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

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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 neurona, neurinoma, or neurilemoma) is a benign nerve sheath tumor composed of Schwann cells, which produce myelin that insulate peripheral nerves. The head and neck are frequent locations for schwannomas, accounting for 25% to 45% of their distribution. Schwannomas can arise from any cranial nerve, with the exception of the first and second cranial nerves, which lack Schwann cells. 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. Schwannomas are also the second most common tumor of the parapharyngeal space, behind tumors of salivary gland origin. The facial nerve is the third most commonly involved cranial nerve, but involvement of the vidian nerve branch is exceedingly rare. There are only 7 reported cases in the literature.

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 sphenoid 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).

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.
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. 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.

Table 1. Timeline of the case

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

MRI = magnetic resonance imaging.

Table 2. Literature review: Characteristics of previous case reports

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

CSF = cerebrospinal fluid.

The most common treatment was a transnasal endoscopic resection (in 5 patients). One patient elected for observation, and another underwent radiation therapy. A maxillary swing approach was used in the remaining patient.

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 parasympathetic 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).

Our case demonstrates that a large vidian nerve schwannoma can present with predominantly ocular symptoms. This was
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ocular symptoms occurred without headache; also lacking. The nasopalatine and lacrimal glands pain, dryness, or excessive secretions of and sympathetic functions in the nasal vidian nerve has parasympathetic function loss and vision disturbance. Because her predominant symptoms were vi-

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, which is a rare occurrence in the later cases where complete gross resection of the schwannoma was feasible with low morbidity. 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.

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.

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References


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