A 32-year-old man was presented to the Department of Otolaryngology, Loghman Hakim Hospital, Shaheed Beheshti University of Medical Sciences, Tehran, Iran complaining of a left-sided neck lump since five months. The mass was located in the posterior part of the left submandibular region (Figure 1). There was no history of infections of upper respiratory tract, dental problems, or inflammations of oral cavity, oropharynx, salivary glands, nose, and paranasal sinuses. There was no pain, malaise, fever, or weight loss. He had no dysphagia, odynophagia, hoarseness, regurgitation, aspiration, or sense of pharyngeal numbness.

On physical examination, there was a mass 4×4 cm in diameter involving level IB and superior level II of the neck. It was firm on palpation and mobile only in the anterior-posterior direction. The skin covering the lesion was normal and no sign of topical inflammation was present. Oral cavity, oropharynx, nose, nasopharynx, and larynx were normal. No other mass or lymphadenopathy was found in the neck.

Laboratory tests including a complete blood count, and erythrocyte sedimentation rate were normal. PPD was 5 mm; mononucleosis test, toxoplasmosis test, and HIV Ab were all negative. Chest X-ray was normal. Contrast enhanced axial computed tomography (CT) of the neck revealed a homogeneous round mass in the left side of the neck located in poststyloid region close to the carotid arteries and jugular vein. It was separated from submandibular and parotid glands. The other parts of the neck were normal (Figure 2). Fine-needle aspiration biopsy revealed only blood elements.

The patient underwent excisional biopsy. The mass was round and well-encapsulated without infiltration of adjacent tissues. It adhered to vagus nerve (Figures 3). Palpation of the mass, caused bradycardia.

What is Your Diagnosis?

See the next pages for the diagnosis.
A schwannoma (neurilemoma) is a benign neurogenic neoplasm composed of Schwann’s cells. This tumor may occur at all ages and does not show a preference for men or women. Approximately 10% of them arise from the vagus nerve.1 Till June 2000, only 95 schwannomas have been reported in the literature, the majority being in patients between 30 and 60 years of age.2 Between 25% and 45% of all reported schwannomas are found in the head and neck region.

These sites include the parapharyngeal space, neck, paranasal sinuses, nasal and oral cavities, face, scalp, intracranial cavity, and larynx.3 Most schwannomas are solitary, but they rarely may be multiple or associated with von Recklinghausen’s disease. Tumors involving the parapharyngeal space most commonly arise from the vagus or cervical sympathetic chain, and may present as a mass displacing the lateral pharyngeal wall or tonsil into the oropharynx. Pain is suggestive of a schwannoma but neurologic defects are unusual.4

The tumors have a distinctive pattern on

Discussion

![Figure 2. CT of the neck demonstrating a homogeneous mass adjacent to the left side of great vessels.](image)

![Figure 3. Intraoperative views of the tumor: A) the tumor is well-encapsulated and B) adheres to vagus nerve.](image)
histologic examination (Figure 4). A palisading array of nuclei around a central mass of cytoplasm which termed “Antoni type A” tissue; and a loose, surrounding stroma with no distinctive fiber and cell pattern the so-called “Antoni type B” tissue. Necrosis, cystic degeneration, and focal thrombosis are prominent in the solitary schwannoma. Malignant changes seldom occur.⁶

Tumors that arise from the vagus or cervical sympathetic chain tend to displace the internal carotid artery anteriorly. Most schwannomas have significant enhancement on postcontrast magnetic resonance imaging (MRI) and thus may be confused with a paraganglioma.⁷

Tumors of the parapharyngeal space can be divided into pre- and poststyloid spaces by the fascia of the tensor veli palatini; tumors of the poststyloid space are almost always either paragangliomas or nerve sheath tumors. Contrast imaging studies assist in differentiation.⁸

A preoperative diagnosis may be made with some certainty based on a high index of suspicion from the history of painless swelling, and characteristically mobile laterally but immobile vertically, reflecting its attachment to the vagus nerve. CT, MRI, and angiography may obtain confirmation for the diagnosis. Incisional biopsy is unnecessary and contraindicated because of the vascular nature of the lesion and the possibility of uncontrolled hemorrhage. It may also make removal of the tumor mass difficult because of obliteration of tissue plane.⁵

Gross total resection remains the treatment of choice for these tumors. The capsule is gently and carefully dissected from the fascicles of the nerve. When it is necessary to debulk the tumor, the capsule is incised longitudinally to preserve the uninvolved fascicles. However, as much as possible of the capsule should be removed to prevent recurrence. If the nerve or some of the fascicles cannot be salvaged, a split repair should be performed using the great auricular or sural nerve. Where it is not possible, vagus nerve is sacrificed along with the tumor. Hoarseness is almost always present after resection and recovers in most cases. Other common complications include pharyngo-laryngeal anesthesia, aspiration, and paralysis of the cranial nerves IX, XI, and XII, which may be transient or permanent.⁵

References


Figure 4. Histopathology of the cervical vagus nerve schwannoma (H&E, ×100).