was described as a fusocellular proliferation, with abundant blood vessels, some dilated, and others with hyalinized walls, which were mixed with spindle cell fascicles with nuclei that presented size variability, some hyperchromatic. Proliferative index was 5% and there was no mitosis. Some areas of intravascular thrombosis and hemosiderin were exhibited. On immunohistochemistry, cells were intensely and diffusely positive for S100. Final anatomopathological diagnosis was ancient schwannoma [Figure 2].

This revealed that the mass did not correspond to a meningioma, as initially thought, but to an accessory nerve ancient schwannoma.

Postoperatively, the patient presented respiratory distress shortly after extubation, caused by abundant respiratory secretions and subsequent aspiration pneumonia. This together with later cardiac arrhythmias prolonged his stay at the intensive care unit. Swallowing difficulty was also notable, with the need of a nasogastric tube for feeding during a short time. At discharge 3 months after surgery, the patient had achieved autonomous oral feeding and deambulation.

The management of the remaining small capsular fragment consisted initially on follow-up. Subsequent MRIs showed

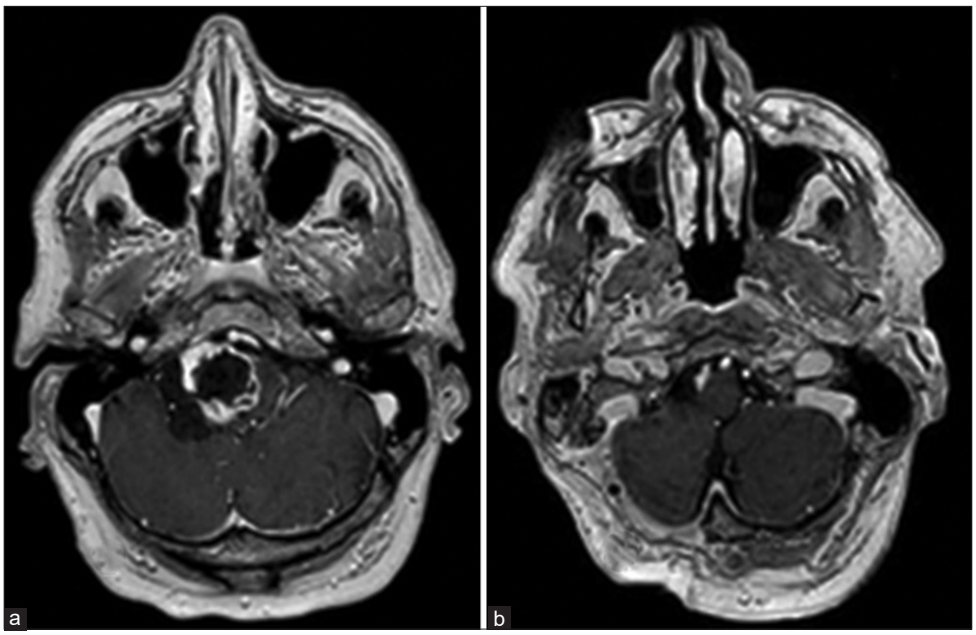

Figure 1: Radiological examinations: (a) preoperative axial T1-weighted contrast-enhanced MRI demonstrating a rim-enhancing lesion. (b) Postoperative axial T1-weighted contrast-enhanced MRI showing the thin layer of tumor left at surgery.

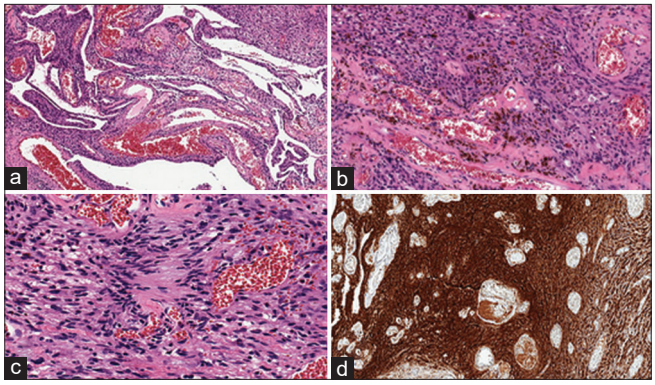

Figure 2: Histological examination: (a) (H&E, ×40) Ancient schwannoma showing cystic change. (b) (H&E, ×100) Hyalinized vessels surrounded by spindle cells and focal hemosiderin pigment. (c) (H&E, ×400) Presence of Verocay bodies. (d) (S100, ×40) Diffuse S100 positivity.

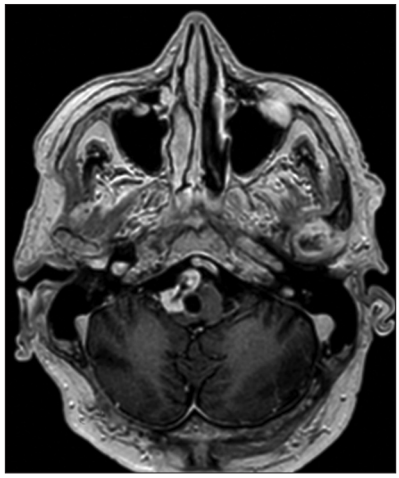

Figure 3: Thirteen-month follow-up axial T1-weighted contrast-enhanced MRI showing regrowth of the tumor.

regrowth of the residual lesion 13 months after surgery [Figure 3], especially of its cystic component. Due to the big cystic part, the decision was to reoperate with a debulking aim before radiotherapy. In the postoperative care unit, the patient suffered cardiac arrest of respiratory origin, which resulted in death after cardiopulmonary resuscitation failure.

DISCUSSION

Schwannomas are tumors that arise from the nerve sheath. Intracranially, most schwannomas originate from the vestibular nerve, being the most common lesion of the cerebellopontine angle, followed by meningiomas. Most of the times, a CT scan or an MRI is enough to make a reliable diagnosis of either lesion, but since only 2% of intracranial schwannomas arise from the lower cranial nerves (IX, X, and XI), this may be difficult, especially when their typical appearance is not exhibited.[2] Accessory nerve schwannoma is the least common of those arising from the lower cranial nerves, and only 63 cases have been reported in the literature. Of those, only 48 were located intracranially.[9,10]

Location of schwannomas is key for imaging diagnosis, as they follow the course of the nerves, may smoothly enlarge their foramina, and may cause fatty amyotrophy of innervated muscles.[3] Julow classified accessory nerve schwannomas attending to their location in two types: an intrajugular type located at the jugular foramen and an intracisternal type, located at the cisterna magna.[6] Later, an extracranial type has been added. According to this classification, our patient would have an intracisternal schwannoma. The most frequent preoperative misdiagnosis among lower cranial nerve schwannomas located mainly intracisternally is acoustic neuromas.[7] This was not the case in our patient, as the lesion was too inferior to be considered one.

At neuroimaging, schwannomas are usually round or oval masses and may display heterogeneous features including possible cystic or hemorrhagic components. Meningiomas are usually hemispheric, semilunar masses that do not follow the nerve course and are attached through a broad base to the meninges.[3] Our patient had two lesions. The suprasellar mass was clearly a meningioma, with its usual characteristics. On the contrary, his cisternal lesion was very ambiguous, not showing the typical characteristics nor location. At imaging, it was not clear that the mass followed the nerve course and the jugular foramen was preserved.

The radiologic findings, as well as the low rate of low cranial nerve schwannomas, and the fact that our patient had a suprasellar meningioma, led radiologists to make a preoperative diagnosis of meningioma. Finally, the histological analyses diagnosed an ancient schwannoma.

Schwannomas usually display a benign behavior, growing for a long time before been detected. This slow growth is even more remarkable in the case of schwannomas with ancient change, such as the one we discuss. Ancient schwannomas were first described by Ackerman and Taylor in 1951. They are described as having fibrous nodules, which are thought to occur due to diffuse overgrowth and vascularization that diminish with time, the vessels thicken, and cellularity of the connective tissue decreases until hyalinized areas appear. These tumors display no necrosis, but may show areas of fatty degeneration. Other features usually observed are areas of calcification, cystic formation, and hemorrhage. These characteristics suggest a longstanding growth.[1] Intracranial location of ancient schwannomas is extremely rare, being this the 8th case reported up to this date, and the first affecting the accessory nerve [Table 1].

Due to their slow growth and location, patients with intracisternal accessory nerve schwannomas do not usually

Table 1: Literature review of intracranial ancient schwannomas.

Authors and year	Age (years), sex	Origin	Maximum size (mm)	Cystic component	Differential diagnosis based on imaging	Resection	Ki-67 index	Outcome
Ugokwe *et al*., 2005	23, M	Trigeminal nerve	NA	Yes	Exophytic cerebellar astrocytoma, ependymoma, atypical schwannoma, abscess	Gross-total resection	NA	Good clinically at 6-month follow-up
Agrawal *et al*., 2010	70, M	Trigeminal nerve	NA	No	High-grade glioma, metastases	Near-total decompression	NA	Good
Micovic *et al*., 2017	73, F	Olfactory nerve	25	No	Aneurysm, meningioma	Total resection	NA	
Junaid *et al*., 2018	18, F	Optic nerve	85	No	NA	Total resection	NA	No recurrence at 2-year follow-up
Al-Shudifat *et al*., 2020	35, F	Trigeminal nerve	70	Yes	Chordoma, chondrosarcoma	Total resection	NA	No recurrence at 2-year follow-up
Takeuchi *et al*., 2022	61, M	Cerebellar hemisphere	65	Yes	Hemangioblastoma, low-grade glioma, metastatic tumor	Total resection	<1%	No recurrence at 5-year follow-up
Tsuchiya *et al*., 2022	53, F	Vestibular nerve	35	Yes	Vestibular schwannoma	Near-total resection	5%	Recurrence after 5 months, treated with stereotactic radiosurgery
Present	61, M	Accessory nerve	45	Yes	Meningioma, schwannoma	Near-total resection	5%	Recurrence after 13 months. Death after 2nd surgery.

F: Female, M: Male, NA: Not applicable

manifest clinical symptoms until the tumor reaches a great size. When present, symptoms usually consist on cerebellar signs and/or myelopathy, caused by the direct compression of the cerebellum, brain stem, and/or spinal cord. On the contrary, intrajugular tumors commonly associate cranial nerve deficits.[5] This is consistent with our patient's clinical findings, with a tumor larger than 4 cm, and a presentation that included dysmetria, ataxic gait, and positive Hoffman and Babinski signs.

Treatment of schwannomas consists on gross-total resection of the mass, with posterior follow-up, to assure no regrowth has occurred.[8] In our patient, a near-total resection was performed, to avoid neurologic complications, due to the adherence of the mass to the surrounding structures. Thus, the management of the remaining small capsular fragment was going to consist on follow-up with periodic clinical examinations and MRIs, until rapid growth of the lesion led to a new surgery.

Kaye *et al*. reported swallowing difficulty and salivary retention as the main postoperative complications in lower cranial nerve schwannoma surgery.[7] Our patient presented as well swallowing difficulty and salivary retention, which caused difficulty for extubation and further complicated his postoperative courses.

Although total resection of schwannomas is considered curative, this may not be achieved in all patients, such as in our case, due to the attachment of the lesion to surrounding structures. Since very few intracranial ancient schwannomas have been reported, no assertions about this subtype can be made. From the reported cases, we can see that the two tumors that showed rapid regrowth after surgery requiring further treatment presented a cystic component, a higher proliferative index, and a nontotal resection. This could suggest that ancient schwannomas, where the cystic component constitutes one of the typical degenerative changes, may require closer surveillance, especially when a higher Ki-67 index is present, and a nontotal resection is performed.

CONCLUSION

In this report, we describe the case of an accessory nerve ancient schwannoma, an extremely uncommon histological

variant, of which only one case affecting the XI nerve has been published, being ours the first arising from the intracisternal component of this nerve. The rarity of schwannomas of the XI nerve makes these lesions hard to diagnose preoperatively on MR imaging, which is especially true for lesions that do not clearly follow the course of the nerve within the jugular foramen, as one would expect from schwannomas. In our case, surgery was performed with a preoperative radiologic diagnosis of meningioma, which was later demonstrated to be an ancient schwannoma. Preoperative radiologic examinations must be carefully studied in advance and the surgeon must take into consideration any location or imaging characteristics that cause uncertainty about the diagnosis, to be prepared for possible new scenarios.

Declaration of patient consent

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

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Ackerman LV, Taylor FH. Neurogenous tumors within the thorax. A clinicopathological evaluation of forty-eight cases. Cancer 1951;4:669-91.
2. Ammirati M, Lamki T, editors. Overview of skull base tumors. In: Youmans and Winn Neurological Surgery. 7th ed., Vol. 2. Amsterdam, Netherlands: Elsevier; 2017. p. 1222-42.
3. Bonneville F, Sarrazin JL, Marsot-Dupuch K, Iffenecker C, Cordoliani YS, Doyon D, *et al*. Unusual lesions of the cerebellopontine angle: A segmental approach. Radiographics 2001;21:419-38.
4. George NA, Balagopal PG, Somanath T, Sebastian P. Multiple ancient schwannoma of the accessory nerve: a case report. Gulf J Oncolog 2012;11:66-9.
5. Jin SW, Park KJ, Park DH, Kang SH. Intracisternal cranial root accessory nerve schwannoma associated with recurrent laryngeal neuropathy. J Korean Neurosurg Soc 2014;56:152-6.
6. Julow J. Neurinoma of spinal accessory nerve. Report of two cases. Acta Neurochir 1983;69:219-24.
7. Kaye AH, Hahn, JF, Kinney SE, Hardy RW, Bay JW. Jugular foramen schwannomas. J Neurosurg 1984;60:1045-53.
8. Samii M, Babu RP, Tatagiba M, Sepehrnia A. Surgical treatment of jugular foramen schwannomas. J Neurosurg 1995;82:924-32.
9. Sarma S, Sekhar LN, Schessel DA. Nonvestibular schwannomas of the brain: A 7-year experience. Neurosurgery 2002;50:437-48; discussion 438-9.
10. Yan P, Wang P. Accessory nerve schwannoma: A new case report and systematic review. Interdiscip Neurosurg 2020;19:100571.

How to cite this article: Otero-Fernández P, Ruiz-Escribano-Menchén L, Herrera-Montoro V, Morcillo-Carratalá R, Calvo-García M, Llumiguano-Zaruma C. Accessory nerve ancient schwannoma: A case report. Surg Neurol Int 2022;13:338.