CASE REPORT

Three Cases of Head and Neck Schwannomas: Varied Clinical Presentation

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SUMMARY

Schwannomas are benign tumors. A series of three unusual cases involving the head and neck region at the Department of ENT, Hospital Ipoh from July 2004 to June 2005 is presented. The first case was a pedunculated schwannoma of the tongue base. The second was a schwannoma of the cervical sympathetic chain who developed a transient Horner's Syndrome upon fine needle aspiration cytology. The third case was a bilobed cervical vagal schwannoma which developed immediate vocal cord palsy post-operatively which was evident at six months follow-up. All tumors were removed surgically.

KEY WORDS:
Schwannomas, Varied clinical presentation, Base of Tongue, Horner's Syndrome, Vocal Cord Palsy

INTRODUCTION

Schwannomas also referred to as neurilemmomas are benign encapsulated nerve sheath tumors composed of Schwann cells. As nerves exit the brain there is a change from myelination by oligodendrocytes to Schwann cells. Schwannomas are formed by proliferating Schwann cells encompassing peripheral motor and sensory nerves. They may arise at any age and there is no gender predilection[1]. About 25% of extracranial schwannomas are located in the head and neck region with the lateral cervical region and the oral cavity being the most common sites[2]. Schwannomas involving the tongue base, cervical sympathetic chain and a bilobed cervical vagal schwannoma are rare entities and have never been reported in Malaysia.

PATIENT 1

A 17 year old Malay boy presented with a one year history of a progressively enlarging growth in the oral cavity causing difficulty in swallowing. On examination a papillomatous, pedunculated lesion measuring 1cm by 1cm was visualised on the left base of tongue. The growth was snared via an intra-oral route after fixation with a modified Boyles-Davies mouth gag under general anaesthesia. The base was then excised by cutting diathermy.

Histological section showed a well encapsulated lesion composed of compact spindle cells with twisted nuclei and occasionally intranuclear vacuoles (Antoni A area) and less orderly and less cellular areas composed of spindle to oval cells (Antoni B area) and Verocay bodies consistent with a schwannoma. (Figure 1) Post-operative recovery was uneventful with no evidence of recurrence noted at one year follow-up.

PATIENT 2

A 35 year old Malay lady presented with a right painless cervical swelling measuring 3cm by 3cm for eight months. FNAC was performed and the patient developed Horner's Syndrome immediately which gradually recovered. The FNAC was unsatisfactory. A CT scan with contrast was performed to rule out a vascular tumor (Figure 2).

The tumor was excised surgically via a cervical incision and the histopathological examination (HPE) confirmed as schwannoma. At six months post-surgery the patient was noticed to have mild right ptosis.

PATIENT 3

A 47 year old Chinese lady presented with a slow growing painless left cervical swelling for 18 years. FNAC was reported as a degenerative cyst. A contrast CT scan was performed. The tumor was excised transcervically and was confirmed a schwannoma on the histopathological report. Grossly the tumor was bilobed and 'egg-like' in appearance (Figure 3).

Post-operatively the patient developed a left vocal cord palsy. At six months follow-up the patient had hoarseness and a weak voice with no evidence of aspiration.

DISCUSSION

Schwannomas were first described by Verocay[3] in 1908. They are benign, slow growing tumors with rare malignant change. These tumors originate from the neural crest which differentiates into the Schwann cell and the sympthicoblast. The latter cell gives rise to paraganglionic cells from which arise carotid body tumors, glomus tumors and gangliomas[4].

Histologically, two tissue forms have been described. The Antoni type A tissue shows well-developed cylindrical bands of Schwann's cells and connective tissue fibres with a tendency towards pallisading of the nuclei about a central mass of cytoplasm (Verocay bodies). Antoni type B tissue is a

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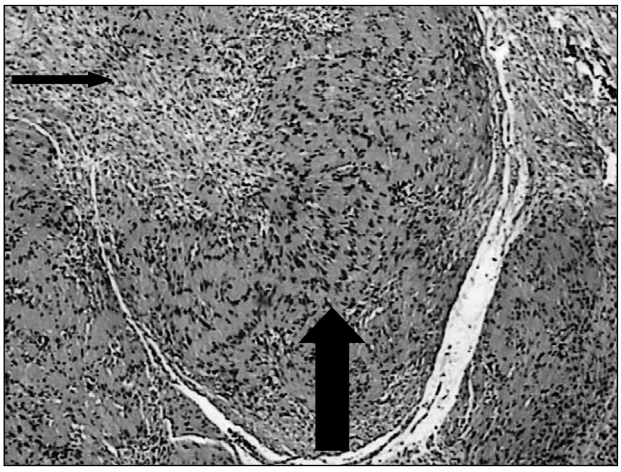

Fig. 1: Well circumscribed schwannoma lesion composed of Antoni A (thick arrow) and Antoni B (thin arrow) areas

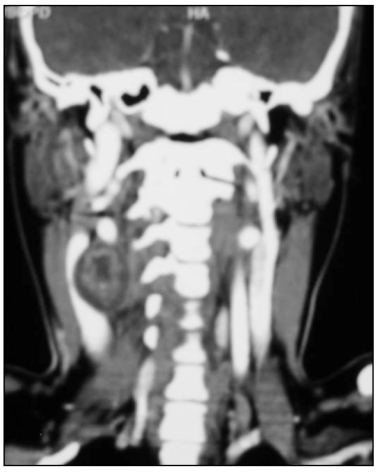

Fig. 2: CT scan showing right cervical tumor

Fig. 3: Bilobed cervical vagal schwannoma

loosely arranged stroma in which the fibres and cells form no distinctive pattern.

Diagnosis of Schwannomas is based on histology. Intra-orally the tongue is the most common site. However, lesions involving the tongue base is extremely rare. For small tumors, as in this case, simple excision is usually curative. However, larger tumors may require excision via a submandibular or lip-splitting approach.

Fewer than 50 cases of schwannomas involving the cervical sympathetic chain have been reported in the English literature[5]. Horner's Syndrome is commonly a feature of these tumors post-operatively or may even be a presenting feature. First bite syndrome was not encountered in this patient. This syndrome consists of pain in the parotid on the first bite of food. It is thought to be due to denervation hypersensitivity of the sympathetic receptors that control myoepithelial cells in the parotid gland.

Cervical schwannomas are nearly all fusiform[4]. A bilobed appearance is noted with skull base involvement. A bilobed cervical vagal schwannoma has not been described previously. Management, as with schwannomas elsewhere consists of complete excision. Post-operatively, the patient developed left vocal cord palsy. This is known complication of vagal schwannoma excision as the nerve and tumor are often inseparable. A medialisation thyroplasty can be performed to overcome this problem. The option was rendered to the patient which she refused as she felt the mild hoarseness was not bothersome.

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