Tumors of the Cervical Sympathetic Chain – Diagnosis and Management

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Objectives: Tumors originating from the cervical sympathetic chain are uncommon but important entities in the differential diagnosis of prelaminar paranganglioma (paraganglioma) space masses. We sought to evaluate the presentation of these tumors and the outcomes of surgical treatment.

Study Design: Retrospective chart review

Methods: We report our experience with 24 patients from 1994 to 2011. Clinic notes, operative and pathology reports, and radiologic images and reports were used to create the study database.

Results: The most common presenting symptoms were dysphagia (29%, n=7), neck mass (n=7, 29%), and throat fullness (n=4, 17%). Two patients (8%) presented with Horner’s syndrome. Although radiologic images showed classic lateral displacement of the carotid arteries in 10 (42%), in 9 (38%) patients the radiologic findings demonstrated splaying of the carotid arteries similar to carotid body tumor and in 5 (20%) the findings were indeterminate. Three patients were observed, two due to small size and patient preference and one because of multiple bilateral cranial nerve involvement. Twenty-one patients underwent surgical removal, with pathology revealing 10 parangangiomas, 10 schwannomas, and 1 neurofibroma. Three patients (14%) had cranial nerve weaknesses (two vagal and one spinal accessory). Although most patients had some degree of Horner’s syndrome postoperatively, this was symptomatic in 12 (57%). With a mean follow-up of 20 months there have been no recurrences.

Conclusions: This represents the largest original series of tumors of the sympathetic chain to date. Although anterolateral displacement of the carotids on imaging is suggestive of a sympathetic tumor, absence of these findings does not rule out this entity. Cervical sympathetic tumors can be safely managed with operative intervention with less than a 15% incidence of cranial nerve weakness.

Table I. Clinical Characteristics for 24 Sympathetic Chain Tumors

<table>
<thead>
<tr>
<th>Tumor Type</th>
<th>Sex</th>
<th>GF</th>
<th>Age [range]</th>
<th>Other Lesions</th>
<th>Familial</th>
</tr>
</thead>
<tbody>
<tr>
<td>Parangangioma</td>
<td>M 8</td>
<td>48</td>
<td>25-86</td>
<td>Synchronous</td>
<td>3 (12.5%)</td>
</tr>
<tr>
<td>Schwannoma</td>
<td>F 16</td>
<td>48</td>
<td>25-86</td>
<td>Synchronous</td>
<td>3 (12.5%)</td>
</tr>
<tr>
<td>Neurofibroma</td>
<td>4</td>
<td>21</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Not-operated</td>
<td>3</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Positive Lymph Nodes</td>
<td>0 (0%)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Operative Steps in Removal of Sympathetic Chain Tumors

1. Both high cervical and facelift-style incisions offer adequate exposure and are planned based on presence of tortuous neck creases and body habitus.
2. A level II selective neck dissection both samples lymph nodes in potential malignant paragangliomas and exposes important landmarks (SCM, digastic, X, XI, jugular vein and carotid)
3. Dissection is carried deep to the posterior belly of the digastric, allowing removal of the styloid process and attached musculature.
4. All nerves and vessels are identified (see Figure 1) and the sympathetic chain is isolated distal and proximal to the tumor.
5. In cases of paragangliomas and neurofibromas, the tumor is removed with the nerve trunk. In cases of schwannomas, attempts may be made to enucleate the tumor and preserve the sympathetic chain.
6. The neck incision is closed in layers over a suction drain.

Figure 1. Anatomy of the Carotid Space. The sympathetic chain is located postero medial to the great vessels and cranial nerves IX, X, XI, and XII. All of these structures are potentially at risk with growth or removal of sympathetic tumors.

Figure 2. Intraoperative Photo of Sympathetic Chain Schwannoma. The vagus nerve (V) and jugular vein (J) can clearly be seen separate from and passing lateral to the tumor. The internal carotid artery (IC) which was overlying the tumor in a position adjacent to the vagus, was mobilized forward for the purposes of this picture and removal of the mass. Upper right inset shows a low-power operative image.

Figure 3. Axial Imaging of a Sympathetic Schwannoma. Contrast-enhanced CT (left panel) demonstrates a minimally-enhancing lesion that splay the bifurcation but appears located medial to the vessels. Gadolinium T1-weighted MRI (right panel) shows the lesion to be heterogeneously enhancing without flow voids.

Figure 4. Coronal Imaging of Sympathetic Schwannoma. Same lesion as in Figure 3. Contrast-enhanced CT (left panel) shows a lesion splaying the left carotid arteries at the bifurcation. MR angiography (right panel) demonstrates splaying of the carotids but without the presence of a hypervascular lesion (“empty lye”).

Table II. Imaging Characteristics Suggestive of a Lesion Other than a Carotid Body Tumor

Both sympathetic chain tumors and vagal tumors can mimic carotid body tumors on imaging studies. The following findings make a carotid body tumor unlikely. Examples are in Figure 5 below.

A. Lack of internal and external artery separation

Carotid body tumors will splay the internal and external carotid and may do so in antero-posterior or less commonly mediolateral directions. Non-carotid body tumors may splay the carotid arteries but typically displace them in the same direction.

B. Location away from the bifurcation

Tumors may splay the carotid arteries, but if they are not located immediately at the junction of the internal and external carotid arteries, suspicion should be raised for a lesion other than a carotid body tumor. Careful evaluation may reveal the lesion to be more superiorly located than the carotid body.

C. Absence of flow-voids on enhanced MRI

Paragangliomas are high-flow lesions that tend to demonstrate flow voids on MRI. Schwannomas also enhance intensely, but tend not to demonstrate flow voids. Whereas a paraganglioma may arise from the carotid body or from other neural structures such as the sympathetic chain and vagus nerve, schwannomas from the carotid body have not been reported.

D. Presence of an ipsilateral carotid body tumor

A second lesion on the same side as a carotid body tumor necessarily indicates a tumor on a nearby structure.

The preoperative diagnosis of a sympathetic chain or vagal tumor ultimately relies on considering these entities in the differential diagnosis of a presumed carotid body tumor.

Figure 5. Radiographic findings in Sympathetic Chain Tumors. A) schwannoma displaces the carotid arteries (gray arrows) laterally; B) paraganglioma located superior to the carotid bifurcation (white arrowhead); C) enhancing lesion on T1-weighted MRI not demonstrating flow voids (a carotid paraganglioma with flow voids is shown in the inset panel); D) small schwannoma (white arrow) seen medial to a larger carotid body tumor.

Works Consulted


