Schwannoma of the Fourth Ventricle: The Eighth Case Report

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Key words
- Brain tumor
- Fourth ventricle tumor
- Intra-axial schwannoma

Abbreviations and Acronyms
MRI: Magnetic resonance imaging
VP: ventriculoperitoneal

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Citation: World Neurosurg. (2015) 84, 5:1493.e9-1493.e13. http://dx.doi.org/10.1016/j.wneu.2015.04.036

Journal homepage: www.WORLDNEUROSURGERY.org

Available online: www.sciencedirect.com

1878-8750/$ - see front matter

OBJECTIVE: **We report an uncommon case of a surgical resection of a fourth ventricle tumor in an adult that proved to be a schwannoma.**

METHODS: **A 53-year-old man presented with a 1.5-year history of gait unsteadiness and vertigo and a few-week history of headache, emesis, and neurogenic dysphagia. A brain magnetic resonance imaging revealed a large, heterogeneously contrast enhancing mass located within the fourth ventricle, compressing the brainstem and causing supratentorial ventricle enlargement. A suboccipital craniotomy and a telovelar approach were performed to resect the tumor. The ventricular system was repermeabilized at the end of the operation.**

RESULTS: **A postoperative magnetic resonance imaging confirmed complete tumor removal. There was an initial worsening of the preoperative deficits, which progressively improved. The tumor was classified as a fourth ventricle schwannoma. There has been no evidence of tumor recurrence during the 6 years of follow-up. At present, the patient is ambulatory and reports an intermittent diplopia on conjugated gaze.**

CONCLUSION: **This case report intends to reveal the eighth case of a fourth ventricle schwannoma since 1957. Schwannomas of the fourth ventricle are infrequent but should be accounted in the differential diagnosis of space-occupying lesions in this location. Gross total resection might be the definite treatment of these tumors if deemed possible.**

INTRODUCTION

Schwannomas arise from the myelin sheath of peripheral nerves and account for 8% of all intracranial tumors. Although most commonly encountered in extra-axial locations, intra-axial schwannomas have been reported, including intraventricular lesions (4, 5, 7, 8, 13, 20, 25, 27). Tumors arising specifically in the fourth ventricle are extremely rare and only 7 case reports have been described to date (14, 16, 18, 22, 29). Because Schwann cells do not cover the central nervous system, theories trying to explain the occurrence of intra-axial schwannomas are speculative (2, 4, 6-8, 12, 13, 16, 17, 19, 20, 22, 28, 29). The aim of this report is to present the eighth case of a fourth ventricle schwannoma, which differs from the previously reported ones for its dimension and enriched neurological examination. A literature review focusing on this particular topography for schwannoma occurrence and its particular etiopathogenesis is also undertaken.

CASE REPORT

A 53-year-old man presented with progressive gait unsteadiness during the past 18 months and chronic vertigo. Few weeks before admission, he reported severe headaches, vomiting, and increasing dysphagia. His past medical history was unremarkable and there was no family history of neurofibromatosis. No café au lait spots were found on skin examination. The neurological examination showed a saccadic horizontal nystagmus, assymetrical palate elevation, broad-based gait, and truncal ataxia. The remaining cranial nerves were normal and there were no signs of motor or sensitive pathways dysfunction. A brain magnetic resonance imaging (MRI) revealed a large, contrast-enhancing, irregularly spherical, fourth ventricular tumor ($4 \times 4 \times 3.9$ cm) suggestive of choroid plexus papilloma or ependymoma. There was evidence of brainstem compression and the ventricles were enlarged despite no signs of active hydrocephalus (Figure 1). Surgical treatment was recommended.

Operation and Postoperative Course

The patient was positioned prone with the head flexed. A midline incision was done extending from the inion to C2. A suboccipital craniotomy was performed down to the foramen magnum and the posterior arch of C1 was removed. After opening the dura mater, the cerebellar tonsils were gently retracted. Opening of the tela choroidea and the inferior portion of the inferior medullary velum exposed a yellowish mass occupying most of the fourth ventricle. After internal debulking of the tumor, it was meticulously dissected from the floor of the fourth ventricle, where it was adherent. A total resection was achieved.

Transient new neurological deficits were induced and included VI, IX and X nerve palsies and ataxia worsening. On postoperative day 9, a percutaneous endoscopic

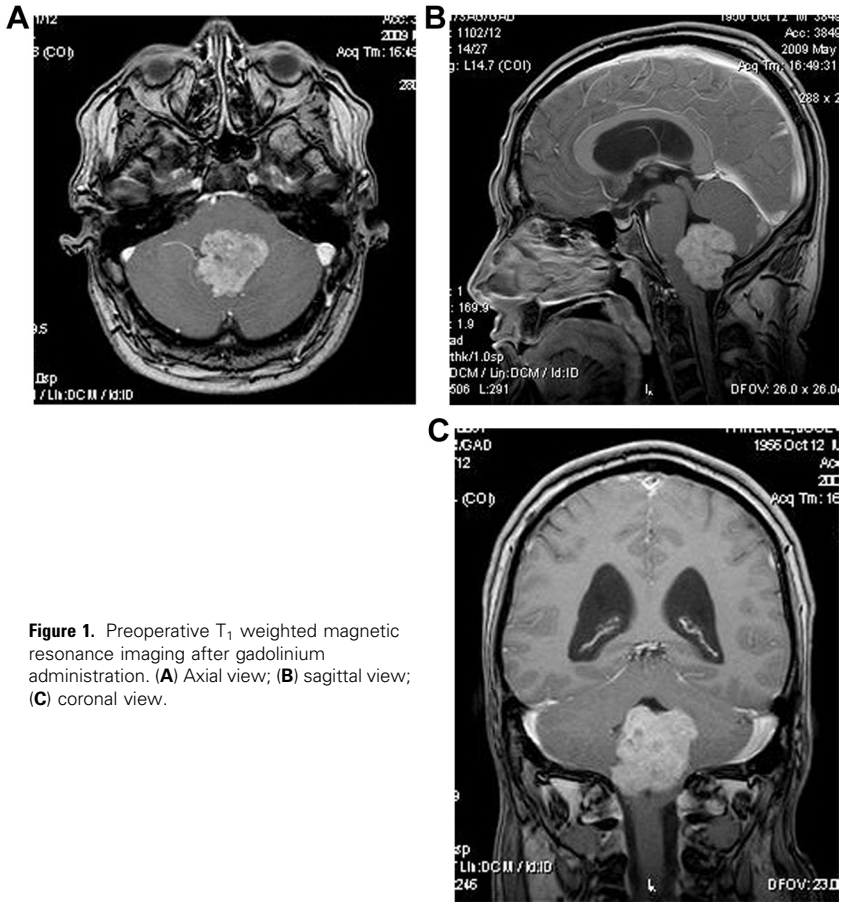

Figure 1. Preoperative T_1 weighted magnetic resonance imaging after gadolinium administration. (**A**) Axial view; (**B**) sagittal view; (**C**) coronal view.

gastrostomy tube was inserted, mechanical ventilation was discontinued, and the patient was started on rehabilitation therapy. He was discharged 5 weeks later to a rehabilitation center.

The gait progressively improved during 6 months, when he could ambulate without assistance. The percutaneous endoscopic gastrostomy tube was removed 1 year after the procedure. A mild intermittent diplopia was unchanged.

The 1-year follow-up brain MRI showed no residual tumor (Figure 2) and there is no evidence of tumor recurrence 6 years after surgery.

Microscopic Examination

The tumor consisted mainly of areas of compact architecture with elongated bipolar cells arranged in fascicles, long club-shaped nuclei, and the so-called Antoni A tissue. Rarer, a loose-textured Antoni B tissue was elicited (Figure 3A). Tumor cells were strongly and uniformly immunoreactive for S-100 protein (Figure 3B). The interface between tumor and cerebral parenchyma from where it probably arose was ill-defined and showed abundant Rosenthal fibers (Figure 3C). Schwannoma of the fourth ventricle was the histologic diagnosis.

DISCUSSION

Schwannomas correspond to 8% of brain tumors and 25% of spinal tumors (22). Most (95%) are extra-axial, but they can also be found within the brain and spinal cord parenchyma or ventricular system (4, 16). The intraventricular location is the least common, with no more than 20 cases reported since 1950 (18), comprising 1 case in the third ventricle (17), 12 in the lateral ventricles (2, 4-8, 11-13, 19, 20, 25, 28), and 7 cases in the fourth ventricle (10, 14, 16, 18, 22, 29). The first fourth ventricle schwannoma ever reported (in 1957) was malignant and diagnosed postmortem (14). The current report is, to our knowledge, the eighth description of a fourth ventricle schwannoma (Table 1).

A presumptive diagnosis based on MRI features is rather difficult, as intraventricular schwannomas do not have unique characteristics. Ependymoma, choroid plexus papilloma, and juvenile pilocytic astrocytoma are common-enhancing tumors of the fourth ventricle and may encompass the differential diagnosis. Age and gender are not contributory to the diagnosis. The symptoms (mainly headaches) and the neurological signs (cerebellar and cranial nerve signs) are unspecific. Duration of symptoms can easily be longer than 1 year. The postoperative course is mostly related to preoperative deficits, tumor size, and floor invasion of the fourth ventricle. In the current case report, significant postoperative deficits were temporarily induced, but a mild diplopia was the only fixed neurological deficit. It should be noted that this tumor was the largest one reported to date. There were no descriptions of tumor recurrence on the previously published 7 surgical cases, which is in accord with the benign nature of these tumors. Although recurrence might be rare, MRI surveillance is warranted, particularly if total resection is not achieved.

To our knowledge, there are no Schwann cells covering central nervous system fibers. Schwann cells are present in peripheral fibers and in autonomic tissue surrounding vessels. A few investigators defend a single etiopathogenic theory (9, 22, 24), which would explain any intra-axial occurrence of a schwannoma, whereas others (4, 29) believe in a plurietiology model to explain the ectopy of this neoplasm. These attempts to explain the finding of intraspinal or intracerebral schwannomas can be extrapolated to the even least common ventricular schwannomas (17).

Benedickt (3), in 1874, first described nerve fibers (covered by Schwann cells) in the choroids plexus of the fourth ventricle. Stöhr (26), in 1922, and Rigs and Clary, in 1957 (23) suggested a neoplastic growth/hyperplasia of Schwann cells of the autonomic nervous fibers covering the arteries and the choroid plexus of the central nervous system. Adelman and