Abstract
Schwannomas of head and neck region commonly arise from the vestibular and vagus nerve. Hypoglossal nerve schwannomas are very rare. They may be intracranial only or have both intra and extracranial extension. Radiology specially, computed tomography (CT) and magnetic resonance (MRI) are useful in not only diagnosis but also identifying the nerve of origin. Total excision of the tumor via an external approach is the treatment of choice, for extracranial hypoglossal nerve schwannoma.

Introduction
Schwannomas are solitary, encapsulated, benign tumors originating from Schwann cells, usually attached to or surrounded by a nerve of origin [1]. Head and neck schwannomas account for 25% to 45% and often involve cranial nerve IV, V, VII, X, XI, XII or the sympathetic and peripheral nerves [2]. About 10% of schwannomas in the neck region originate from vagus or sympathetic nervous system [3].

Schwannoma arising from hypoglossal nerve is very rare and may develop in intracranial part or may involve both the intracranial and the extracranial components assuming a dumb-bell shape [4]. Peripheral i.e. extracranial hypoglossal schwannomas are extremely rare, and only few cases have been reported in literature till date [5].

Pre-operative computed tomography (CT) and magnetic resonance (MRI) are used to identify its origin. We present herein a rare case of hypoglossal nerve schwannoma presenting as right submandibular space mass.

Case Report
A 45-year-old male presented with gradually progressive mass in the right submandibular region since 6 years. It was not associated with increased in size following food ingestion. There was no history of dental pain, difficulty or painful swallowing or discharge of purulent saliva from the floor of the mouth. Past, family and personal history were not contributory.

The local physical examination revealed a 4.5 x 5cm soft, non-tender, non-pulsatile, well-defined mass in the right submandibular region. Oral examination was normal.

Computed tomography imaging (CT) showed well-circumscribed oval mass in the right carotid space measuring 4.2cm x 3.2cm x 3cm anterior to carotid sheath vessels displacing and slightly compressing the right submandibular gland anteriorly (Figures 1, 2). All the routine blood and urine investigations were normal.

Trans-cervical fine needle aspiration cytology (FNAC) of the mass suggested Schwannoma. Patient underwent surgical excision of the mass via transcervical approach under general anesthesia. Nerve of origin was identified, dissected free from the mass and preserved (Figures 3, 4).

Figure 1: Axial section of Contrast enhanced computed tomogram (CECT) showing, well-circumscribed, 4.2cm x 3.2cm x 3cm oval mass in the right submandibular space.

Figure 2: Coronal section CECT showing same soft tissue mass in right submandibular space.
Macroscopically, the excised mass was oval, smooth, encapsulated and yellowish, measuring 5cm in its greatest diameter. The patient made an uneventful postoperative recovery with complete resolution of symptoms without any cranial nerve deficit. Histopathology examination of excised mass, reported spindle cells arranged in both Antoni A and B pattern.

Discussion

Schwannomas originate from schwann cells of peripheral, cranial or autonomic nerves [1]. They are typically seen between fourth and sixth decade of life. While most studies suggest female preponderance [3], many others have reported report no age or sex predilection [1,4,5]. Head and neck schwannomas usually involve sensory divisions of cranial nerves, commonly the vestibular and vagal nerve [6]. Hypoglossal schwannoma is very rare, since the nerve consists only of a motor component [6].

Hypoglossal nerve emerges from the medulla oblongata between the pyramid and the olive. During its extracranial course, the nerve traverses through the hypoglossal canal, curves ventrally between the pyramid and the olive. During its extracranial course, only of a motor component [6].

Hypoglossal nerve may be considered in differential diagnosis. We advocate that total excision of the tumor via an external approach must be the treatment of choice.

Clinically schwannomas may present as an asymptomatic mass, a newly developed neurogenic symptom or sign that may be related to the nerve of origin [3]. In our case, the initial clinical impression included differentials like salaladenitis, paraganglioma, branchial cleft cyst, malignant lymphoma and metastatic cervical lymphadenopathy. Salaladenitis usually has association with meals however there was no such complaint and submandibular gland appeared normal on intraoral palpation as well as on CT imaging. The metastatic lymph nodes are often multiple, with an evident primary, making the diagnosis easy. However, a solitary metastatic node from an unknown primary cancer may be difficult to differentiate from a schwannoma in the absence of peridenitis or extra nodal extension of disease [8]. Paragangliomas show early arterial enhancement on CT, they are hypervascular lesions, while schwannomas are hypovascular [9].

Deep lobe parotid tumors, lipomas and lymphadenopathy are usually seen arising in pre-styloid compartment; moreover the loss of the fat plane between the mass and the parotid gland suggests a deep lobe parotid origin. MRI must be the treatment of choice. Yonsei Med J 51: 938-942.

We report a case of hypoglossal schwannomas in submandibular space. Although schwannoma in submandibular space is very rare, but when a well circumscribed mass is observed, as in the present case, schwannoma may be considered in differential diagnosis. We advocate that total excision of the tumor via an external approach must be the treatment of choice.

References