Schwannomas are tumors arising from the myelin-producing Schwann cells of the peripheral sensory nervous system. Of those occurring in the head and neck region, most are intracranial. Involvement of cranial nerves containing motor fibers alone is uncharacteristic. Schwannomas of the 12th cranial nerve are rare, with most originating intracranially, although some may extend extracranially with prolonged growth. Hypoglossal nerve schwannomas may rarely arise exclusively along the extracranial portion of the nerve, and to our knowledge eight such cases have been reported [1-3]. Schwannomas involving the cervical sympathetic chain are equally rare, with only a handful of cases reported in the literature [4].

We describe the unique case of a patient with hypoglossal nerve and cervical sympathetic chain schwannomas presenting concomitantly as submandibular and carotid space masses, respectively. Because of the rarity of schwannomas in these particular locations, this diagnosis was not considered in the interpretation of the preoperative imaging studies.

Case Report

A 53-year-old woman, who had a medical history significant for bilateral hearing loss unassociated with acoustic neuromas, presented with a 4-week history of a palpable right-sided neck mass. She had no pain or sensory loss, dysphagia, dysphonia, or constitutional symptoms. Physical examination revealed a 3-cm firm, nontender, mobile mass in the mid anterior right side of the neck, as well as a second mass in the right submandibular space. No associated tongue asymmetry, vocal cord paralysis, or oral cavity lesion was found.

Review of a CT scan obtained at another medical facility showed two discrete masses, one appearing to arise from the submandibular gland, the other a cystic mass in the carotid space. Fine-needle aspiration of the carotid space mass suggested a branchial cleft cyst; however, an epithelial lining that would confirm this diagnosis was not identified. To better characterize the masses, MR imaging was performed and showed a 1.8-cm mass contiguous with the anteromedial right submandibular gland below the floor of the mouth, as well as a 3-cm complex mass posterior to the gland in the carotid space (Fig. 1). Both masses displayed areas of necrosis or cyst formation. These lesions were interpreted as a submandibular gland tumor and a branchial cleft cyst or necrotic lymph node, respectively. CT-guided fine-needle aspiration of the submandibular mass did not yield adequate tissue for diagnosis.

Because malignancy could not be excluded, the patient underwent surgical excision. Intraoperatively, the mass that radiologically appeared to arise from the submandibular gland represented a hypoglossal nerve lesion (the submandibular gland was normal), and the carotid space mass represented a schwannoma of the cervical sympathetic chain. The posterior division of the hypoglossal nerve was dissected free from the tumor. The anterior divisions of the hypoglossal nerve and of the cervical sympathetic chain were sacrificed because they were intimately involved by tumor. A greater auricular nerve graft was used to repair the anterior branch of the hypoglossal nerve. Pathologic examination revealed well-encapsulated solid and cystic masses with small nerve fascicles entering their periphery. Microscopically, both masses had characteristic Antoni A and B type patterns consistent with schwannomas. The histologic diagnosis was confirmed by immunohistochemistry, which showed neoplastic cells diffusely positive for S-100 protein.

After surgery, the patient developed mild right-sided Horner's syndrome and hypoglossal nerve palsy. On clinical follow-up 8 days later, the patient had moderate dysar-
thria associated with paralysis of the 12th nerve. The patient’s workup was negative for neurofibromatosis type II.

Discussion
Schwannomas are benign, slow-growing, encapsulated tumors of neural crest origin. In the head and neck, schwannomas usually arise from the sensory divisions of cranial nerves, most commonly the vestibular nerve.
Synchronous Schwannomas

Less commonly the vagus, trigeminal, and facial nerves are affected. Involvement of motor nerves or multiple nerves is uncommon and when present may indicate the diagnosis of neurofibromatosis type II.

Hypoglossal nerve schwannomas have been described along various segments of the nerve from its origin in the medulla to its termination in the submandibular space. For diagnostic purposes the hypoglossal nerve may be divided into five segments: the medullary, cisternal, skull base, pharyngeal, and sublingual segments [5]. To our knowledge, only 49 cases have been reported; of these, 28 were intracranial, 13 transcranial (arising intracranially), and eight extracranial in origin (four were located in the parapharyngeal space and four in the submandibular space) [1-3].

Schwannomas of the hypoglossal nerve affect women approximately 2.5 times more often than men, often presenting in the fourth and fifth decades of life. Subcapsular and generalized headaches account for 44% and 33% of initial symptoms, respectively [3]. Although hypoglossal nerve palsy is an uncommon initial symptom, it is present in 94% of patients at clinical presentation [1]. Furthermore, depending on the affected nerve segment, clinical presentation may also include glossopharyngeal, vagus, or spinal accessory nerve palsies (50%), cerebellar dysfunction (48%), and motor (41%) or sensory (37%) long tract signs [1]. Cranial nerve palsies result from the stretching of axons around the tumor mass. Because neighboring nerves may be affected by direct mass effect, nerve dysfunction does not necessarily indicate the nerve of tumor origin.

Schwannomas of the cervical sympathetic chain are equally rare, with only 14 reported cases that we are aware of [6]. These lesions present as parapharyngeal or carotid space masses. Large lesions may be associated with dysphagia, dysarthria, lower cranial nerve palsies, and trismus [4].

As in the case we present, schwannomas of the head and neck in unusual locations are often mistaken for other disease processes. A review of extracranial schwannomas involving the cranial and cervical nerves revealed that only 29% of cases were correctly diagnosed preoperatively [7]. CT and MR imaging are critical in the evaluation of head and neck masses because they allow for accurate anatomic detail and spatial localization. Schwannomas are well-defined masses that enhance avidly and have variable signal intensity on T2-weighted images depending on the presence of tissue necrosis and the degree of tumor cellularity classified as Antoni A or Antoni B type tissue. Histologically, Antoni type A is characterized by densely populated palisades of nuclei that may be hypointense on T2-weighted imaging. Antoni type B corresponds to a loose meshwork with an indistinct cellular pattern that appears hyperintense on T2-weighted imaging because of its increased water content.

Secondary imaging findings of hypoglossal nerve palsy include ipsilateral tongue-base enlargement and posterior retraction, as well as hyperintensity on T2- and T1-weighted images reflecting derervation with fatty infiltration and atrophy [8]. Lesions arising from the vagus nerve and cervical sympathetic chain may be distinguished on cross-sectional imaging according to their relationship with the neck vasculature. Although vagus nerve masses characteristically grow between and therefore splay the carotid artery and jugular vein, cervical sympathetic chain masses grow posterior to both vessels and do not produce this separation. However, both lesions may displace the carotid artery anteriorly.

In the evaluation of a carotid space mass, branchial cleft cysts, paragangliomas, and inflammatory or neoplastic lymph nodes should be considered. Branchial cleft cysts are well-demarcated, unicellular lesions. Necrotic lymph nodes may be distinguished from cysts by their thick walls, which represent residual lymphoid stroma. Paragangliomas are hypervascular tumors with a characteristic salt-and-pepper appearance on T2-weighted images, representing areas of solid tumor intermingled with vascular flow voids.

Masses in the submandibular space commonly represent infectious disease, salivary gland tumors, extension of squamous cell carcinoma of the floor of the mouth, or nodal metastases. Frequently, benign mixed salivary gland tumors are isointense on T1-weighted imaging, are hyperintense on T2-weighted imaging, and enhance avidly. Although malignant salivary gland lesions may be hypointense on T2-weighted imaging and display variable enhancement, they often appear similar to benign lesions, making definitive diagnosis difficult. Therefore, biopsy or excision may be required.

In people who are elderly or severely ill, schwannomas may be monitored. Otherwise, the treatment is complete surgical excision. Often, the nerve can be dissected from the encapsulated tumor mass, thus preserving nerve function. When a nerve segment must be sacrificed because of tumor involvement, immediate nerve grafting may be performed. After excision, recurrence is rare. As was the case in our patient, resection of hypoglossal nerve tumors may result in palsy of the 12th nerve and consequent speech and swallowing difficulties. Postoperative Horner’s syndrome, the most common complication after excision of masses along the sympathetic chain, may be partial or complete and may result in a permanent neurologic deficit [6]. Dissection in the parapharyngeal space may be complicated by lower cranial nerve injury, which is manifested by vocal cord paralysis, dysphagia, or shoulder weakness.

References