Interdisciplinary Neurosurgery: Advanced Techniques and Case Management 2 (2015) 111–114

Contents lists available at ScienceDirect

Interdisciplinary Neurosurgery: Advanced Techniques and Case Management

journal homepage: www.inat-journal.com

Case Reports & Case Series (CRP)

Isolated trochlear nerve schwannoma presenting with diplopia: A case report and literature review

Mohammad Samadian, Navid Farzin, Mehrdad Hosseinzadeh Bakhtevari *, Mohammad Hallajnejad, Omidvar Rezaei

Department of Neurosurgery, Loghman e Hakim Hospital, Shahid Beheshti University of Medical Sciences, Tehran, Iran

ARTICLE INFO

Article history:
Received 2 February 2015
Revised 25 March 2015
Accepted 29 March 2015

Keywords:
Intraoperative diagnosis
Neurofibromatosis 2
Schwannoma
Trochlear nerve

ABSTRACT

Background: Trochlear nerve schwannoma is a very rare tumor encountered especially in patients without type 2 neurofibromatosis (NF2). Most of the time, this tumor is diagnosed intraoperatively. We describe a rare case of trochlear nerve schwannoma.

Clinical Presentation: A 63-year-old male presented with generalized headache from 8 months earlier, without nausea and vomiting. The headache had worsened during the last months. Clinically, he suffered from transient diplopia. Magnetic resonance imaging (MRI) demonstrated a mass in the left ambient and interpeduncular cisterns that was compressing the midbrain and upper pons. The preoperative impression was of trigeminal schwannoma. However, intraoperatively, the tumor originated in the trochlear nerve. The mass was removed entirely via left suboccipital retrosigmoid approach. The histopathology diagnosis confirmed schwannoma. The patient's diplopia did not improve postoperatively.

Conclusion: Trochlear nerve schwannoma is a rare type of schwannoma and in almost all of the cases is diagnosed intraoperatively. There are three types of trochlear nerve schwannoma according to the classification proposed for trigeminal schwannoma: cisternal type—confined to the precavernous segment of the trochlear nerve; cistocavernous type—invading the cavernous sinus and the retroclival and retropetrosal cistern; cavernous type—located in the middle cranial fossa on the cavernous or paracavernous segment of the fourth cranial nerves, with or without cavernous sinus invasion. The cisternal type was the most common type encountered in previous studies. The clinical signs and symptoms of trochlear nerve schwannoma are similar to the trigeminal schwannoma and should be considered in the differential diagnosis of trigeminal schwannoma.

Introduction

Intracranial schwannomas account for 8% of all intracranial tumors and most frequently arise from the vestibular nerve of the trigeminal nerve [1]. Intracranial schwannomas originate in a number of mixed cranial nerves and rarely originate from pure motor nerves, such as the trochlear nerve, especially in the absence of neurofibromatosis type 2 (NF2) [1–3]. Trochlear nerve schwannoma does not always present with trochlear nerve palsy and therefore, preoperative diagnosis based on neuroimaging finding is difficult [1]. We describe a rare case of trochlear nerve schwannoma that was diagnosed by surgical and histological findings.

Material and methods

Case presentation

A 63-year-old male presented with non-positional non-pulsatile headache for 8 months in duration, without nausea and vomiting. The headache had worsened recently and was refractory to common medications. Also, he suffered from transient diplopia. He had never experienced any facial pain or facial palsy. Eye movement was intact symmetrically and other cranial nerves examinations were unremarkable. Brain computed tomography (CT) revealed a round hypo-density in the left ambient cistern, with compression of the brain stem (Fig. 1A). Magnetic resonance imaging (MRI) demonstrated a cystic mass in the left ambient cistern, with extension to interpeduncular cistern, which was is intense to hypo-intense on T1-weighted (Fig. 1B) images and is intense to hyper intense on T2-weighted images (Fig. 1C, D). A heterogeneous enhancement was observed after administration of gadolinium (Fig. 2).

The patient underwent surgery via left sub-occipital retro-sigmoid approach (Fig. 3), in lateral position with neuromonitoring assistance

* Corresponding author at: Neurosurgery Department, Loghman e Hakim Hospital, Kamali Avenue, Kargar Jonubi Street, Tehran, Iran. Tel.: +98 912 1874506 (Mobile); Tel./fax: +98 21 55414065.

E-mail addresses: mdsamadian@gmail.com (M. Samadian), navidfarzin2015@gmail.com (N. Farzin), mehrdaada@yahoo.com (M.H. Bakhtevari), hallajnejad@gmail.com (M. Hallajnejad), info@drrezaee.ir (O. Rezaei).

http://dx.doi.org/10.1016/j.inat.2015.03.006

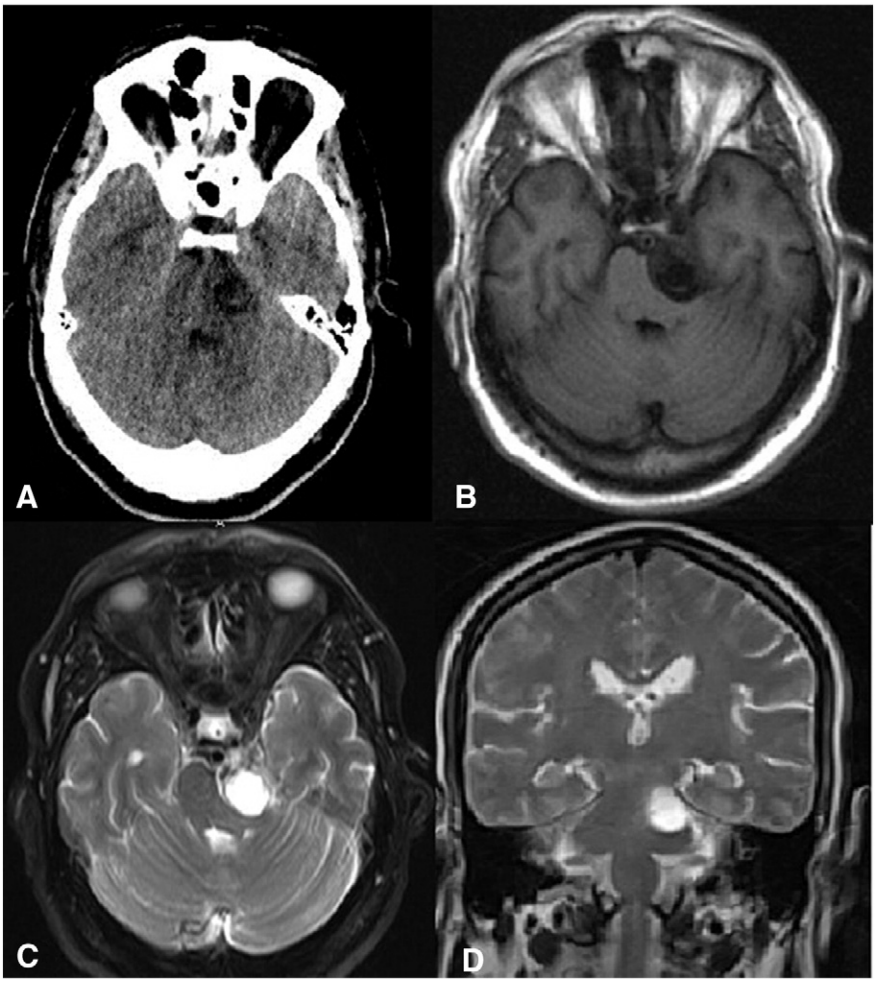

Fig. 1. Brain CT scan reveals a round hypodensity in the left ambient cistern in compression to the brain stem (A), which is isointense to hypointense on T1-weighted MRI (B), and isointense to hyperintense on T2-weighted MRI (C, D).

of the 5th and 7th cranial nerves. This approach was selected because of the extension of the tumor to cerebellopontine angle. During surgery, the tumor was dissected from the surrounding structures and after observation of trigeminal nerve, the surgeons noted that the tumor was superior to the trigeminal nerve and had no adhesion to it. The tumor was partially resected and after the dissection of the tentorium, the trochlear nerve continued to the residual tumor. Gross total mass removal was achieved, together with the trochlear nerve

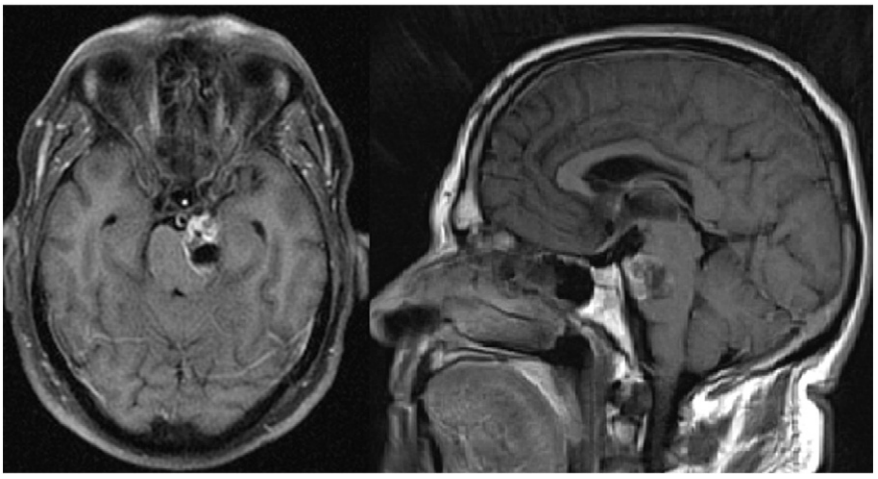

Fig. 2. The T2-weighted MRI (right) demonstrated a cystic lesion in the left side of the midbrain with heterogeneous enhancement (left).

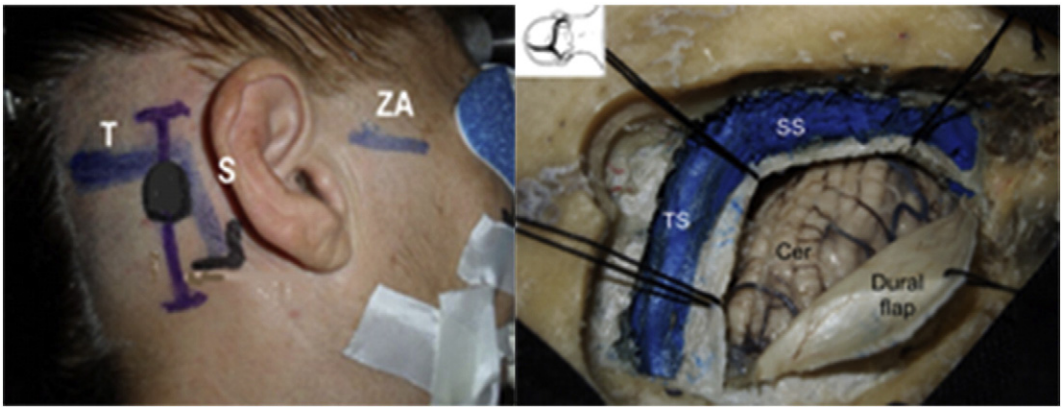

Fig. 3. View of the retrosigmoid suboccipital approach. C: cerebellum, S: sigmoid sinus, T transverse sinus, ZA: zygomatic arch.

(Fig. 4A, B). Because of the manipulation of the trochlear nerve during surgery in addition to probable injury to the nerve with postoperative interventions such as gamma knife and their potential hazardous to the trochlear nerve, we decided to sacrifice trochlear nerve to gain total resection and prevent further recurrence and using ophthalmic corrective surgery to repair the deficit. The histological examination established the diagnosis of schwannoma. After surgery, the headache improved. Unfortunately, diplopia persisted because of trochlear nerve palsy. Follow-up MRI demonstrated total resection of the tumor without any residue (Fig. 5). The patient was referred to an orbital surgeon for corrective surgery.

Discussion

The trochlear nerve schwannoma is a very rare tumor. Only 31 cases were reported in the literature so far [1–15]. The first case was reported in 1976 by Kings and associates [18]. Preoperative diagnosis of trochlear schwannoma is very difficult and the origin of the tumor is identified by surgical findings (Table 1). Of all cases up to now, 22 cases were diagnosed after intraoperative findings, including our case.

Trochlear nerve schwannoma is of three types, according to the classification proposed for trigeminal schwannoma. The cisternal type is confined to the precavernous segment of the trochlear nerve. The cistocavernous type invades the cavernous sinus and retroclival and retropetrosal cistern, whereas the cavernous type is located in the middle cranial fossa on the cavernous or paracavernous segment of the fourth cranial nerves, with or without cavernous sinus invasion. The cisternal type was the most common type in the previous cases [1].

The most MRI findings compatible with trochlear schwannoma consist of an isointense mass in both T1 and T2 weighted images, with avid enhancement after gadolinium injection [1,6,7]. The most frequent location for trochlear nerve schwannoma is the ambient or cerebropointine cistern [1,12].

According to the literature review of Kohma et al. [1], the characteristic symptoms of trochlear schwannoma are represented by diplopia, hemiparesis and cerebellar signs, such as ataxia, dysmetria and nystagmus. Trochlear nerve palsy was detected only in less than half of the cases [1] and absence of diplopia in several cases could be explained by a gradual compensation by other extraocular muscles for the loss of the superior oblique nerve function [3]. Hemiparesis is more common in the trochlear schwannoma rather than the trigeminal schwannoma [1].

Trochlear schwannomas are usually located beneath the edge of the tentorium, and therefore they may extend to the interpeduncular cistern, in contrast to trigeminal schwannoma, and cause motor dysfunction due to cerebral peduncles compression [1,11].

Regarding the location of tumor and imaging findings, trigeminal schwannoma, epidermoid cyst, ependymoma and intrinsic brain tumors, represent the differential diagnoses of trochlear schwannoma [1,2,6–11,14].

The cisternal type was the most common type of trochlear schwannoma, while the sub-temporal approach was the most common approach that has been used [1]. The posterior transpetrosal approach is another approach that may be used, as it provides a wide exposure of the cerebellopontine angle, ambient cistern and the identification of the involved cranial nerves, while the major drawback is that it requires complicated skull base surgery [1,16].

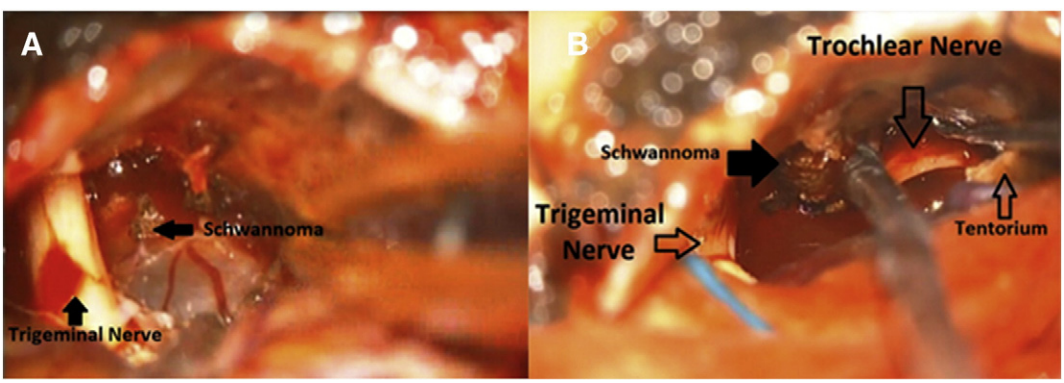

Fig. 4. (A) Intraoperative view of the trigeminal nerve and position of the tumor. (B) After resection of the infratentorial part of the tumor and dissection of tentorium, the tumor was adhered to the trochlear nerve.

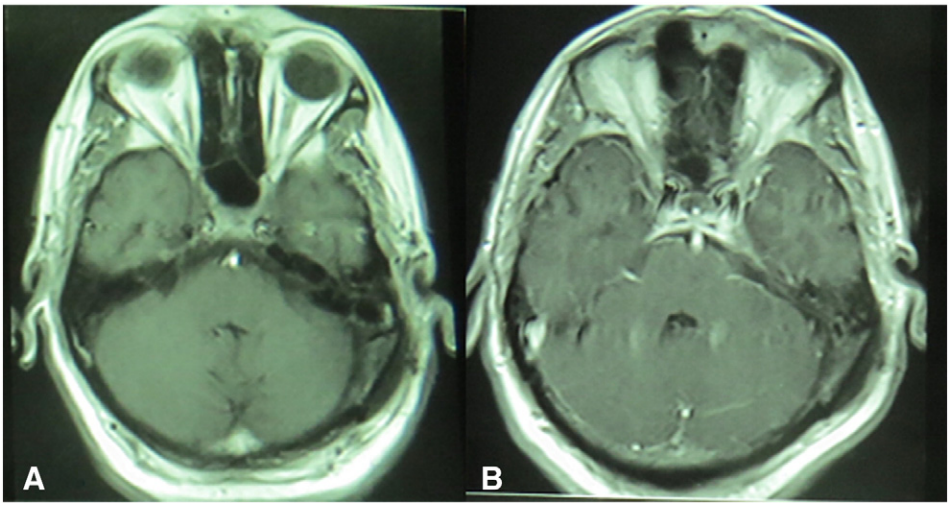

Fig. 5. T1-weighted MRI before (A) and after gadolinium injection (B) one month after surgery.

We operate the trochlear schwannoma via retro-sigmoid suboccipital approach because of tumor extension to the left cerebellopontine angle. Abe et al. [6] and Mauice-Williams [17] also used this approach.

There was only one case in which the trochlear nerve function improved postoperatively after 2 years of follow-up [5]. Fourteen cases presented with diplopia before operation whereas 12 cases developed diplopia after surgery. Our case had diplopia before surgery that did not improved significantly after surgery [1].

Conclusion

Trochlear nerve schwannoma is a rare type of schwannoma and in almost all of the cases is diagnosed intraoperatively. There are three types of trochlear nerve schwannoma according to the classification proposed for trigeminal schwannoma: cisternal type—confined to the precavernous segment of the trochlear nerve; cistocavernous type—invading the cavernous sinus and the retroclival and retropetrosal cistern; cavernous type—located in the middle cranial fossa on the cavernous or paracavernous segment of the fourth cranial nerves, with or without cavernous sinus invasion. The cisternal type was the most common type encountered in previous studies. The clinical signs and symptoms of trochlear nerve schwannoma are similar to the trigeminal schwannoma and should be considered in the differential diagnosis of trigeminal schwannoma.

The trochlear nerve schwannoma should be considered in all cases of brain stem tumors that have diplopia without significant facial paresthesia or palsy, especially those tumors which extend to the ambient or interpeduncular cisterns. Regarding the similarity of imaging findings in trochlear and trigeminal schwannoma, history, physical examination and intraoperative findings remain our trustworthy instruments for distinguishing between trochlear and trigeminal schwannomas.

Table 1
Summary of 31 surgical cases of trochlear schwannoma according to type and surgical approach.

Regions of tumors (type)	
Cisternal	28 (91%)
Cisternocavernous	2 (6%)
Cavernous	1 (3%)
Surgical approach	
Subtemporal transtentorial	18
Lateral suboccipital	2
Petrosal	4
Pterional	5
Other	2

Conflict of interest

All authors certify that they have no affiliations with or involvement in any organization or entity with any financial interest or non-financial interest in the subject matter or materials discussed in this manuscript.

There is no funding or conflict of interest.
There are no financial disclosures.

References

[1] Kohma M, Murakami K, Endo T, Watanabe M, Tominaga T. Surgical and histological observations of trochlear schwannoma: case report. Neurol Med Chir 2009;49(5):217–20.
[2] Matusi T, Morikawa E, Morimoto T, Asano T. Presigmoid transpetrosal approach for treatment of a large trochlear nerve schwannoma. Neurol Med Chir 2002;42:31–5.
[3] Du R, Dhoot J, McDermott MW, Gupa N. Cystic schwannoma of anterior tentorial hiatus: case report and review of the literature. Pediatr Neurosurg 2003;38:167–73.
[4] Barealena T, Leoni C, Trossello MP, Rinaldi MF, Cianfoni A, Caprara G, et al. Hourglass cystic schwannoma of the trochlear nerve. Acta Biomed 2010;81(2):147–50.
[5] Younes WM, Hermann EJ, Krauss JK. Cisternal trochlear nerve schwannoma: improvement of diplopia after subtotal tumour excision. 2012;26(1):107–9.
[6] Abe T, Iwata T, Shimazu M, Iida M, Izumiyama H, Matsumoto K, et al. Two cases of trochlear nerve schwannoma. No Shinkei Geka 1994;22:371–5 [Jpn, with Eng abstract].
[7] Beppu T, Yoshida Y, Wada T, Arai H, Suzuki M, Kuroda K, et al. Trochlear and abducens nerve schwannomas accompanied by a cerebellopontine angle meningioma: case report. Neurol Med Chir (Tokyo) 1997;37:416–21.
[8] Celli P, Ferrante L, Acqui M, Mastronardi L, Fortuna A, Palma L. Schwannoma of the third, fourth, and sixth cranial nerves: a survey and report of a new fourth nerve case. Surg Neurol 1992;38:216–24.
[9] Garen PD, Harper CG, Teo C, Johnston IH. Cystic schwannoma of the trochlear nerve mimicking a brain-stem tumor. Case report. J Neurosurg 1987;67:928–30.
[10] Jackowski A, Weiner G, O'Reilly G. Trochlear nerve schwannomas: a case report and literature review. Br J Neurosurg 1994;8:219–23.
[11] Nadkami TD, Goel A. Trochlear nerve schwannoma presenting as pathological laughter. Br J Neurosurg 1999;13:212–3.
[12] Ohba S, Miwa T, Kawase T. Trochlear nerve schwannoma with intratumoral hemorrhage: case report. Neurosurgery 2006;58:791.
[13] Santoreneos S, Hanieh A, Jorgensen RE. Trochlear nerve schwannomas occurring in patients without neurofibromatosis: case report and review of the literature. Neurosurgery 1997;41:282–7.
[14] Shenoy SN, Raja A. Cystic trochlear nerve schwannoma mimicking intrinsic brain stem tumour. Br J Neurosurg 2004;18:183–6.
[15] Tokuriki Y, Yamashita J, Kikuchi H, Asato R, Handa H. Trochlear nerve schwannoma: case report. Neurol Med Chir (Tokyo) 1988;28:70–3.
[16] Sincoff EH, McMenomey SO, Delashaw Jr JB. Posterior transpetrosal approach: less is more. Neurosurgery 2007;60(2 Suppl. 1):ONS53–9.
[17] mauice-williams RS. Isolated schwannoma of the fourth cranial nerve: case report. J Neurol Neurosurg Psychiatry 1989;52(12):1442–3.
[18] King JS. Trochlear nerve sheath tumor: case report. J Neurosurg 1976;44(2):245–7.