Cervical Sympathetic Chain Schwannoma

Chang-Chun Lin, Chen-Chi Wang,* Shi-An Liu, Ching-Ping Wang, Wen-Hsien Chen

Schwannomas are benign, slow-growing tumors that arise from Schwann cells of the nerve sheath. Those originating from the sympathetic cervical chain are rare. Herein, we present three cases of cervical sympathetic chain schwannoma (CSCS) and describe our experience with the clinical presentation, surgical management, and outcomes of patients with this pathology. All three patients were found to have a pulsatile, non-tender mass located at the level of the angle of the mandible without cervical nerve function deficit. On imaging study, the location of the mass between the prevertebral muscle and carotid artery with outward displacement of the carotid sheath is a typical characteristic of CSCS. Operative excision and sacrifice of a portion of the sympathetic chain remains the treatment of choice. Postoperative Horner’s syndrome is predictable and vagus nerve dysfunction or first bite syndrome is also possible. [J Formos Med Assoc 2007;106(11):956–960]

Key Words: carotid body tumor, cervical sympathetic chain, neck mass, neurilemmoma, parapharyngeal space, schwannoma

Schwannomas in the head and neck region may originate from any of the peripheral, cranial, or autonomic nerves, including the optic and olfactory nerves.1 This tumor arises from the Schwann cells, which are precursor cells of the nerve sheath. They can be categorized according to their nerve origin and location within the head and neck. Parapharyngeal space neoplasms are uncommon lesions in the adult population. Approximately 20–27% of parapharyngeal masses are benign nerve sheath tumors.2 Schwannomas that originate from the cervical sympathetic chain are a rare subgroup of these tumors. Herein, we report three cases of cervical sympathetic chain schwannoma (CSCS) and present the clinical characteristics, management, and treatment outcomes along with a review of the literature.

Case Reports

Case 1

A 38-year-old man presented with a slowly enlarging, painless mass in the right upper neck, which he had had for 12 months, without any other symptoms such as tinnitus, hearing loss, dysphagia, dysphonia, dyspnea, pain or weight loss. Physical examination revealed a firm, non-tender ovoid mass measuring about 4 × 4 × 5 cm situated just below the right submandibular gland. Complete neurologic assessment revealed all cranial nerve function to be normal and no Horner’s syndrome was noted. Upper aerodigestive tract examination by flexible laryngoscopy was also normal. Fine needle aspiration showed no malignant cells. Computed tomography (CT) revealed...
one heterogeneous enhanced mass lesion about 45 mm in diameter at the carotid space of the right upper neck just posterior to the right submandibular gland and medial to the right carotid bifurcation (Figure 1).

Case 2
A 33-year-old woman had a history of neurofibromatosis type II with neurofibroma over the left axilla, which was excised when she was 10 years old. Typical café-au-lait macula was noted over the neck and trunk. She had had a mass lesion over the left upper neck for 5 years. This firm mass lesion had become larger without tenderness in the half year before the current admission. CT showed a heterogeneous enhancing mass lesion about $4 \times 5 \times 7$ cm in size over the left neck in the parapharyngeal space. Magnetic resonance angiography (MRA) revealed a mass lesion about $3.7 \times 1.8 \times 2.5$ cm with high signal intensity on T2WI and well-contrasted enhancement over the right carotid bifurcation region of the parapharyngeal space at the oropharynx level, and no definite abnormal vascular stain was noted from MRA. Balloon test occlusion suggested that the patient would tolerate ligation of the internal carotid artery (ICA) with a low probability of significant neurologic sequelae.

All three patients received surgical excision of the tumor via cervical approach. The tumor of Case 1 was well-encapsulated and located between the carotid sheath and prevertebral muscle (Figure 2). It caused outward displacement of the internal jugular vein, carotid artery and vagus nerve. The vagus and hypoglossal nerves were identified and preserved. The mass was resected (Figure 3) and we
repaired the nerve with end-to-end anastomosis and without using graft.

Microscopic examination of the lesion revealed both Antoni type A and type B histology (Figure 4). After the operation, Horner’s syndrome with mild ptosis was noted in all three patients and first bite syndrome was noted in Case 1. Case 3 had mild hoarseness due to right vocal fold paralysis. There was no other neurologic deficit.

**Discussion**

Schwannomas are typically benign, slow-growing, solitary tumors arising from Schwann cells of the nerve sheath. The specific details of their genesis are not known. Since the initial report, the tumor has been given a number of names, including neurilemmoma, solitary nerve sheath tumor, perineural fibroblast tumor and, more recently, schwannoma.3–6 Schwannomas of the sympathetic chain are quite rare, with approximately 55 cases of CSCS reported in the literature.7 CSCS are reported to occur in patients between the ages of 20 and 50 years. Males and females are equally affected, and approximately 25–45% of these neoplasms are found in the head and neck region.8 Malone et al found that the parapharyngeal space (31%) was the most common nonvestibular head and neck location.9

The majority of schwannomas are asymptomatic at the time of presentation. Patients usually experience an indistinct neck mass accompanied by some vague symptoms such as lump sensation. The incidence of Horner’s syndrome before excision has only been reported previously in six cases of cervical schwannoma.10,11 Schwannomas, by their nature, rarely impair nerve function. Because the cervical sympathetic chain runs in a relatively loose fascial compartment, nerve injury due to compression, such as that seen with acoustic neuromas or facial nerve schwannomas, is extremely rare. Even in infiltrative neurofibromas, dysfunction of the nerve of origin is rare.12

Most tumors of the parapharyngeal space are metastatic disease or direct extension from adjacent spaces. Primary parapharyngeal tumor is uncommon. In the literature, 80% of parapharyngeal space tumors are benign and 20% are malignant. The location of the tumor can be helpful for differential diagnosis. Tumors in the pre-styloid compartment are most likely to be salivary gland tumor, lipoma, or rare neurogenic tumors. And all the structures in the post-styloid compartment are potential sources for post-styloid tumors.13 Thus, masses that could arise in the post-styloid compartment include carotid artery lesions, paragangliomas arising from the vagus nerve or the carotid body, neurogenic tumors of cranial nerves IX to XII, or sympathetic chain. In our cases, the tumors were all located in the post-styloid compartment.

As patients do not usually present with Horner’s syndrome, imaging studies play a central role in the diagnosis of head and neck schwannomas. Regardless of the nerve of origin, schwannomas in general are hypodense in comparison to muscle on CT without contrast.14 With contrast, these lesions show at least some degree of enhancement, often at the periphery.15 Paragangliomas, on the other hand, are classically isodense when compared to muscle on pre-contrast CT, with more reliable homogeneous enhancement post-contrast. MRI of schwannomas reveals relatively low signal intensity on T1-weighted images and high signal intensity on T2-weighted images.16

**Figure 4.** A loose, hypocellular, myxoid pattern characterizes Antoni type B histology (open arrow). The Antoni A (black arrow) regions comprise tightly packed, spindle-shaped cells. (Hematoxylin & eosin, 100×.)
With significant enhancement, a vascular lesion must be ruled out. The angiographic appearance of schwannoma has been characterized by Abramowitz et al, who described a vascular pattern consisting of scattered “puddling” of contrast medium throughout the lesion, with no evidence of arteriovenous shunting. The relationship of the mass to the surrounding vascular structures is important in ruling out a carotid body tumor. Classically, a carotid body tumor will splay the internal and external carotid arteries, producing a “lyre” sign. This is rarely associated with a schwannoma.

In our cases, the workup included CT, MRI and MRA. CSCS can mildly splay the carotid bifurcation but does not separate the internal jugular vein and the ICA. Vagus nerve schwannomas (VNS) can separate the internal jugular vein and the ICA, but they cannot widen the carotid bifurcation. Previous studies have shown that anterior displacement of the common carotid artery and of the ICA is a characteristic finding of both VNS and CSCS. This is because both schwannomas grow posteriorly to these blood vessels. In VNS, the schwannoma grows between the common carotid artery or the ICA and the jugular vein, resulting in an increase in the distance between the artery and the vein (separation). In CSCS, the schwannoma grows posteriorly to the common carotid artery, ICA, and jugular vein, and no separation can be observed between the jugular vein and the common carotid artery or ICA (Figure 1). However, we found that the jugular vein and the ICA are displaced laterally. The internal and external carotid arteries are mildly splayed and the external carotid artery is displaced anteriorly, which presented in all of our three cases. This result is consistent with the report of Wang et al, which reported one case and reviewed seven cases of CSCS in the literature. Wang et al concluded that splaying of the carotid bifurcation with hypervascularity suggests a carotid body tumor, where as in cases without hypervascularity, a CSCS is another possibility. Vagus nerve schwannoma can separate the internal jugular vein and ICA, but seldom widen the carotid bifurcation. Additionally, we contribute three more cases of evidence for this radiologic characteristic of CSCS.

Several surgical approaches have been recommended for excision of tumors in the parapharyngeal space, including transcervical (with or without mandibulotomy), transparotid, infratemporal, and transoral approaches separately or in combination. It has been our experience that the transcervical approach provides adequate exposure for excision of the parapharyngeal space CSCS. All vascular structures and neural structures, including cranial nerves IX to XII and the superior laryngeal nerve, should be identified and preserved (Figure 2). By careful identification and preservation of these neurovascular structures, morbidity in the postoperative period may be avoided. Once these structures have been identified, the tumor can be excised from the sympathetic chain. In the present cases, we had to transect the nerve to remove the tumor. End-to-end anastomosis was performed in all three cases. However, Horner’s syndrome with ptosis, miosis, and anhidrosis were still noted even though the patients seldom complained of symptoms. Transient vagus nerve paralysis with hoarseness was noted in Case 3, possibly due to traction of the vagus nerve during operation or postoperative ischemic neuropathy.

Case 1 experienced pain in the parotid area while eating food at the first bite after surgery, which is the typical symptom of first bite syndrome. First bite syndrome is a complication of parapharyngeal space surgery that presents as pain in the parotid and jaw, often with radiation to the ear, which lasts a few seconds and occurs with the first bite of a meal. The symptoms are always worst with the first bite and improve to resolution with subsequent bites. The symptoms may recur at the start of the next meal. It is thought that first bite syndrome may be caused by loss of sympathetic input to the parotid gland after severing of the cervical sympathetic chain. This results in a denervation hypersensitivity of the sympathetic receptors that control myoepithelial cells in the parotid gland. A supramaximal response by the myoepithelial cells is elicited on
cross-stimulation by the release of parasympathetic neurotransmitters induced by chewing and biting.\textsuperscript{21} We found that the symptoms in Case 1 resolved slowly over time after 6 months of follow up.

In summary, schwannomas of the cervical sympathetic chain are rare neoplasms that most often present as asymptomatic unilateral neck masses without Horner’s syndrome. Imaging studies play a central role in the diagnosis of CSCS. Parapharyngeal tumor without hypervascularity and mild splaying bifurcation of the carotid artery that does not separate the internal jugular vein and the ICA should be considered as CSCS. In our patients, the tumors were intimately involved with the nerve, requiring partial resection of the sympathetic chain. Postoperative Horner’s syndrome is common, but does not appear to cause problems and usually at least partially resolves over time. Other neurologic deficits, such as vagus nerve paralysis and first bite syndrome, may occur, and this possibility should be carefully discussed with the patient preoperatively.

References