Cervical Schwannoma: Report of Four Cases

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SUMMARY

Extracranial schwannomas in the head and neck region are rare neoplasms. The tumours often present as asymptomatic, slowly enlarging lateral neck masses and determination of the nerve origin is not often made until the time of surgery. Preoperative diagnosis maybe aided by imaging studies such as magnetic resonance imaging or computed tomography, while open biopsy is no longer recommended. The accepted treatment for these tumors is surgical resection with preservation of the neural pathway. We report four cases of cervical schwannomas that we encountered at our center during four years of period. The clinical features, diagnosis and origin, management and pathological findings of these benign tumors are discussed.

KEY WORDS:
Schwannoma, Cervical schwannoma, Extracranial schwannoma

INTRODUCTION

Schwannomas, also known as neurilemoma or neurinomas are benign nerve sheath neoplasm that may originate from any peripheral, cranial or autonomic nerve of the body. About 25% to 45% of schwannomas are located in the head, and it may involves the cranial nerve such as V, VII, IV, X, XI and XII or the sympathetic and peripheral nerve [3, 5]. About 10% of schwannomas that occur in the head and neck region generally originate from the vagus or sympathetic nervous system [2]. Preoperative imaging studies such as magnetic resonance imaging and computed tomography are used to distinguish its location and origin, while open biopsy is not recommended [3, 4]. The treatment of schwannoma is surgical resection, with several surgical modalities have been introduced to preserve the neurological function [5].

We report four cases of schwannoma that we encountered at our center during four years of period, between 2007 and 2010. The clinical features, diagnosis dilemma and origin, management and pathological findings of these tumors are discussed. (Table I)

CASE REPORTS

Case 1

A twenty-six-year-old lady, a known case of Neurofibromatosis Type II presented with a progressively enlarging left neck swelling for four months duration. The swelling was painless but gradually increasing in size. Otherwise, she denied dysphagia, odynophagia, tongue numbness or slurred speech.

Physical examination revealed a 3 x 3 cm mass located at right lateral upper third of the cervical region. The mass was firm, mobile and non-tender. She had a right facial nerve palsy (House-Brackman Grade IV) due to a previous operation for a right acoustic neuroma in 2000 and a right supraorbital wound for a plexiform schwannoma in 2007.

A computed tomography of the neck and thorax was performed and showed a well defined, minimally enhancing lesion at the left parapharyngeal space measuring 2.4 x 4.3 x 10 cm. The left carotid sheath vessel is displaced medially. The lesion had similar density to a previous computed tomography done, giving the impression of a schwannoma. No tissue biopsy was taken and the patient was treated conservatively as it involved major vessels such as the carotid and jugular vein.

The patient is still asymptomatic upon the last review in early 2011.

Case 2

A sixty-nine-year-old lady presented with a right neck swelling for more than one-year duration. The swelling was painless but gradually increasing in size. Otherwise, she denied odynophagia, slurred speech or loss of facial sensation.

Physical examination revealed a 4 x 5 cm mass located at right upper third of the cervical region. The mass was firm, mobile and non-tender. Other ear, throat and nose examinations were normal. All neurological examinations were intact.

A computed tomography of the neck and thorax was performed and showed a well-defined mass in the posterior triangle region, not involving the intra-spinal muscle measuring 5.0 x 4.0 cm.The mass was totally excised via a right transcervical approach. The well-circumscribed mass was located between sternocleidomastoid muscle and scaleneus muscle. It measured approximately 4.0 x 4.0 x 5.0cm.

The histological examination confirmed a schwannoma with Antoni A and Antoni B areas, neoplastic Schwann cells with hemorrhages and necrosis. Verocay bodies are seen.

There was no recurrence noted at a sixth month postoperative visit but neurological deficit such as numbness at right ventral aspect of third and forth fingers remained unchanged with no sign of recovery.

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Table I: Clinical Patient Summary; epidemiological data, clinical findings, diagnostic modalities, nerve of origin and post-operative complications

Patient number/ Year	Age(yrs) / Sex	Tumor size (cm)	Diagnostic modalities	Resection	Origin	Postoperative Deficit
1/2007	26/F	3.0x3.0x10.0	CT - Schwannoma	Conservative	C2	Nil
2/2008	69/F	4.0x4.0x5.0	CT and histopathology-	Gross total	C7	Hypoesthesia over hand at C7 distribution, no sign of improvement
3/2010	34/F	3.0x3.0x3.0	CT and histopathology-Schwannoma	Gross total	Hypoglossal	Right hemiatrophy of tongue, no sign of recovery
4/2010	47/M	3.0x4.0x3.0	CT and histopathology-Schwannoma	Gross total	Cervical plexus	Hypoesthesia over right pinna; no sign of improvement

*CT; computed tomography

Case 3
A thirty-four-year-old lady presented with a right neck swelling for ten months duration. The swelling was painless but gradually increasing in size. Otherwise, she denied odynophagia, tongue numbness or paraesthesia or slurred speech.

Physical examination revealed a 3 x 3 cm mass located at right upper third of the cervical region. The mass was firm, mobile and non-tender. Other ear, throat and nose examinations were normal.

A computed tomography of the neck and thorax was performed and showed a well-defined, minimally enhancing lesion in the right carotid space, measuring 3.6 x 6.0 x 4.2 cm. It extended from ramus of mandible downwards to the vocal cord level and medially extended to hyoid bone (Fig. 1).

The mass was totally excised via a right transcervical approach. The well-circumscribed mass was located at the anterior third of the stenoclaidomastoid muscle, just below the submandibular gland. It measured approximately 3 x 3 x 3 cm. The mass pushed the right internal jugular vein laterally and carotid artery posterolaterally. The pedicle of the mass had arisen anteriorly and posteriorly in a horizontal plane. (Fig. 2)

The histological examination confirmed a schwannoma with Antoni A and Antoni B cells with hemorrhages and necrosis. There was no recurrence or complications noted at the fifth month postoperative visit but neurological deficit such as paralysis of the right side of the tongue remained unchanged with no sign of recovery.

Case 4
A forty-seven-year-old man presented with a right neck swelling for one-year duration. The swelling was painless but gradually increasing in size. Otherwise, he denied dysphagia, odynophagia or changes of voice.

Physical examination revealed a 3 x 40 cm mass located at right upper third of the cervical region. The mass was firm, mobile and non-tender. Other ear, throat and nose examinations were normal.

A computed tomography of the neck was performed and showed a well-defined lesion in the right posterior cervical space measuring 2.8 x 3.8 x 6.0 cm. The mass was completely resected via right transcervical approach. The well-circumscribed mass was located at the posterior triagle, posterior to the sternocleidomastoid. It measured approximately 3 x 4 x 3 cm. The mass had arisen from multiple cervical nerves with multiple exits noted.

The histological examination confirmed a schwannoma with Antoni A and Antoni B cells with focal area of cystic degeneration and dilated blood vessels with hyalinized wall. There was no evidence of the disease noted at third month postoperative visit but neurological deficit such as hypoesthesia over the right pinna is still present. No hoarseness or voice changes noted.

DISCUSSION

Schwannomas are benign, slow growing peripheral nerve tumors, only about 25% to 45% occur in the head and neck region [1, 3]. Schwannomas are also referred to as neurilemmomas and neuromas. Schwannomas typically present between the forth and sixth decade of life but can occur at any age. There is no sex predisposition for head and neck schwannomas but we observe female predominance in our series, as well as been reported by Kang *et al* [2, 3]. Clinically, schwannomas presented with asymptomatic neck masses that caused little concern other than the possibility of malignancy and cosmesis [1, 3]. It correlates with our series that all our patients presented with an asymptomatic slow-growing lateral neck masses that can be palpated along the medial border of sternocleidomastoid muscle.

Pre-operative diagnosis of schwannomas is difficult because many schwannomas do not present with neurological deficits and several differential diagnoses for tumour of the neck may be considered, including paraganglioma, branchial cleft cyst, malignant lymphoma and metastatic cervical lymphadenopathy. Furthermore, due to their rarity, these tumors are often not even taken into consideration in the differential diagnosis [2, 3]. The possible neurogenic symptoms and signs related to cervical schwannomas depends on the origin of the tumour, such as pain and tenderness, coughing upon exerting pressure on tumor which is unique presentation of vagal schwannomas, headache, Horner's syndrome, cranial verve palsies (particularly V, VI, VII, XII), tinnitus or hearing loss, which are not observed in our series[2,3].

There is general agreement concerning the value of magnetic resonance imaging in the pre-operative work-up as it is helpful in defining diagnosis and in evaluating the extent and the relationship of the tumor with the jugular vein and the carotid artery. Magnetic resonance imaging is the best diagnostic aid for these tumors [4]. The magnetic resonance imaging appearance of these nerve sheath tumors is quite typical and diagnosis can often be made, or at least strongly suspected although it cannot distinguish between schwannoma and neurofibroma [2, 3, 4]. However, due to lack of recourses, only computed tomography scan was done to assess the tumor. On computed tomography scan, schwannoma appear as a well and fusiform mass with a relatively homogenous contrast enhancement with internal cystic changes that becomes more prominent as the tumour enlarges. This cystic change is associated with mucinous degeneration, haemorrhage, necrosis and microcystic formation [2].

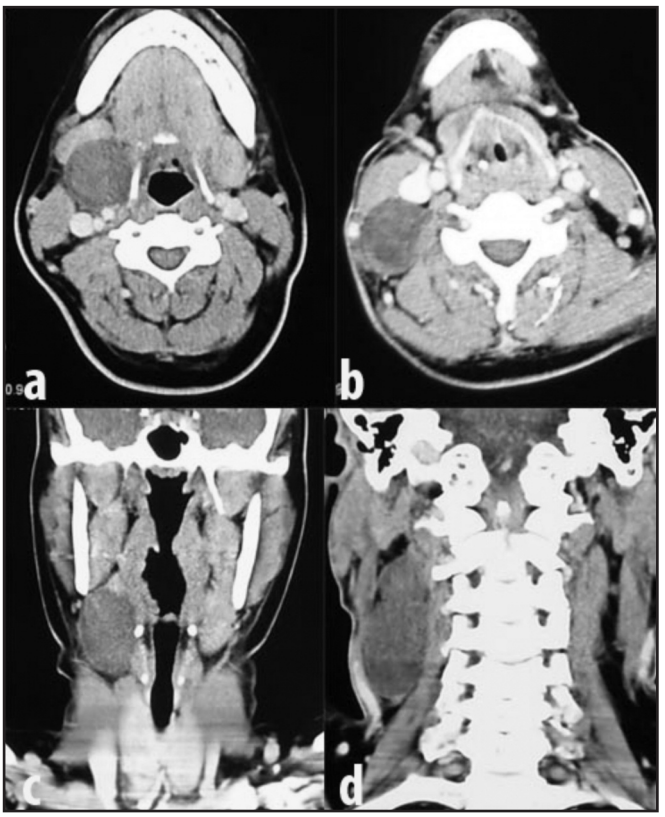

Fig. 1: Computed tomography axial (a) and coronal (c) of Patient 3 demonstrates a well-defined, enhancing lesion in the right carotid space, extending from the ramus of mandible downwards to the vocal cord level and medially extends to the hyoid bone; axial (b) and coronal (d) of Patient 4 demonstrates a well-defined lesion in the right posterior cervical space.

The useful of fine-needle aspiration and cytology is still controversial; the majority of authors does not recommend open or needle biopsy for these masses [3]. Incisional biopsy will obliterate tissue plane thus make removal of tumour mass difficult. In our series, all patients had undergone prior fine-needle aspiration and cytology and no conclusive diagnosis could be obtained. Microscopically, both tumour types display well-differentiated, fusiform, spindle cell without frequent mitoses [1]. Schwannomas exhibit a distinctive cylindrical structures and tendency towards palisading of the nuclei (Verocay bodies) which called Antoni A tissue, and a nondistinctive, loose stroma of fibres cells called Antoni B tissue. Typical features include hemorrhage and cystic degeneration.

Treatment of vagal nerve tumours is a complete surgical excision because they are relatively radioresistant [1, 3]. Total extirpation of the tumor, with preservation of intact fascicles, is always the goal for surgical treatment of nerve sheath tumours, and surgery should be planned with this objective in mind. Incomplete treatment, such an open biopsy or exploration without tumor removal should not be performed. Recently, intracapsular enucleation has been introduced to preserve the neurological functions. Intracapsular enucleation preserves the nerve fibres function more than

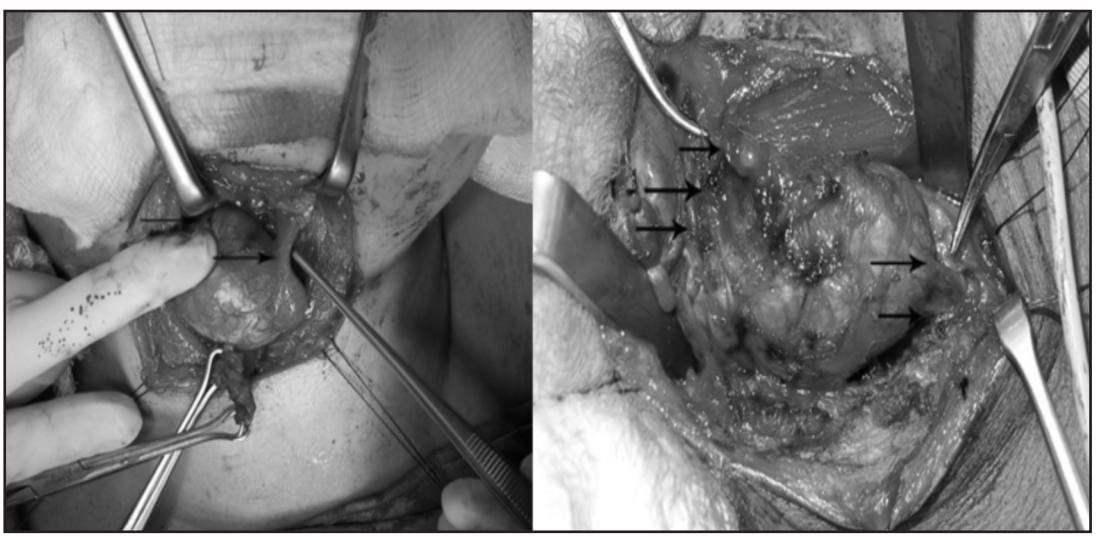

Fig. 2: Operative findings of Patient 3 (left) showing a pedicle from the hypoglossal nerve and Patient 4 (right) showing multiple exits of the nerve.

30% when compared to tumour resection with primary anastomosis [5].

We used a lateral cervical approach, via carotid-type incision, for total resection. The key anatomic point is that schwannomas arise from a solitary entering nerve fascicle at the proximal pole; gross total removal can be accomplished with sacrifice the entering and exiting fasciles. If it is impossible to find an adequate plane and is technique difficult to preserve the integrity of the nerve trunk, the involved segment may be resected and an end-to-end anastomosis performed using microsurgical technique [4].

Patients should be informed about the possible post-operative complications. Vocal cord palsy has been reported as high as 85% in the literature [3]. Other common complications include pharyngolaryngeal anesthesia, aspiration and cranial nerve IX, X, XII palsies, which maybe transient or permanent. Cardiac event such as electrocardiogram abnormality, severe bradycardia and event cardiac arrest during tumor dissection have been reported. Therefore, patient should be monitored carefully for electrocardiogram abnormalities during manipulation of the tumour, and the anesthetic team should be prepared to address any resultant hemodynamic instability[3].

CONCLUSION

Extracranial schwannomas in the head and neck region are rare neoplasm. Diagnosis is establish by imaging studies such as magnetic resonance imaging or computed tomography, while FNAC is used o rule out other condition. The accepted treatment for these tumors is surgical resection with preservation of the neural pathway.

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