World Journal of Advanced Research and Reviews
eISSN: 2581-9615 CODEN (USA): WJARAI
Cross Ref DOI: 10.30574/wjarr
Journal homepage: https://wjarr.com/

(CASE REPORT)

Abducens nerve schwannoma: A case report and review of literature

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World Journal of Advanced Research and Reviews, 2025, 25(02), 1152-1157

Publication history: Received on 22 December 2024; revised on 31 January 2025; accepted on 02 February 2025

Article DOI: https://doi.org/10.30574/wjarr.2025.25.2.0329

Abstract

Background: Abducens nerve schwannomas are rare skull base tumors, frequently affect the cavernous sinus and develop near the brainstem. There is limited literature on their optimal management. Surgery and/or stereotactic radiosurgery are available as therapeutic options. Significant morbidity, including nerve function loss, can result from resection. Stereotactic radiosurgery is an option for treating other schwannomas.

Case Description: We report a case of a 30-year-old woman with no known medical history presented with a 4-year history of recurrent double vision with limited abduction on lateral gaze secondary to paresis of the right abducens nerve. A heterogeneous mass at the right cavernous sinus was found from neuro-imaging (MRI).

Considering the clinical status of patient and imaging findings, we decided to proceed with stereotactic radiosurgery. After She presented an improvement with absence of the diplopia and complete neurological recovery. The control MRI was carried out after 6 months, showing a clear reduction in tumor volume of at least 50%.

Conclusion: Gamma Knife Radio Surgery (GKRS) looks to be a safe and effective minimally invasive treatment option for such lesions, with excellent rates of tumor control and symptomatic improvement possible. Consequently, it is reasonable to consider GKRS as a first-line treatment option for oculomotor nerve schwannomas.

Keywords: Abducens nerve; Gamma Knife radiosurgery; Schwannomas; Stereotactic radiosurgery

1. Introduction

Schwannomas are slow-growing, benign tumors that originate from the Schwann cells of the neural sheath surrounding motor and sensory nerves. While they typically develop in motor nerve fibers, especially in association with neurofibromatosis, they can also sporadically affect other nerves, including the facial, hypoglossal, abducens, and trochlear nerves. They account for between 6 and 8% of all primary intracranial neoplasms; of these, vestibular schwannomas are the most common, followed by schwannomas arising from the trigeminal and lower cranial nerves. Abducens nerve schwannomas are very rare, and they are characterized by sensitive nervous disorders and pain of varying intensity.

We report a case of a patient who presented with persistent cephalalgia and diplopia for 2 months, whose neurological exam demonstrated a right abducens nerve paresis. Investigation demonstrated a heterogeneous tumor process centered on the right cavernous sinus responsible for invasion of the right VI nerve. the patient benefited from Gamma

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Knife radiosurgical treatment based on 12 Gy for her schwannoma of the right abducens nerve with complete clinical recovery revealed by the absence of diplopia and disappearance of headaches and a clear reduction in tumor volume noted by comparing pre-and post-GKRS images was considered significant.

2. Case report

A 30-year-old previously healthy woman was referred to the consultation department of neurosurgery for investigation of recurrent diplopia for approximately 4 years before and was treated by symptomatic treatment. For two months she has been complaining of an evolution of headache whose intensity was progressively increasing, associated with persistent diplopia and some episodes of vomiting.

The neurological exam of the patient demonstrated a horizontal binocular diplopia with limitation of abduction of the right eye secondary to paresis of the abducens nerve. Other ocular movements were preserved, without any loss of vision.

Magnetic resonance imaging (MRI) demonstrated a heterogeneous lesion with irregular contours, centered on the right cavernous sinus. The tumor was predominantly hyperintense on T1 images and Diffusion. On T2 images, the tumor was with an intermediate signal. After gadolinium infusion the tumor presented a heterogeneous enhancement, and measuring: 28x26X22; invading the right abducens nerve and the clivus anteriorly; pushing back the right internal carotid. The lesion suggests a schwannoma of the right abducens nerve.

The Patient underwent a Gamma Knife Radio Surgery (GKRS) with a Leksell Model G at the National Center for Rehabilitation and Neurosciences in Rabat. At this center, GKRS is performed on an outpatient basis, with the Leksell stereotactic frame placed after a local anesthetic is applied. After MRI studies are acquired under stereotactic conditions, the required MR images (plain T1, T2, and contrast images) are transferred to the planning software. Leksell Gamma Plan (Elekta) is then used to devise a plan, taking into consideration the prescription isodose, tumor margin doses, and maximal dosages, which are decided and confirmed after consulting with a radiation oncologist. Tumor volume was calculated. A multi-isocenter technique is employed to ensure at least 90% tumor coverage, with a prescription isodose of 50% and a tumor margin dose of 12 Gy. The duration of the procedure was 107 minutes. On that same day, the patient was released with a prescription for oral analgesics to be taken if needed.

During the early post radio-surgical treatment period, the patient presented absence of the diplopia with and the ocular motricity was completely recovered.

In order to compare the pre- and post-images GKRS and assess the efficacy of the treatment, the patient underwent a series of clinical examinations and had a first control MRI six months after the intervention, which revealed a significant reduction in tumor volume. A subsequent yearly radiological control was then planned.

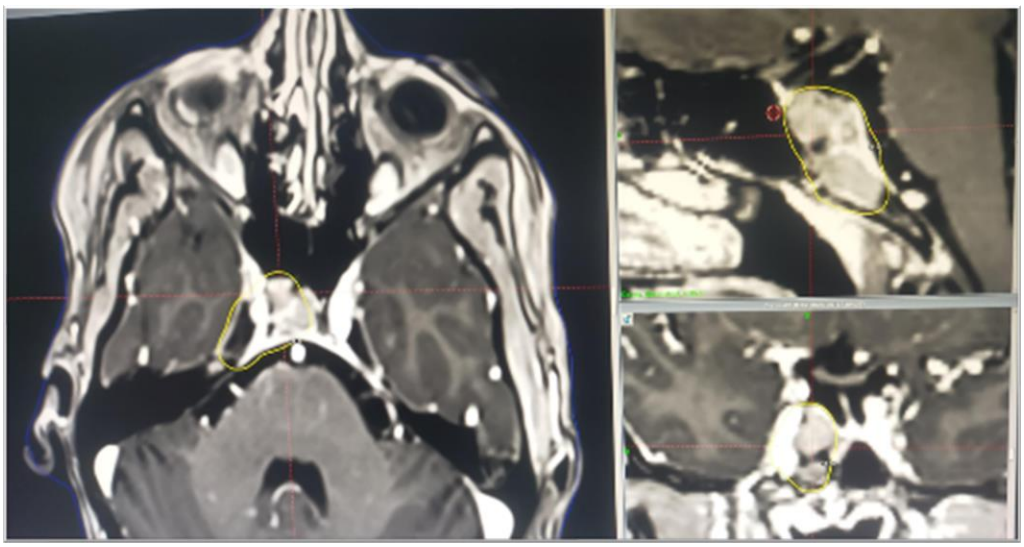

Figure 1 MRI of the brain obtained prior to GKRS: appearance suggestive of schwannoma of the right abducens nerve (measuring 28x26x22) and invading the right abducens nerve and the clivus anteriorly

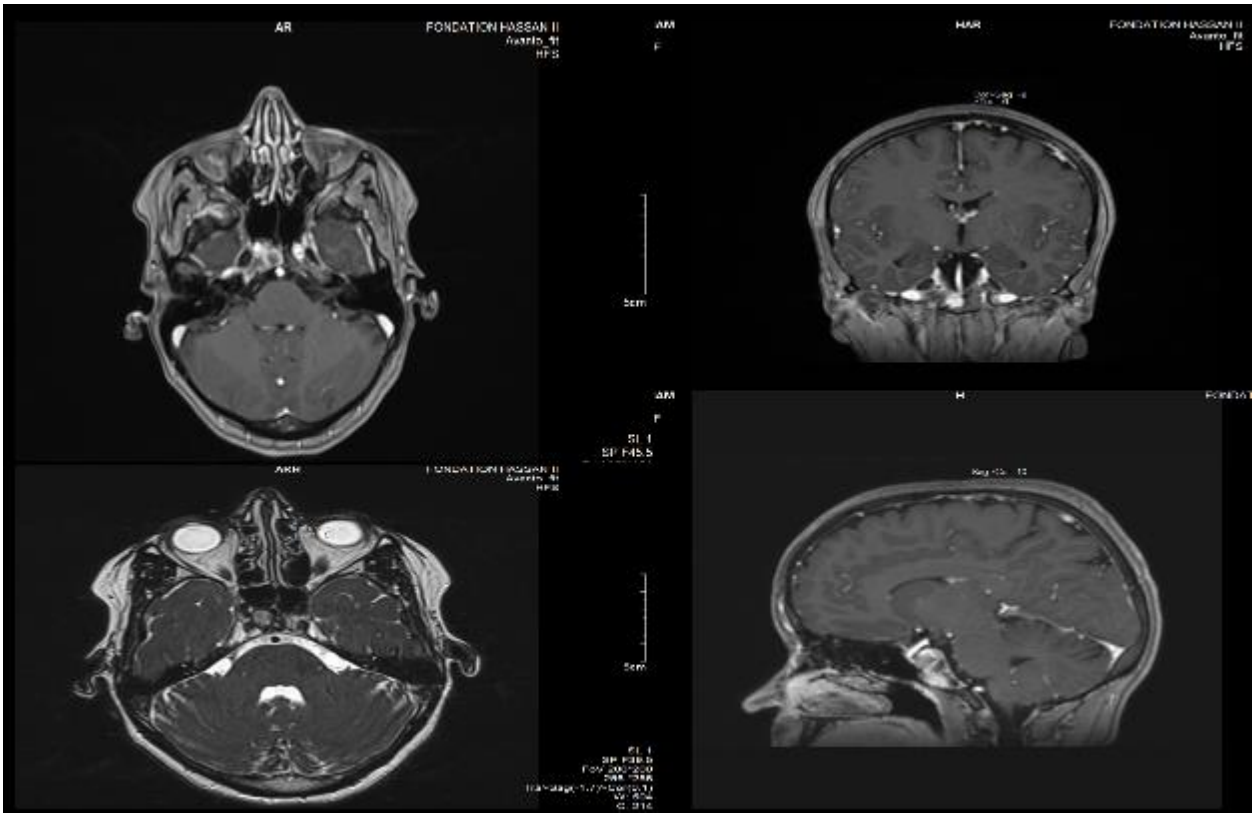

Figure 2 MRI images obtained 6 months after GKRS revealing a reduction in tumor size

3. Discussion

Intracranial schwannomas are a type of benign tumor that make up 8–10% of all primary brain tumors. The vestibular cranial nerves are the most common source of them, followed by the trigeminal, facial, and lower cranial nerves; pure motor CN (3rd, 4th, and 6th) is rarely affected. Schwannomas arising from the ocular motor nerves are uncommon, and AN schwannomas, in particular, are very rare. They can be found in cisternal, cavernous, or cisterno-cavernous are, but more common in women in their fourth or fifth decade of life (1-15).

In the English literature, only 34 cases have been reported of intracranial schwannomas of the 6th cranial nerve pathologically confirmed to date (20, 21-24). and the Radiosurgery's role in treating nonvestibular schwannomas has been well-documented with positive outcomes (25-26).

The pre-pontine cistern, cavernous sinus, and orbit are the usual locations for sixth CN schwannoma (11,16,23). They are more common in females in their fourth or fifth decade of life, but they are equally likely to occur in the prepontine or cavernous sinus locations. Although coexisting trigeminal, facial, and vestibulocochlear nerve dysfunction can also occur, the most common presentation is horizontal diplopia, which is the general presenting feature of isolated sixth CN palsy. Additional symptoms that have been reported include headache, blurred vision, proptosis, and emotional instability (5, 11, 18, 23). The neurological examination revealed a right abducens nerve paresis in our patient, who also had a headache and horizontal diplopia.

Schwannomas are extra-axial tumors that show up on CT with strong contrast enhancement as iso-dense to gray matter. On T1-weighted MRI images, these tumors are usually hypo to isointense; on T2-weighted images, they are hyperintense; and they intensify following the administration of contrast. Secondary hemorrhagic and cystic transformations are uncommon (8,27,28). Even though isolated AN palsy aids in identifying the nerve of origin, linked trochlear or trigeminal nerve involvement—particularly in cases of large tumors in the cavernous sinus—can cause confusion. The sixth CN schwannomas, which can occasionally resemble meningiomas, are followed by chordoma, chondrosarcoma, angiofibroma, pituitary adenomas, and trigeminal schwannomas. Other nerve-derived schwannomas, hemangiomas, plasmacytomas, and epidermoid are among them. 24 Additionally, there have been reports of metastatic lesions from renal or pulmonary sources. (1,14,24).

In our case, the imaging appearance matched the radiological features of schwannomas that had been previously described (29). In Heterogeneous T1 and diffusion hyperintensity, an intermediate signal on T2 weighted images, and strong contrast enhancement are highlights of MRI. One of the first cases of abducens nerve schwannoma involving the cerebellopontine segment was reported by Bing-Huan in 1981. The first case of cavernous sinus schwannoma of the sixth cranial nerve was documented by Leunda et al. a year later (30). Since then, there have only been 16 documented cases of abducens schwannomas originating from the cavernous segment.

Tung and al. divided these lesions into two categories. Type 1 was caused by the cavernous sinus and manifested as diplopia and abducens palsy. With symptoms of elevated intracranial pressure, obstructive hydrocephalus, and sixth nerve palsy, type 2 was primarily caused by the prepontine cistern prior to the cavernous segment; other cranial nerves may also be affected (31). This classification aids in determining the viability of nonsurgical alternatives, the anticipated extent of resection, and the surgical approach.

Currently, radiosurgery and/or surgery are used to treat intracranial schwannomas. There are currently no guidelines for the preferred management of AN schwannomas due to their rarity and the resulting paucity of literature. Previously, the only treatment options for Tung Type I and II tumors were surgery through the transcavernous and subtemporal/suboccipital corridors, respectively. For obvious reasons, such as perioperative mortality, hemorrhage, damage to CNs, exacerbation of preexisting deficits, aphasia, and motor deficits, Tung et al. claimed that Type II lesions in the cavernous sinus were simpler to remove than Type I lesions (32-35).

For patients who would rather have a less invasive treatment option or who are at intolerable risk from surgery, radiosurgery is a good choice. It is advised to counsel the patient on less invasive and surgical options (10,36,37). In stereotactic radiosurgery, radiation beams are conformally focused onto a specific target to reduce spillover onto nearby normal brain parenchyma. GKRS, Cyber Knife, and linear accelerator (LINAC)-based platforms are among the modalities. GKRS is arguably the most widely used of these. Numerous studies have confirmed the safety and effectiveness of GKRS in treating vestibular schwannomas, making it one of the most frequently treated tumors. The effectiveness of stereotactic radiosurgery in nonvestibular schwannomas has been reported in far fewer cases (6,12,38-42).

4. Conclusion

Abducens nerve schwannomas are rare intracranial tumors. GKRS appears to be a safe and effective minimally invasive method for treating such lesions, with excellent tumor control rates and symptomatic improvement achievable. Therefore, it is reasonable to regard GKRS as the preferred initial treatment for this uncommon pathology.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent and verbal permission were obtained from the patient before the submission of this article. In addition, this article follows both the Consensus-based Clinical Case Reporting Guideline and the Recommendations for the Conducting, Reporting, Editing, and Publication of Scholarly Work in Medical Journals.

References

[1] Acharya R, Husain S, Chhabra SS, Patir R, Bhalla S, Seghal AD: Sixth nerve schwannoma: a case report with literature review. Neurol Sci 24:74–79, 2003

[2] Beppu T, Yoshida Y, Wada T, Arai H, Suzuki M, Kuroda K, et al: Trochlear and abducens nerve neurinomas accompa- nied by a cerebellopontine angle meningioma.

[3] Celli P, Ferrante L, Acqui M, Mastronardi L, Fortuna A, Palma L: Neurinoma of the third, fourth, and sixth cranial nerves: a survey and report of a new fourth nerve case. Surg Neurol 38:216–224, 1992

[4] Chakrabarti KB, Doughty D, Plowman PN: Stereotactic mul- tiple arc radiotherapy. II. Cranial neuroma. Br J Neurosurg 10:577–583, 1996

[5] Chen BH: Neurinoma of the abducens nerve. Neurosurgery 9:64–66, 1981

[6] Elsharkawy M, Xu Z, Schlesinger D, Sheehan JP: Gamma Knife surgery for nonvestibular schwannomas: radiological and clinical outcomes. J Neurosurg 116:66–72, 2012

[7] Erlich SA, Tymianski M, Kiehl TR: Cellular schwannoma of the abducens nerve: case report and review of the literature. Clin Neurol Neurosurg 111:467–471, 2009

[8] Ginsberg F, Peyster RG, Rose WS, Drapkin AJ: Sixth nerve schwannoma: MR and CT demonstration. J Comput Assist Tomogr 12:482–484, 1988

[9] Hayashi M, Chernov M, Tamura N, Yomo S, Ochiai T, Nagai M, et al: Gamma Knife surgery for abducent nerve schwan- noma. Report of 4 cases. J Neurosurg 113 Suppl:136–143, 2010

[10] Ichimi K, Yoshida J, Inao S, Wakabayashi T: Abducens nerve neurinoma—case report. Neurol Med Chir (Tokyo) 37:197– 200, 1997

[11] Irace C, Davì G, Corona C, Candino M, Usai S, Gambacorta M: Isolated intraorbital schwannoma arising from the abdu- cens nerve. Acta Neurochir (Wien) 150:1209–1210, 2008

[12] Kim IY, Kondziolka D, Niranjan A, Flickinger JC, Lunsford LD: Gamma Knife surgery for schwannomas originating from cranial nerves III, IV, and VI. J Neurosurg 109 Sup- pl:149–153, 2008

[13] Lanotte M, Giordana MT, Forni C, Pagni CA: Schwannoma of the cavernous sinus. Case report and review of the litera- ture. J Neurosurg Sci 36:233–238, 1992

[14] Lo PA, Harper CG, Besser M: Intracavernous schwannoma of the abducens nerve: a review of the clinical features, ra- diology and pathology of an unusual case. J Clin Neurosci 8:357–360, 2001

[15] Mascarenhas L, Magalhaes Z, Honavar M, Romao H, Re- sende M, Resende Pereira J, et al: Schwannoma of the abdu- cens nerve in the cavernous sinus. Acta Neurochir (Wien) 146:389–392, 2004

[16] Moses JE, Vermani N, Bansal SK: Preoperative clinico-ra- diological diagnosis of schwannoma arising from cavernous segment of abducens nerve. Neurol India 59:471–473, 2011

[17] Nakagawa T, Uchida K, Ozveren MF, Kawase T: Abducens schwannoma inside the cavernous sinus proper: case report. Surg Neurol 61:559–563, 2004

[18] Nakamura M, Carvalho GA, Samii M: Abducens nerve schwannoma: a case report and review of the literature. Surg Neurol 57:183–189, 2002

[19] Okada Y, Shima T, Nishida M, Okita S: Large sixth nerve neuroma involving the prepontine region: case report. Neu- rosurgery 40:608–610, 1997

[20] Park JH, Cho YH, Kim JH, Lee JK, Kim CJ: Abducens nerve schwannoma: case report and review of the literature. Neuro- surg Rev 32:375–378, 2009

[21] Showalter TN, Werner-Wasik M, Curran WJ Jr, Friedman DP, Xu X, Andrews DW: Stereotactic radiosurgery and fraction- ated stereotactic radiotherapy for the treatment of nonacous- tic cranial nerve schwannomas. Neurosurgery 63:734–740, 2008

[22] Suetake K, Kurokawa Y, Uede T, Momota H, Hashi K: A case of abducens neurinoma mimicking acoustic neurinoma. Comput Med Imaging Graph 22:257–261, 1998

[23] Tung H, Chen T, Weiss MH: Sixth nerve schwannomas. Re- port of two cases. J Neurosurg 75:638–641, 1991

[24] Vachata P, Sames M: Abducens nerve schwannoma mim- icking intrinsic brainstem tumor. Acta Neurochir (Wien) 151:1281–1287, 2009

[25] Hasegawa T, Kida Y, Kobayashi T, Yoshimoto M, Mori Y, Yoshida J: Long-term outcomes in patients with vestibular schwannomas treated using gamma knife surgery: 10-year follow up. J Neurosurg 102:10–16, 2005

[26] Kondziolka D, Lunsford LD, McLaughlin MR, Flickinger JC: Long-term outcomes after radiosurgery for acoustic neuro- mas. N Engl J Med 339:1426–1433, 1998

[27] Hansman ML, Hoover ED, Peyster RG: Sixth nerve neuroma in the cavernous sinus: CT features. J Comput Assist To- mogr 10:1030–1032, 1986

[28] Kingsley DP, Brooks GB, Leung AW, Johnson MA: Acoustic neuromas: evaluation by magnetic resonance imaging. AJNR Am J Neuroradiol 6:1–5, 1985

[29] Korchi AM, Cuvinciuc V, Caetano J, Becker M, Lovblad KO, Vargas MI. Imaging of the cavernous sinus lesions. Diagn Interv Imaging 2014;95:849-59.

[30] Leunda G, Vaquero J, Cabezudo J, Garcia-Uria J, Bravo G. Schwannoma of the oculomotor nerves. Report of four cases. J Neurosurg 1982;57:563-5.

[31] Sarma S, Sekhar LN, Schessel DA: Nonvestibular schwan- nomas of the brain: a 7-year experience. Neurosurgery 50:437–448, 438–439, 2002

[32] Lee JD, Kim SH, Song MH, Lee HK, Lee WS: Management of facial nerve schwannoma in patients with favorable facial function. Laryngoscope 117:1063–1068, 2007

[33] Samii M, Migliori MM, Tatagiba M, Babu R: Surgical treat- ment of trigeminal schwannomas. J Neurosurg 82:711–718, 1995

[34] Sharma BS, Ahmad FU, Chandra PS, Mahapatra AK: Tri- geminal schwannomas: experience with 68 cases. J Clin Neurosci 15:738–743, 2008

[35] Tung H, Chen T, Weiss MH: Sixth nerve schwannomas. Re- port of two cases. J Neurosurg 75:638–641, 1991

[36] Peciu-Florianu I, Tuleasca C, Comps JN, Schiappacasse L, Zeverino M, Daniel RT, et al. Radiosurgery in trochlear and abducens nerve schwannomas: Case series and systematic review. Acta Neurochir (Wien) 2017;159:2409-18.

[37] Prasad GL, Sharma MS, Kale SS, Agrawal D, Singh M, Sharma BS. Gamma knife radiosurgery in the treatment of abducens nerve schwannomas: A retrospective study. J Neurosurg 2016;125:832-7.

[38] Mabanta SR, Buatti JM, Friedman WA, Meeks SL, Men- denhall WM, Bova FJ: Linear accelerator radiosurgery for nonacoustic schwannomas. Int J Radiat Oncol Biol Phys 43:545–548, 1999

[39] Phi JH, Paek SH, Chung HT, Jeong SS, Park CK, Jung HW, et al: Gamma Knife surgery and trigeminal schwannoma: is it possible to preserve cranial nerve function? J Neurosurg 107:727–732, 2007

[40] Sheehan J, Yen CP, Arkha Y, Schlesinger D, Steiner L: Gam- ma knife surgery for trigeminal schwannoma. J Neurosurg 106:839–845, 2007

[41] Showalter TN, Werner-Wasik M, Curran WJ Jr, Friedman DP, Xu X, Andrews DW: Stereotactic radiosurgery and fraction- ated stereotactic radiotherapy for the treatment of nonacous- tic cranial nerve schwannomas. Neurosurgery 63:734–740, 2008

[42] Zhang N, Pan L, Dai JZ, Wang BJ, Wang EM, Cai PW: Gam- ma knife radiosurgery for jugular foramen schwannomas. J Neurosurg 97 (5 Suppl):456–458, 2002.