

Vidian Nerve Schwannoma: A Rare Skull-Base Neoplasm Presenting with Ocular Manifestations: A Case Report and Literature Review

Farzad A Masroor, MD; Jason Gilde, MD; Jonathan Liang, MD

Perm J 2018;22:18-021

E-pub: 10/01/2018

<https://doi.org/10.7812/TPP/18-021>

ABSTRACT

Introduction: Vidian nerve schwannomas are exceedingly rare, with only 7 cases reported since 2006. Patients presenting with ocular symptoms have been reported in only 1 case.

Case Presentation: A 54-year-old woman presented with a 3-month history of right periorbital pressure, third cranial nerve palsy, and visual field defect. Imaging results showed a right sphenoid skull-base mass with obliteration of the vidian canal that extended into the pterygopalatine fossa. The patient underwent an extended endoscopic resection with pterygopalatine fossa dissection. Pathologic findings demonstrated a schwannoma.

Discussion: A literature review showed that this is the second reported case of a vidian nerve schwannoma presenting with ocular symptoms and that endoscopic resections are becoming the standard of care. Practitioners should be aware that vidian nerve schwannomas can present as a skull-base mass with predominantly ocular symptoms, including vision loss, secondary to mass effect. Consideration should be given to this entity in the setting of typical radiographic and histopathologic characteristics. Endoscopic approaches to resection are safe and have low morbidity.

INTRODUCTION

A schwannoma (also known as a neuroma, neurinoma, or neurilemoma) is a benign nerve sheath tumor composed of Schwann cells, which produce myelin that insulate peripheral nerves.¹ The head and neck are frequent locations for schwannomas, accounting for 25% to 45% of their distribution.² Schwannomas can arise from any cranial nerve, with the exception of the first and second cranial nerves, which lack Schwann cells.³ The most frequently affected cranial nerves are the vestibular and trigeminal nerves, with the former accounting for 8% of intracranial neoplasms and 80% to 90% of cerebellopontine angle tumors in adults.⁴ Schwannomas are also the second most common tumor of the parapharyngeal space, behind tumors of salivary gland origin.⁵ The facial nerve is the third most commonly involved cranial nerve,⁶ but involvement of the vidian nerve branch is exceedingly rare. There are only 7 reported cases in the literature.^{3,7-12}

Schwannomas, although benign, lead to morbidity by way of mass effect on the involved nerve and nearby structures.

We present a case of a large vidian nerve schwannoma primarily manifesting with ocular symptoms, which had not yet been described in the literature, according to our literature review.

CASE PRESENTATION

Presenting Concerns

A 54-year-old woman presented to our Otolaryngology Department with 3 months of right-sided periorbital pressure, diplopia, and visual disturbances. Physical examination revealed a right third cranial nerve palsy, numbness in the right cheek, and drooping of the right eyelid. Ophthalmologic findings demonstrated a right third cranial nerve palsy, superior visual field defect, alterations in hue, and difficulty with accommodation. Nasal endoscopic findings showed a fibrous mass extending from the right sphenothmoidal recess.

On computed tomography, a 4.1 cm × 3.6 cm × 2.1 cm expansile mass was noted in the right sphenoid region with evidence of bony remodeling. The cavernous portion of the right carotid artery was dehiscent, and the orbital apex was

compressed by the mass. On magnetic resonance imaging, there was a mass that was hypointense on T1-weighted imaging and hyperintense on T2-weighted imaging that demonstrated the same findings as did computed tomography without evidence of intracranial invasion (Figure 1).



Figure 1. Axial T2-weighted magnetic resonance image of the cranium. Note the hyperintensity of the mass (white asterisk) with expansion and mass effect on the cavernous carotid artery (red arrow) and orbital apex (yellow arrow).

Therapeutic Intervention and Treatment

The patient underwent an extended endoscopic resection that included bilateral ethmoidectomies and sphenoidotomies, posterior septectomy, and pterygopalatine fossa (PPF) dissection. Intraoperatively, multiple islands of bony dehiscence were noted along the skull base. The dura mater was intact, and there was no evidence of a cerebrospinal fluid leak. Middle turbinate and septal mucosal free grafts were used to cover the cavernous dehiscence, and a posteriorly based septal flap was used to cover the carotid dehiscence. The posterior lamina papyracea was opened for decompression of the orbital apex.

Farzad A Masroor, MD, is a Resident Physician in the Department of Otolaryngology-Head and Neck Surgery at the Oakland Medical Center in CA (farzad.a.masroor@kp.org). Jason Gilde, MD, is a Fellow Physician in the Department of Otolaryngology-Head and Neck Surgery at the Loma Linda University Medical Center in CA (jason.gilde@gmail.com). Jonathan Liang, MD, is a Surgeon in the Department of Otolaryngology-Head and Neck Surgery at the Oakland Medical Center in CA (jonathan.liang@kp.org).

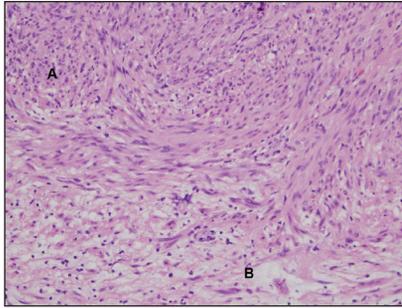


Figure 2. Hematoxylin and eosin-stained section, high-power view (magnification x400), showing Antoni Type A pattern with densely packed spindle cells having palisading nuclei (A), and Antoni Type B pattern with sparse cellularity in a myxoid stroma (B).

Follow-up and Outcomes

Pathologic analysis demonstrated uniform S100-positive spindle cells arranged in intersecting fascicles, consistent with a schwannoma. Antoni Type A and Type B patterns were seen on biopsy (Figure 2). Type A patterns consist of densely packed spindle cells with palisading nuclei, and Type B patterns feature paucicellular areas in a loose myxoid stroma.¹ Complete gross resection was achieved, but microscopic margins were positive at the PPF. At follow-up 5 months later, the patient had improvement in her symptoms with

no evidence of gross disease on endoscopy and magnetic resonance imaging. Table 1 shows a timeline of the case.

REVIEW OF THE LITERATURE

A literature review was performed using PubMed and Ovid MEDLINE databases from January 1, 1970, to September 20, 2015. Search terms included *vidian nerve, facial nerve, schwannoma, neuroma, neurinoma, neurilemoma, and head and neck*. Two reviewers (FM, JL) performed a thorough review of the titles, abstracts, and full texts where appropriate. The presenting symptoms, pathologic findings, and methods of management of the patients with vidian nerve schwannomas were analyzed and compared with those of our case report.

Our literature review yielded 7 case reports documenting 8 patients with vidian nerve schwannomas.^{3,7-12} The age of the patients, their presenting symptoms, and the methods of management are shown in Table 2. Headache was present in 3 of the 8 patients.^{3,8,10} Oculomotor palsy with a sphenoid sinus mucopyocele and cerebrospinal fluid leakage was seen in 1 patient.⁹ Another patient was asymptomatic.¹¹ Facial nerve palsy, palate pain, and hearing loss were seen in the others.^{3,7,8}

The most common treatment was a transnasal endoscopic resection (in 5 patients).^{3,8-10,12} One patient elected for observation,⁸ and another underwent radiation therapy.¹¹ A maxillary swing approach was used in the remaining patient.⁷

DISCUSSION

The vidian nerve (also known as the nerve of the pterygoid canal) is formed by the union of the deep and greater petrosal nerves, the latter being a branch of the facial nerve at the level of the geniculate ganglion. The greater petrosal nerve consists of presynaptic parasympathetic fibers that synapse with postganglionic neurons at the sphenopalatine ganglion to innervate the nasopalatine mucosa and lacrimal gland. Likewise, the deep petrosal nerve consists of postsynaptic sympathetic nerve fibers from the internal carotid artery plexus to innervate the nasopalatine mucosa and lacrimal gland. The vidian nerve travels in the vidian (pterygoid) canal, which runs from an area just anterior to the foramen lacerum in the middle cranial fossa to the pterygopalatine fossa, just posterolateral to the sphenopalatine foramen. This course traverses the medial pterygoid plate of the sphenoid bone, in the floor of the sphenoid sinus (Figure 3).^{3,13,14}

Given the vidian canal’s intimation with the floor of the middle cranial fossa, auditory apparatus, cavernous sinus, orbital apex, sphenoid sinus, and PPF, it is understandable how a vidian nerve schwannoma can cause the myriad symptoms reported in the literature.^{3,7-11} Our case demonstrates that a large vidian nerve schwannoma can present with predominantly ocular symptoms. This was

Date	Event
4/3/2015	Evaluated in the otolaryngology clinic for 3 mo of right-sided periorbital pressure and visual disturbances
4/7/2015	MRI demonstrated 4-cm mass in the right sphenoid region
4/15/2015	Transnasal endoscopic resection of mass performed with pathology showing schwannoma
9/7/2015	Clinic follow-up showed no evidence of recurrence, with significant improvement in symptoms
8/16/2017	MRI demonstrated no recurrence of disease

MRI = magnetic resonance imaging.

Source	Age, y/sex	Presentation	Treatment
Cheong et al, ³ 2006	13/female	Headache with unilateral facial nerve palsy	Transnasal endoscopic resection
Honda et al, ⁷ 2008	49/female	Unilateral hearing loss with serous otitis media	Maxillary swing with endoscopic assistance
Hackman et al, ⁸ 2011	Case 1: 49/male Case 2: 58/male	Case 1: Occipital headache Case 2: Unilateral palate pain and lip numbness	Case 1: Observation Case 2: Transnasal endoscopic resection
Wu et al, ⁹ 2012	78/female	Unilateral oculomotor palsy with CSF leakage	Transnasal endoscopic resection
Hong et al, ¹⁰ 2014	41/male	Occipital headache	Transnasal endoscopic resection
Yamasaki et al, ¹¹ 2015	49/female	Asymptomatic	Radiation therapy
Fortes et al, ¹² 2016	60/female	Unilateral facial hypoesthesia	Transnasal endoscopic resection

CSF = cerebrospinal fluid.

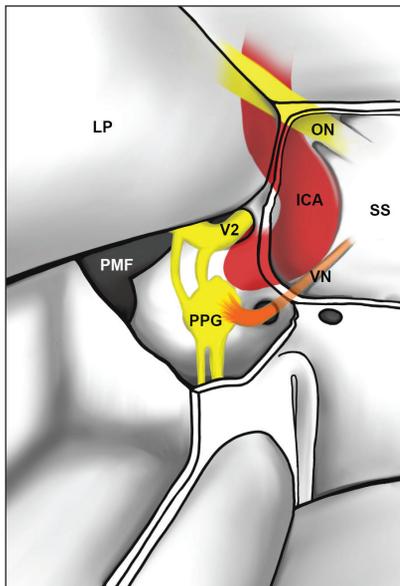


Figure 3. Anatomical schematic of the pterygopalatine fossa with anterior and medial walls removed, demonstrating the lamina papyracea (LP), vidian nerve (VN) in the vidian canal, maxillary nerve (V2) division of the trigeminal nerve exiting the foramen rotundum, pterygomaxillary fissure (PMF), pterygopalatine (sphenopalatine) ganglion (PPG), internal carotid artery (ICA), optic nerve (ON), and sphenoid sinus (SS).

caused by mass effect on the orbital apex and cavernous sinus. To our knowledge, this is the first case reported in the literature to present with visual field deficits, the second to present with oculomotor nerve palsy,⁹ and the only case in which ocular symptoms occurred without headache, hearing loss, facial weakness, or nasal obstruction.

On the basis of the previously reported cases, headache is the most common symptom at presentation. Although our patient did experience periorbital pressure, her predominant symptoms were vision loss and vision disturbance. Because the vidian nerve has parasympathetic and sympathetic functions in the nasal cavity, it is surprising that nasal cavity pain, dryness, or excessive secretions of the nasopalatine and lacrimal glands were not noted much in the previous reported cases. With our patient, nasal cavity and lacrimal gland symptoms were also lacking.

Surgery is the traditional modality of treatment of schwannomas when it can achieve gross resection with low

morbidity,¹¹ which is of particular concern with skull base lesions. An earlier case report used a maxillary swing approach for resection, which is a traditional open technique that was once commonplace along with lateral rhinotomy incisions, osteotomies, and infratemporal fossa approaches.⁷ Given the advancements in transnasal endoscopic techniques in approaching the skull base, this became the preferred method of resection in the later case reports where complete gross resection of the schwannoma was feasible with low morbidity.^{3,7-12} The most recent case report involved treatment with fractionated radiation therapy, which is reasonable given that radiotherapy has demonstrated durable tumor control in vestibular schwannomas.¹¹

CONCLUSION

Vidian nerve schwannomas are a rare subset of facial nerve schwannomas that can present with ocular manifestations caused by mass effect. Clinicians should be aware of the possibility of an anterior skull base neoplasm in patients with persistent ocular signs and symptoms. Endoscopic resection carries less morbidity than traditional open approaches and permits orbital decompression with preservation of ocular function. In cases in which microscopic margins are positive at the PPF, endoscopic resection also allows for subsequent endoscopic surveillance. Endoscopic resection is a safe and effective treatment modality for vidian nerve schwannomas. ♦

Disclosure Statement

The author(s) have no conflicts of interest to disclose.

Acknowledgment

We would like to acknowledge Karen Axelsson, MD, in the Department of Pathology at the Oakland Medical Center for assistance in interpretation and preparation of the pathology images.

Kathleen Loudon, ELS, of Loudon Health Communications provided editorial assistance.

How to Cite this Article

Masroor FA, Gilde J, Liang J. Vidian nerve schwannoma: A rare skull-base neoplasm presenting with ocular manifestations: A case report and literature review. *Perm J* 2018;22:18-021. DOI: <https://doi.org/10.7812/TPP/18-021>

References

- Pickett BP, Crawley BK. Neoplasms of the ear and lateral skull base. In: Johnson JT, Rosen CA, editors. *Bailey's Head & Neck Surgery: Otolaryngology*. 5th ed. Baltimore, MD: Lippincott Williams & Wilkins; 2014. p 2376-8.
- Malone JP, Lee WJ, Levin RJ. Clinical characteristics and treatment outcome for nonvestibular schwannomas of the head and neck. *Am J Otolaryngol* 2005 Mar-Apr;26(2):108-12. DOI: <https://doi.org/10.1016/j.amjoto.2004.08.011>.
- Cheong JH, Kim JM, Bak KH, Kim CH, Oh YH, Park DW. Bilateral vidian nerve schwannomas associated with facial palsy: Case report and review of the literature. *J Neurosurg* 2006 May;104(5):835-9. DOI: <https://doi.org/10.3171/jns.2006.104.5.835>.
- Russell DS, Rubinstein LJ. Tumors of the cranial, spinal, and peripheral nerves. In: *Pathology of tumours of the nervous system*. 5th ed. London, UK: Edward Arnold; 1989. p 533-89.
- Girardi G, Vanaki SS, Puranik RS. Schwannoma of parapharyngeal space. *J Maxillofac Oral Surg* 2010 Jun;9(2):182-5. DOI: <https://doi.org/10.1007/s12663-010-0059-y>.
- Samii M, Migliori MM, Tatagiba M, Babu R. Surgical treatment of trigeminal schwannomas. *J Neurosurg* 1995 May;82(5):711-8. DOI: <https://doi.org/10.3171/jns.1995.82.5.0711>.
- Honda K, Asato R, Tanaka S, Endo T, Nishimura K, Ito J. Vidian nerve schwannoma with middle cranial fossa extension resected via a maxillary swing approach. *Head Neck* 2008 Oct;30(10):1389-93. DOI: <https://doi.org/10.1002/hed.20793>.
- Hackman T, Rickert CG, Getz AE, Uppaluri R. Endoscopic surgical management of vidian nerve schwannoma. *Laryngoscope* 2011 Feb;121(2):241-4.
- Wu SW, Chen WL, Chen WL, Chen MK. Transnasal endoscopic resection of vidian nerve schwannoma accompanied by sphenoid mucopyocele and oculomotor palsy: A case report. *B-ENT* 2012;8(3):207-11.
- Hong HP, Yoon SW, Park MJ, Jung SC. A case of vidian nerve schwannoma: Resection by endoscopic sinus surgery. *Korean J Otorhinolaryngol Head Neck Surg* 2014;57(1):50-3. DOI: <https://doi.org/10.3342/kjori-hns.2014.57.1.50>.
- Yamasaki A, Sedaghat AR, Lin GC, Curry WT, Shih HA, Gray ST. A rare finding of schwannoma of the vidian canal: A case report. *J Neurol Surg Rep* 2015 Jul;76(1):48-51. DOI: <https://doi.org/10.1055/s-0034-1544112>.
- Fortes B, Beer-Furlan A, Balsalobre L, Vellutini E, Stamm A. Endoscopic endonasal access for the treatment of vidian nerve schwannoma: A case report. *Braz J Otorhinolaryngol* 2016 Jun 4. DOI: <https://doi.org/10.1016/j.bjori.2016.04.015>.
- Jacopo G, Micaela I, Italo C, Luigi C, Larocca LM, Gaetano P. Atypical sinonasal schwannomas: A difficult diagnostic challenge. *Auris Nasus Larynx* 2009 Aug;36(4):482-6. DOI: <https://doi.org/10.1016/j.anl.2008.11.010>.
- Kridel R, Sturm-O'Brien A. Nasal septum. In: Flint PW, Haughey BH, Lund VJ, et al, editors. *Cummings otolaryngology: Head and neck surgery*. 6th ed. Philadelphia, PA: Saunders Elsevier; 2015. p 476.

Keywords: cranial neuroma, cranial neurilemoma, endoscopic sinus surgery, free mucosal graft, nasoseptal flap, oculomotor palsy, pterygopalatine fossa, schwannoma, skull-base dehiscence, skull-base surgery, sphenoid sinus, Vidian nerve