Retrospective study on extracranial neck schwannomas in tertiary care hospital in Nepal

Shrestha UK, Shrestha KR

Department of Cardiothoracic and Vascular Surgery,

Manmohan Cardiothoracic Vascular and Transplant Center, Kathmandu, Nepal

Correspondence to: Dr. Uttam Krishna Shrestha

Manmohan Cardiothoracic Vascular and Transplant Center

Maharajgunj, Kathmandu, Nepal

E mail: u.nayaju@gmail.com

Abstract

Introduction: Schwannomas are relatively rare tumors arising from Schwann cells that ensheath the peripheral nerves. The study aim was to identify extracranial neck schwannomas treated in a tertiary care hospital in Nepal.

Methods: We reviewed medical records of all patients with extracranial head and neck schwannomas treated at our department from July 2004 to June 2014.

Results: Ten such patients consisted of 7 males and 3 females were identified, with a mean age of 37.8 +/- 10.7 years (range: 20 to 54 years) with solitary schwannomas. Mean size of tumor was 5 cm (range: 2-10 cm). Most of the patients (60%) presented with asymptomatic palpable mass. Of these ten, 50% had a left sided neck mass. Seven originated from vagus nerve while three from sympathetic chain. The average duration of symptoms ranged from one month to two and half years. In all cases, tumor was completely resected through cervical approach. All tumors were enucleated keeping nerve of origin intact. In all cases, the tumor was completely resected surgically. The average follow up period ranged from one month to 108 months (median: 24 months). No major postoperative complications were noted. Two patients (20%) developed Horner's syndrome and one (10%) had temporary hypoglossal nerve palsy which recovered within three months.

Conclusion: Non-vestibular extracranial head and neck schwannomas most frequently present as an innocuous longstanding unilateral neck mass. The mainstay of treatment is complete excision preserving the nerve of origin. Early recognition of Schwannomas is key to optimal treatment.

Keywords: cranial nerves, Horner's syndrome, Schwannoma.

Introduction

Schwannomas known as neurilemmomas, neuromas, or neurinomas are relatively rare tumors arising from Schwann cells that ensheath the peripheral nerves. These are encapsulated, highly vascular and slow growing benign neural sheath tumour.[1] Malignant change is unusual.[2] They may be found anywhere in the body but 25 to 45 % of extracranial schwannomas occur in the head and neck region.[1,2] Extracranial head and neck schwannoma is a challenging condition to the head and neck surgeons. It can involve the cranial nerves with the exception of the olfactory and optic nerve as well as sympathetic and peripheral nerves.[3]

Preoperative diagnostic investigations included ultrasonography (US), computed tomography (CT), magnetic resonance imaging (MRI) and fine needle

aspiration cytology (FNAC)[1,2,4]. However, the preoperative diagnosis of schwannoma is difficult and should be suggested by clinical features and supported by investigations. As for the management of schwannomas, multiple treatment options exist including observation, complete tumor excision and intracapsular enucleation.[4] For tumours arising from the major cranial nerves, excision of tumour with the division of the nerve of origin renders lifelong morbidity to the patients.

Methods

All patients with head and neck schwannomas treated at our department between July 2004 and June 2014 were retrospectively reviewed from the medical records of the Tribhuvan University Teaching Hospital and Manmohan Cardiothoracic Vascular and Transplant Center, the largest tertiary hospital in Nepal. Data collected included age, sex, race, presenting signs and symptoms, anatomical location of the tumour, tumour size, nerve of origin, diagnostic modality, surgical approach, intraoperative finding, histopathological finding and outcome after treatment.

Fine needle aspiration cytology (FNAC) was performed for tumours located at the superficial part of the neck. For tumours which were located at the deeper part of the neck, e.g. parapharyngeal space, it was usually not possible to obtain tissue for cytological or histological diagnosis before operation. In all patients, computed tomography (CT) neck was performed.

Ten patients with head and neck schwannomas were identified. One patient with no information other than age and diagnosis of a recurrent head and neck schwannoma was excluded from the study. Our study set of schwannomas was necessarily extracranial non-vestibular and mainly non-trigeminal because the vestibular and trigeminal schwannoma patients usually receive tertiary treatment through either otolaryngology or neurosurgery services.

The study population consisted of seven males and three females, with a mean age of 37.8 +/- 10.7 years (range: 20 to 54 years) with a solitary schwannoma. All patients underwent preoperative computed tomography (CT) to facilitate diagnosis and delineate extent and related anatomy. All patients underwent surgical excision of the tumour. Table I shows the epidemiological data along with the presenting signs and symptoms and initial diagnosis.

Mean size of tumour was 5 cm (range 2- 10 cm). Most of the tumours (60%) presented themselves as asymptomatic palpable mass, two (20%) presented with painful neck swelling while one (10%) presented with facial weakness and in one patient details of presentation were not available. Most of the patients presented with only one sign or symptom. Five of the ten lumps were left sided. Seven originated from vagus nerve while three arose from sympathetic chain. For diagnosis, Fine needle aspiration was performed in 3 cases and gave a cytological diagnosis in one case only. The predominant feature was presence of spindle cells. Radiological investigations included carotid doppler ultrasound which resulted in a misdiagnosis of a carotid body tumour in one case. Computed tomography (CT) scans were performed in all cases and showed intensely contrast-enhancing mass lesions abutting and compressing the carotid artery (Fig 1 and 2) system in keeping with a vascular tumour within the parapharyngeal space.

Time before seeking medical attention ranged from one month to two and a half years. In all cases, tumour was successfully and completely resected through cervical approach. All tumours were enucleated preserving nerve of origin. Till date there has not been any recurrence after excision of schwannoma. In all cases, the tumour was completely resected surgically.(Fig 3)

The presence of characteristic Antoni A or B histologic patterns with or without S-100 stain was used to identify schwannomas. S-100 immunohistochemical staining shows mitoses, necrosis, invasiveness and specific features such as nuclear hyperchromia and pleomorphism or large atypical cells thereby confirming the benign nature of the schwannoma.[2] (Fig 4) Grossly schwannomas are well circumscribed encapsulated firm, grey myxoid masses attached to nerve but may have areas of cystic and xanthomatous change.

Results

Complete removal of schwannoma were possible in all cases. None of the patients suffered wound infection or hematomas. Two patients (20%) developed Horner's syndrome and one (10%) had temporary hypoglossal nerve palsy which recovered within three months. Follow-up ranged from one month to 108 months (median: 24 months). All remained free of disease at last follow-up. None of the Schwannomas was malignant. Epidemiological data, presenting signs and symptoms, diagnostic modality, preoperative diagnosis and nerve of origin are shown in table 1.

Table 1. Status of clinical history and epidemiological information

Pts	Age/ Sex	Symptoms	Initial diagnosis	Diagnostic modality	Nerve of origin
SP	35/M	Rt neck mass– 2 and half years	Soft tissue tumour	FNA – blood, degenerate cells, spindle cells CT scan- neurogenic tumour	Rt Vagus
RK	21/M	Lt Neck mass- Two and half years	Enlarged lymph node	CT scan- vagal schwannoma	Lt Vagus
TB	20/M	Pain in neck	Branchial cyst	FNAC- bloody aspirate CT scan- hemorrhagic thyroid cyst	Lt Vagus
RBT	47/M	Rt neck mass- 2yrs	Neurogenic tumour	CT scan- neurogenic mass	Rt Vagus
MK	46/F	Rt neck mass- 1yr	Soft tissue mass	CT scan- neurogenic tumour	Rt Vagus
NMM	54/F	Lt neck mass and pain- 1yrs	Enlarged lymph node	FNAC- bloody aspirate CT scan- multiple enlarged lymph node with ?Neurogenic mass	Lt Vagus
NR	36/M	Weakness in facial muscle	Neurogenic tumour	CT scan- neurogenic tumour	Lt Sympathetic chain
LNJ	38/M	Pain in lt neck	Enlarged lymph node	CT scan- enlarged lymph node	Lt Vagus
AS	39/M	Rt neck mass- 2 years	Neurogenic tumour	CT scan- neurogenic tumour	Rt Sympathetic chain
BK	42/F	unknown	Rt Carotid body tumour	CT scan- vascular mass at bifurcation of carotid artery	Rt sympathetic chain

On reviewing the histology, there was little difficulty in histological diagnosis in the benign group. The typical features of spindle cells with elongated nuclei and alternating areas of organized, compact cells (Antoni A Configuration) and loosely arranged, relatively acellular tissue (Antoni B Configuration) being present in all cases (Figure 4). The classic feature of compact groups of parallel spindle-shaped nuclei (Verocay bodies) were, however, only present in one of the ten cases (10%).

Figure 1. CT showing left neck schwannoma lying posterior to carotid arteries

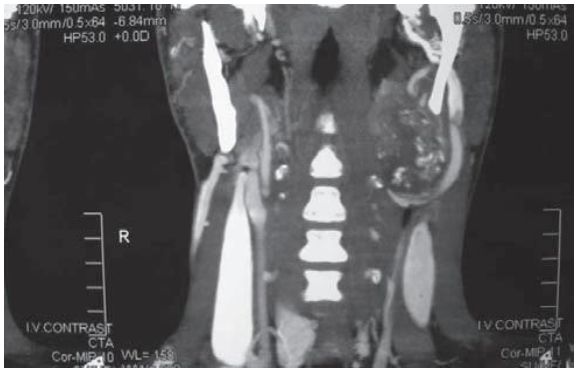

Figure 2. CT angiogram depicting highly vascular schwannoma in the left neck

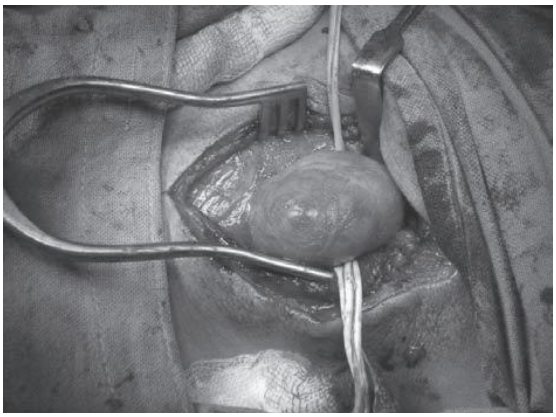

Figure 3. Well encapsulated sympathetic chain schwannoma in left neck

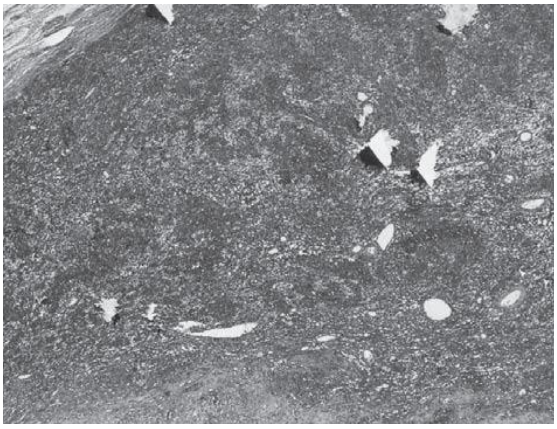

Figure 4. Histopathological feature of schwannoma3 showing spindle cells with elongated nuclei and alternating areas of organized cells (NSE stain)

Discussion

Schwannoma are diagnosed incidentally while investigating for nonspecific complaints as they remain quiescent for long periods of time. Between 25-45% of all reported extracranial schwannomas are found in head and neck region. In the study by Torossian[4] et al 2/3rd of the study population were females while in the study by Leu and Chang,[5] there was male predominance which is similar to our study. Most supraclavicular schwanomas were left sided (64%) however in our series right and left incidence were equal.

It often presents as slow growing solitary, asymptomatic cervical masses occurring within the upper carotid sheath or parapharyngeal space for long time. [6,7] Obstructive symptoms may occur if the tumour gains enough size to compress the upper aerodigestive tract. The pressure effect on the nerve of origin can also cause nerve dysfunction. If a major nerve is involved, nerve palsy, e.g. vocal cord palsy, Horner's syndrome, sensory or motor dysfunction of the upper limb may result.[8,9]

The diagnosis of schwannoma is suggested by the clinical features, e.g. long-standing history, firm mass which is more mobile along the plane perpendicular to the course of the nerve and nerve palsy.[10,11] FNAC and imaging studies are often ordered for diagnosis and planning of operation.[12] The specificity of imaging studies (i.e. CT or MRI) was 38%. The use of CT for diagnosis is not supported by the literature as there is no specific CT feature of schwannoma. Moreover, schwannoma may not be easily distinguished from paraganglioma in CT. Currently, MRI has superseded CT because of better soft tissue delineation. Studies on MRI also found the nerve of origin can be determined from the spatial relationship between the tumour and the great vessels of the neck. In MRI, the tumours are hypointense on T1-weighted images and heterogeneously hyperintense on T2-weighted images. The heterogeneity of intensity may be due to a mixture of haemorrhage, fibrosis and calcium deposits inside the tumour. The nerve of origin could be visualized in some cases with MRI but none with CT. On noncontrast CT, it was reported that schwannomas were typically hypodense versus muscle; with contrast, these lesions tended to show some peripheral enhancement. Miller et al. also reported that MRI was especially useful for the diagnosis with peripheral hyperintense rim with central low intensity on enhanced T1 images.[13]

Schwannomas are frequently difficult to characterize on FNAC. Liu et al.[14] reported that the accuracy of FNAC was only 20%. Our results also showed that of the three in which FNAC was done only one had diagnosis of Schwannoma.

The mainstay of treatment ideally remains complete surgical excision with preservation of the affected nerve. There was no recurrence for all the cases after complete surgical removal. If it is not possible to preserve the nerve of origin, the surgeon must be prepared to perform an end to end anastomosis or interposition nerve graft.[14]

Conclusion

In conclusion, cervical schwannomas are rare neck tumors. An accurate preoperative diagnosis with identification of the nerve of origin, therefore, allows safe resection with minimal loss of function,[15] and nerve sparing excision is the treatment of choice.

Conflict of interest: None declared

References

1. Colreavy MP, Lacy PD, Hughes J, Bouchier-Hayes D, Brennan P, O'Dwyer AJ, et al. Head and neck schwannomas—a 10 year review. J Laryngol Otol 2000;114:119-24.
2. Gavin CW, Khee-Chee S, Dennis TH. Extracranial non vestibularhead and neck scwannomas: A ten year experience. Ann Acad Med Singapore 2007; 36: 233-40
3. Torre V, Bucolo S, Galletti B, Fera G, Mangione AO, Suraci G, et al. Benign extracranial cervical-facial schwannomas: anatomo-clinical and diagnostic considerations on our case reports and review of literature. Acta Otorhinolaryngol Ital 1999;19:160-5.
4. Torossian JM, Beziat JL, AbouChebel N, Devouassoux-Shisheboran M, Fischer G. Extracranial cephalic schwannomas: a series of 15 patients. J Craniofac Surg 1999; 10:389-94.
5. Malone JP, Lee WJ, Levin RJ. Clinical characteristics and treatment outcome for nonvestibular schwannomas of the head and neck. Am J Otolaryngol 2005;26:108-12.
6. Sheridan MF, Yim DW. Cervical sympathetic schwannoma: a case report and review of the English literature. Otolaryngol Head Neck Surg1997;**117**:S206–S10
7. Rosner M, Fisher W, Mulligan L. Cervical sympathetic schwannoma: case report. Neurosurgery 2001;49:1452-4.
8. Gilmer-Hill HS, Kline DG. Neurogenic tumors of the cervical vagus nerve: report of four cases and review of the literature.Neurosurgery 2000;46:1498-503.
9. St Pierre S, Theriault R, Leclerc JE. Schwannomas of the vagus nerve in the head and neck. J Otolaryngol 1985;14:167-70.
10. Moukarbel RV, Sabri AN. Current management of head and neck schwannomas. Curr Opin Otolaryngol Head Neck Surg 2005;13:117-22.
11. Al-Ghamdi S, Black MJ, Lafond G. Extracranial head and neck schwannomas. J Otolaryngol 1992; 21:186-8.
12. Yu GH, Sack MJ, Baloch Z, Gupta PK. Difficulties in thefine needle aspiration (FNA) diagnosis of schwannoma. Cytopathology 1999;10:186–94
13. Miller FR, Wanamaker JR, Lavertu P, Wood BG. Magnetic resonance imaging and the management of parapharyngealspace tumors. Head Neck 1996;18:67–77
14. Liu R, Fagan P. Facial nerve schwannoma: surgical excision versus conservative management. Ann Otol Rhinol Laryngol 2001; 110:1025-9.
15. Myssiorek DJ, Silver CE, Valdes ME. Schwannoma of the cervical sympathetic chain. J Laryngol Otol 1988;102:962–5