Neurofibroma of the internal carotid artery cavernous sympathetic plexus: illustrative case

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BACKGROUND Intracranial carotid sympathetic plexus (CSP) nerve sheath tumors have rarely been reported in the literature. This study describes the first reported case of a CSP neurofibroma and the first case of a CSP nerve sheath tumor treated via an endoscopic endonasal approach followed by adjuvant radiosurgery.

OBSERVATIONS A 53-year-old man presented with 3 days of headaches and diplopia and was found to have a complete left abducens nerve palsy. Computed tomography (CT) revealed a smoothly dilated left carotid canal, CT angiography revealed a superiorly displaced left internal carotid artery (ICA), and magnetic resonance imaging revealed a T2-hyperintense and avidly enhancing lesion in the left cavernous sinus encasing the ICA. The patient underwent subtotal resection via an endoscopic transsphenoidal trans cavernous approach followed by Gamma Knife radiosurgery.

LESSONS Nerve sheath tumors arising from the CSP are extremely rare but need to be considered when assessing unusual cavernous sinus lesions. The clinical presentation is dependent on the anatomical location of the tumor and its relationship to the ICA. The optimal treatment paradigm is unknown.

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KEYWORDS cavernous sinus; sympathetic plexus; neurofibroma; carotid canal; schwannoma

Although neurofibromas are the most common benign nerve sheath tumors, they rarely present as an intracranial tumor. They arise from the nerve sheath and may occur along any peripheral nerve. Unlike schwannomas, which tend to be encapsulated and displace associated nerve fibers, neurofibromas are classically embedded in the axons and endoneurium. They are characterized by a germline mutation in the NF1 gene in up to 10% of patients.

The internal carotid artery (ICA) sympathetic plexus contains postganglionic sympathetic fibers arising from the superior cervical ganglion. These fibers accompany the ICA throughout its petrous and cavernous segments. Nerve sheath tumors of the intracranial carotid sympathetic plexus (CSP) are extremely rare, with only eight CSP schwannomas reported in the literature.1–8

We describe the interesting case of a neurofibroma arising from the cavernous CSP treated via an endoscopic endonasal approach. This is the first documented case of a CSP neurofibroma as well as a CSP lesion treated via an endoscopic endonasal approach.

Illustrative Case

A 53-year-old man presented with a 3-day history of headache and diplopia. He had experienced recurring headaches during the preceding year. He had no significant medical history. Clinical examination demonstrated a complete left abducens nerve palsy. He had no further cranial neuropathies and no peripheral stigmata of neurocutaneous disorders.

Imaging revealed a circumferential, heterogeneously enhancing lesion of the left ICA (Fig. 1). The lesion was located between the horizontal segment of the petrous ICA and the anterior genu of the cavernous segment and was eccentrically located inferomedial to the ICA. There was dilation and remodeling of the carotid canal and foramen lacerum, with the lesion abutting the lateral sphenoid sinus.

ABBREVIATIONS CSP = carotid sympathetic plexus; CT = computed tomography; ICA = internal carotid artery; MR = magnetic resonance.

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wall. There was no calcification of the lesion or narrowing of the ICA (Fig. 2).

The patient underwent debulking of the lesion via an endoscopic transsphenoidal transcavernous approach. A modified expanded endonasal approach was performed with a right-sided ethmoidectomy, including resection of the right middle turbinate and wide bilateral sphenoidotomies in addition to a posterior septectomy window. Widening of the right nasal passageway allowed for a direct path to the cavernous sinus and lateral sphenoid wall. The anterior wall of the sella was removed on the left side, and this was extended to include the floor of the sella. The bony exposure was then extended in a medial to lateral fashion, uncovering the anterior genu of the ICA and the medial cavernous sinus wall. Stereotaxis and Doppler ultrasonography were used to identify the course of the ICA. After the medial wall of the cavernous sinus was widely decompressed, a dural opening was made over the tumor. The tumor was found to be firm and fibrous. Significant resection was not possible given the nature of the tumor and the intimate relationship with the ICA (Fig. 3).

Histopathology demonstrated a spindle cell lesion with a myxoid background with fascicles of spindle cells and wavy nuclei. Immunophenotype was positive for S100 and neurofilament and negative for EMA, SMA, CD34, PR, and p53. The tumor was diagnosed as a neurofibroma (Fig. 4).

The patient had an uncomplicated postoperative course, with the persistence of abducens nerve palsy at the last follow-up. He underwent Gamma Knife radiosurgery 6 months following resective surgery.

Discussion

We report the first case of a CSP neurofibroma documented in the literature. The patient was treated with subtotal resection via an endoscopic transsphenoidal transcavernous approach followed by stereotactic radiosurgery.

Observations

Anatomical Considerations

The ICA sympathetic plexus arises from the superior cervical ganglion before entering the carotid canal, in association with the ICA. These postganglionic sympathetic fibers derive from the ganglion via a medial and lateral branch, contributing to the cavernous plexus and the deep petrosal nerve, respectively.9

The sympathetic plexus forms two areas of increased density along its course: the first related to the distal petrous segment of the ICA and the second to the cavernous segment of the ICA. More detailed anatomical dissections have shown anterosuperior and posteroinferior bundles along the petrous carotid. The deep petrosal branch originates from the anterosuperior bundle.10 In the cavernous segment, multiple smaller bundles form10 before providing small branches to the abducens nerve and the ophthalmic nerve.11,12

CSP schwannomas have been rarely described, with eight cases reported in the literature (Table 1). The tumor location dictates the clinical presentation, and distinct entities have been described. Takase et al.7 proposed a classification scheme for CSP schwannomas based

FIG. 1. Axial T2-weighted (A) and T1-weighted with contrast (B) and coronal T2-weighted (C) and T1-weighted with contrast (D) images demonstrate a T2-hyperintense lesion with avid homogeneous contrast enhancement. The tumor is intimately related to the horizontal segment of the cavernous ICA extending to the anterior genu.

FIG. 2. Coronal CT bone window imaging (A) demonstrating remodeling of the carotid canal (dashed circles). CT angiography (B) shows superior displacement of the horizontal segment of the cavernous ICA.

FIG. 3. Intraoperative endoscopic images. The green dashed line indicates the course of the cavernous ICA. CR = clival recess; OCR = opticocarotid recess; S = sella; T = tumor.
FIG. 4. Histological findings consistent with neurofibroma. A: Hematoxylin and eosin staining shows fascicles of spindle cells in a myxoid background. B: S100 was diffusely positive. C: Neurofilament was positive, highlighting axons. D: Ki-67 showing a low proliferative index. Original magnification ×100 (A–D).

on the anatomical location of the tumor as either type A, arising from the cavernous CSP, or type B, arising from the petrous CSP within the carotid canal of the temporal bone. This anatomical classification scheme correlates with clinical presentation and may help dictate the surgical approach. Although originally described for schwannoma, it is applicable in our case due to similarities in tumor behavior and anatomical origin.

Type A tumors are intracavernous in location and arise from the cavernous plexus. Three cases, including our case, have been described, all presenting with abducens nerve palsy. Within the literature, one patient underwent resection via an extradural approach to the lateral cavernous sinus and one was managed without surgery. We present the first type A tumor to be approached via an endoscopic endonasal approach.

Type B tumors arise from the internal carotid plexus and are located at the petrous apex. These lesions have been described as intraosseous. Six cases have been reported in the literature and present with hearing disturbance and tinnitus secondary to damage to the eustachian tube, inner ear, or tympanic cavity. The extracavernous location of these lesions facilitates surgical access. In the literature, four type B tumors have been operated on via an extradural middle fossa approach with gross-total resection.

Radiological Considerations

The differential diagnosis for a cavernous sinus lesion is broad, with multiple diagnoses having nonspecific or overlapping imaging features. Our patient’s computed tomography (CT) scan demonstrated dilation and remodeling of the left carotid canal without frank erosion or destruction of surrounding bone (Fig. 2). CT angiography and magnetic resonance (MR) T2-weighted flow voids revealed a superiorly displaced cavernous segment of the ICA. MR imaging demonstrated a slightly hyperintense, smooth lesion on T2-weighted sequences with homogeneous, avid, postcontrast enhancement; the lesion was expanding the cavernous sinus and abutted the sphenoid sinus while displacing the ICA superiorly and following the ICA from the cavernous segment to the petrous segment in the carotid canal (Figs. 1 and 2).

When reviewing the radiological features of CSP schwannomas, Hamilton et al.3 highlighted the significance of a smoothly expanded carotid canal as compared with other pathologies that occur in the region. This bone remodeling is attributed to the slow growth of the tumor and occurs along the axis of the ICA.

Endoscopic Endonasal Considerations

Advancement in endoscopic techniques, such as angled endoscopes and image guidance, has allowed access to a broader region of the central skull base, including the cavernous sinus. Fernandez-Miranda et al.13 defined the cavernous sinus compartments when accessing them via an endonasal approach. This tumor was in the inferior compartment, located inferior to both the anterior genu and the horizontal segment of the cavernous ICA. The compartment is bounded anteriorly by the anterior wall of the cavernous sinus and in anatomical dissections was found to contain the sympathetic plexus.

The group proposed a transpterygoid supravidian approach to this region,13 with exposure of the anterior and lateral wall of the cavernous sinus. This was not required in our case given the enlargement of the inferior compartment by the tumor and the

TABLE 1. Summary of previous case reports.

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Pathology</th>
<th>Age (yrs)/Sex</th>
<th>Diplopia</th>
<th>Hearing</th>
<th>ICA Displacement</th>
<th>Approach</th>
<th>Classification Type</th>
<th>EOR</th>
</tr>
</thead>
<tbody>
<tr>
<td>Solodnik et al., 19866</td>
<td>S</td>
<td>59/M</td>
<td>—</td>
<td>Tinnitus</td>
<td>Normal</td>
<td>Suboccipital</td>
<td>B</td>
<td>ND</td>
</tr>
<tr>
<td>Horn et al., 19954</td>
<td>S</td>
<td>46/F</td>
<td>—</td>
<td>Fullness</td>
<td>Normal</td>
<td>Postauricular</td>
<td>B</td>
<td>GTR</td>
</tr>
<tr>
<td>Türe et al., 20033</td>
<td>S</td>
<td>29/M</td>
<td>CN6</td>
<td>—</td>
<td>Hearing loss</td>
<td>Lat</td>
<td>Middle fossa</td>
<td>B</td>
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<tr>
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<td>CN6</td>
<td>Hearing loss</td>
<td>Lat</td>
<td>—</td>
<td>B</td>
<td>—</td>
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<tr>
<td>Goiney et al., 20111</td>
<td>S</td>
<td>48/F</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>NO</td>
<td>A</td>
<td>NO</td>
</tr>
<tr>
<td>Takase et al., 20177</td>
<td>S</td>
<td>54/M</td>
<td>CN6</td>
<td>Hearing loss</td>
<td>—</td>
<td>Superolat</td>
<td>Middle fossa</td>
<td>B</td>
</tr>
<tr>
<td>Present case</td>
<td>N</td>
<td>53/M</td>
<td>CN6</td>
<td>—</td>
<td>—</td>
<td>Superolat</td>
<td>Endoscopic</td>
<td>A</td>
</tr>
</tbody>
</table>

CN = cranial nerve; EOR = extent of resection; GTR = gross-total resection; N = neurofibroma; NO = no operation; S = schwannoma; STR = subtotal resection; Superolat = superolateral.

Adopted and updated from Takase et al.7
superolateral displacement of the cavernous ICA. Instead, a medial to lateral decompression approach to the medial cavernous sinus wall was performed. This approach has previously been described and has been shown to be safe. Although exposure is limited laterally with this approach, it was of no consequence in our case. However, this is an important consideration if gross-total resection is intended.

Lessons
CSP nerve sheath tumors are extremely rare and may arise from either of the distinct sympathetic plexus bundles of the intracranial ICA. The location of the tumor and its relationship to the structures at the skull base dictate the clinical presentation and guide the operative approach. We present the first case of a CSP neurofibroma and the first lesion treated with an endoscopic endonasal approach. Optimal management of these lesions is not yet known because of their rarity.

References

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Conception and design: Blackburn, Trimble, Dawes, Zeineddine.
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Analysis and interpretation of data: Blackburn, Trimble, Zeineddine, Yao. Drafting of the article: Blackburn, Trimble, Dawes, Zeineddine, Yao. Critically revising the article: Blackburn, Trimble, Dawes, Zeineddine, Guttenberg. Reviewed submitted version of the manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Blackburn. Study supervision: Blackburn.

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