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ORIGINAL ARTICLE

Extracranial Head and Neck Schwannomas: Our Experience

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Abstract Schwannomas are benign neoplasms of the peripheral nerves originating in the Schwann cells. They are rare and usually solitary, with clearly delimited capsules. They occur in the head and neck region in only 25 % of the cases, and may be associated with Von Recklinghausen's disease. Schwannomas are always a diagnostic dilemma as they are asymptomatic for long time and histopathology is the gold standard for diagnosis. The present study retrospectively analysed data of 4 patients with schwannomas and reviewed the literature on the subject. Retrospective study at ENT & Head and Neck Surgery Department of Navodaya Medical College, Raichur. Data of 4 patients between 2008 and 2014 were reviewed. The sites of cervical schwannomas and the intraoperative, histopathological and postoperative clinical status of these cases were studied. Diagnostic methods, type of surgery and associated nerve of origin (NOO) were evaluated. The patients' age ranged from 18 to 50 years. None of them had type I neurofibromatosis or Von Recklinghausen's disease. The nerves affected included the brachial plexus, vagus nerve, sympathetic chain and lingual nerve. The nerve of origin was identified based on intra-operative findings and post-operative neurological deficits. Tumour was removed by debulk operation with the preservation of NOO method. Schwannomas are generally benign, and rarely recur. An accurate preoperative workup with the identification of NOO is very important not only for a correct diagnosis, but also for surgical planning and informing the patient about the possible complications.

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Introduction

Schwannoma is a benign, slow growing encapsulated perineural tumour of neuro ectodermal derivation, originating from Schwann cells of the neural sheath of motor and sensitive peripheral nerves [1]. As a slowly growing benign tumour, it has been reported that 25–45 % of schwannomas were located in the extracranial head and neck region [2]. It involves the cranial nerves such as V, VII, X, XI, and XII or sympathetic and peripheral nerves [3]. Extracranial head and neck schwannoma is a challenging condition to the head and neck surgeons. For tumours arising from the major cranial nerves, excision of tumour with the division of the nerve of origin (NOO) renders lifelong morbidity to the patients. On the other hand, other nerve-preserving excision method, e.g. intracapsular enucleation, does not guarantee intact nerve function after surgery. Because of the substantial chance of nerve palsy after operation, obtaining an accurate preoperative diagnosis, and preferably, with the identification of the NOO is crucial in the management of the disease [4].

Objective

The present study aimed to describe our experience with 4 patients with head and neck schwannomas, the diagnostic methods used, the surgical decisions and the treatment outcome.

Materials and Methods

Four cases of benign masses in the head and neck that were confirmed by postoperative histopathological examination at the ENT & Head and Neck Surgery Department of Navodaya Medical College, Raichur between 2008 and 2014 are described in our study.

The inclusion criteria for this study were the patients who had their postoperative histological diagnosis confirmed to be schwannoma. Pre-operative diagnostic tools included Fine Needle Aspiration Cytology, Contrast CT or MRI scan. The treatment of choice was debulking of the tumour with preservation of the nerve of origin. Postoperatively, the data on surgical complications and nerve deficits caused by tumour resection were evaluated. A review of the literature available on the topic was also carried out.

Case 1

A 28 year old male presented with painless swelling in left submandibular region, of 1 year duration. There was no numbness of tongue or weakness in movements of the tongue. Examination revealed a solitary non-tender swelling measuring 3 cm × 3 cm. FNAC was non-diagnostic. Contrast enhanced computed tomographic scan revealed an isodense mass in left submandibular space with nonhomogeneous enhancement measuring 3 cm × 3 cm. MRI was not done due to non-affordability of the patient. Surgery was done by transcervical approach. The tumour was found to be well encapsulated and was found to be attached to lingual nerve. The tumour was excised by debulk operation with the preservation of NOO method. Postoperatively, there was no neurological deficit involving the lingual nerve. Microscopic sections of the tumour areas revealed hypocellular and hypercellular areas with interspread thick and thin walled blood vessels. The hypercellular areas show plump spindle-shaped tumour cells arranged in interlacing fascicles, whorls and palisading pattern with formation of verocay bodies (Figs. 1, 2). Impression was of a schwannoma. The patient was followed up for 2 years with no signs of recurrence.

Case 2

A 18-year-old male presented with complaints of asymptomatic swelling in the left side of the neck since 4 years (Fig. 3). Examination revealed a diffuse swelling in the left side of the neck in carotid triangle. Throat examination revealed a visible bulge in the left tonsil and posterior pillar. Left lateral pharyngeal wall was displaced medially. On indirect laryngoscopy, both vocal cords were mobile.

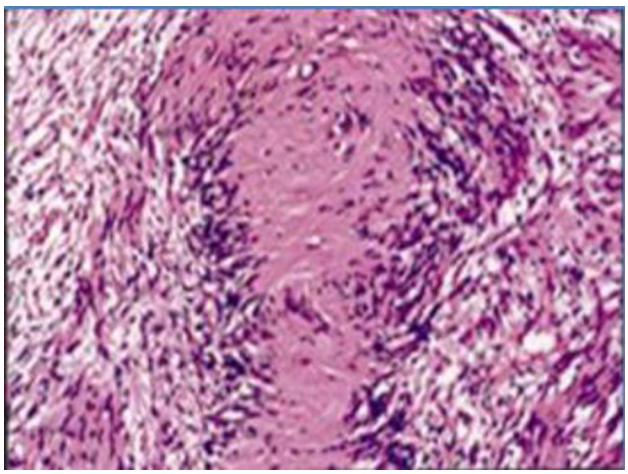

Fig. 1 Verocay body with palisaded rows on either sides (H&E stain, ×40)

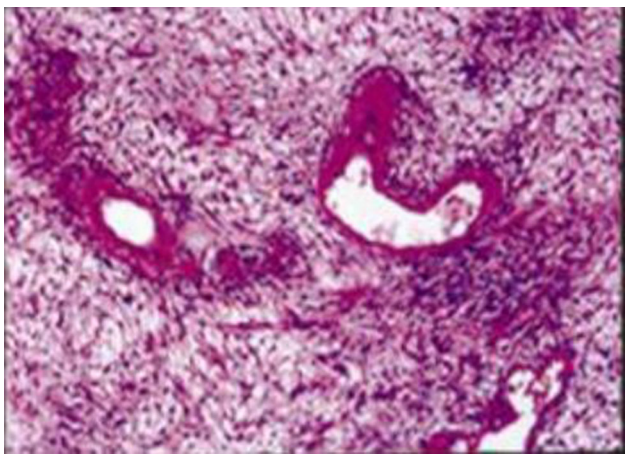

Fig. 2 Hypocellular areas-loose stroma with spaced spindle cells (H&E stain, ×40)

Cranial nerve examination was normal. A provisional diagnosis of left parapharyngeal mass was made. Contrast enhanced computed tomographic (CECT) scan revealed an isodense mass in left carotid space with nonhomogeneous enhancement measuring 7.5 × 4.8 cm with central hypodense area pushing the carotid artery anteriorly and compressing the jugular vein with dilatation of the vein below the mass. The lesion is pushing medially the glottis and the lateral pharynx with mild compression of trachea (Fig. 4). Ultrasound guided fine needle aspiration cytology (FNAC) showed few clusters of oval to spindle cells having bland nuclear chromatin in myxoid stroma against a haemorrhagic background. Magnetic resonance (MR) angiography was done which showed a well-defined space occupying lesion in the left carotid space displacing left common carotid artery and carotid artery bifurcation anteriorly and internal jugular vein anterolaterally suggestive of vagal

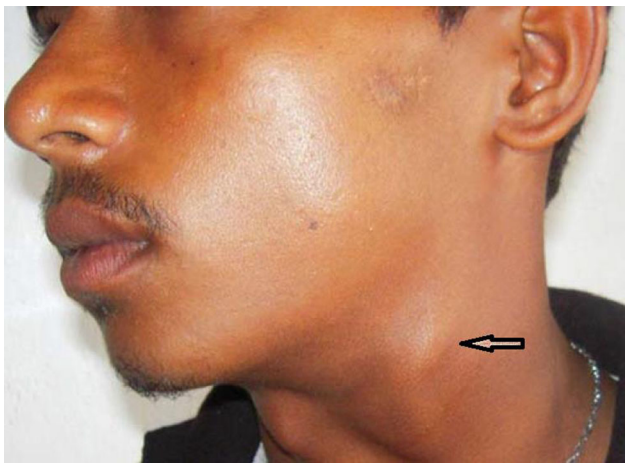

Fig. 3 Pre-operative photograph of the patient with *arrow* pointing at vagal schwannoma

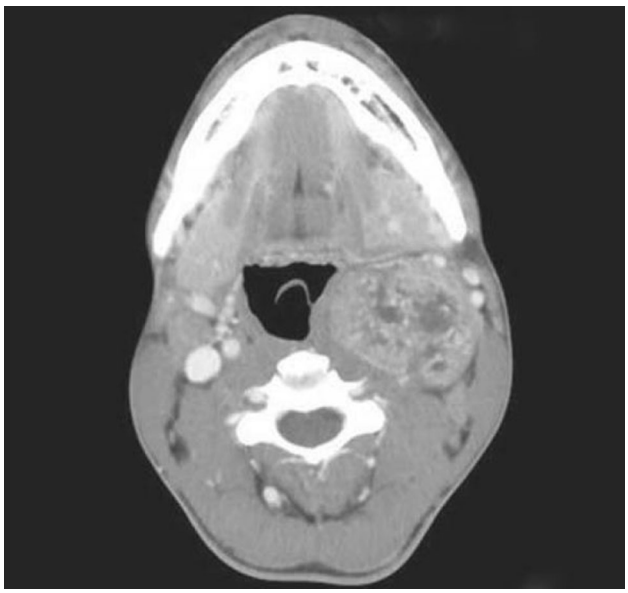

Fig. 4 CECT neck showing the parapharyngeal mass occupying the poststyloid compartment in the vagal schwannoma patient

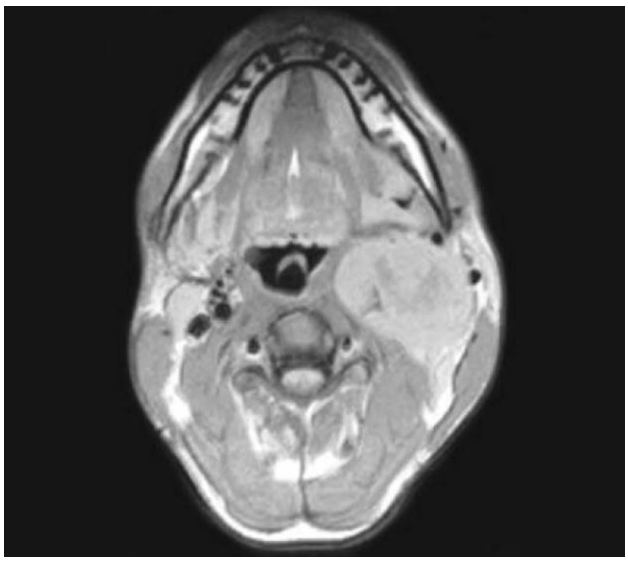

Fig. 5 MR angiography T1-weighted axial section image showing vagal schwannoma occupying left carotid space

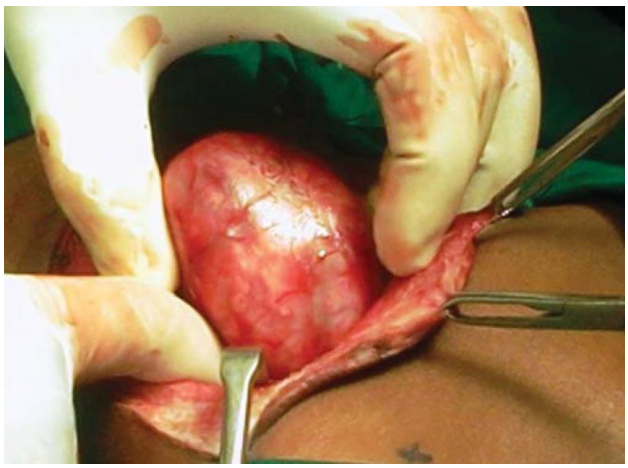

Fig. 6 Intraoperative photograph showing the vagal schwannoma being exposed

schwannoma (Fig. 5). Twenty-four hours urinary vanilyl mandelic acid assay found to be within normal range. Patient was taken up for resection of the mass through a transcervical approach. A curvilinear incision made from behind the angle of the mandible to cricoid cartilage. Dissection was made in subplatysmal plane. Left sternocleidomastoid was retracted and carotid sheath was opened. A small nerve twig was attached to the tumour in its inferior aspects. The tumour was found to be well-encapsulated and was excised by debulk operation with the preservation of NOO method (Fig. 6). Postoperatively, patient was found to have left sided vocal cord paralysis. Grossly, the tumour measured 7.5 × 4.5 × 4 cm. Microscopic sections of the tumour areas revealed features of a schwannoma. Patient was followed up for 2 years and there was no sign of recurrence of the tumour. The left sided vocal cord paralysis did not improve even after 2 years.

Case 3

A 20 year old female presented as a painless slow growing swelling since 2 years in the left side of neck, with no associated symptoms like dysphagia or dysphonia. Local examination revealed a 4 × 3 cm swelling on left side of

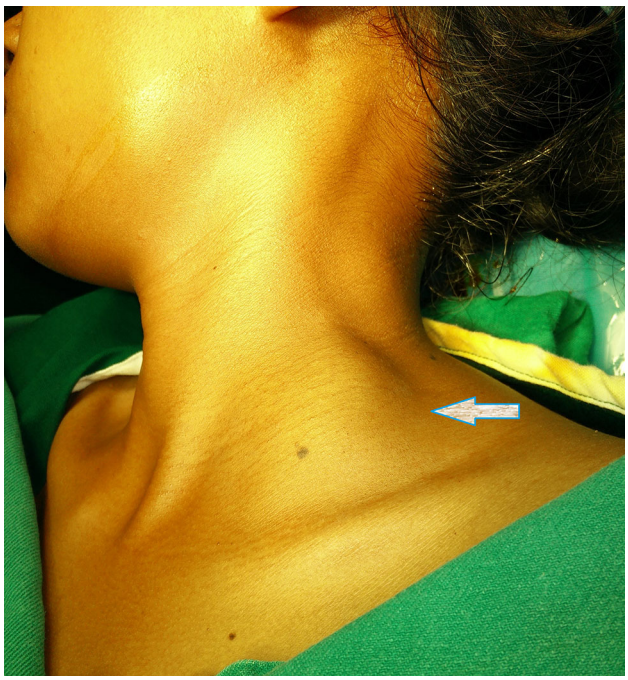

Fig. 7 Pre-operative photograph of the patient with *arrow* pointing at axillary nerve schwannoma

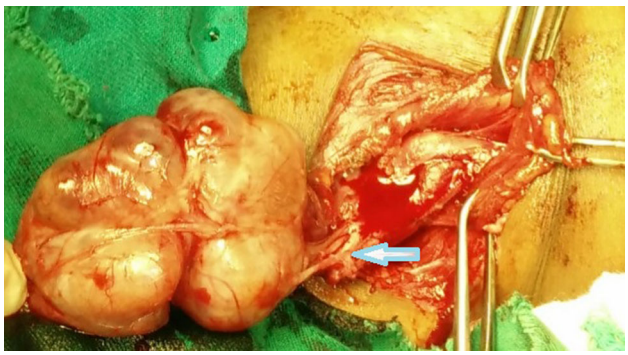

Fig. 8 Axillary nerve schwannoma before resection, with tumour having attachment to a branch of posterior cord of brachial plexus (*arrow*)

the neck posterior to the lower third of the sternocleidomastoid in the supraclavicular region (Fig. 7), well circumscribed and was firm in consistency. It was mobile over the deeper soft tissues in an anteroposterior plane. Skin over the mass appeared normal. Ultrasonography was done and reported as enlarged supraclavicular lymph node. Fine needle aspiration cytology report was necrotizing lymphadenitis. Contrast enhanced computed tomographic scan revealed an isodense mass in left supraclavicular region with nonhomogeneous enhancement measuring 4 cm × 3 cm. MRI was not done due to no-affordability by the patient. Brachial plexus examination revealed no neurological deficits. Debulk operation with the preservation of NOO was done through a transcervical approach. A branch of posterior cord of brachial plexus was seen attached to the tumour (Fig. 8). Histopathological examination of the tumour was suggestive of schwannoma. Postoperatively, patient developed left sided deltoid muscle weakness. Nerve conduction studies reported as weakness of the muscles supplied by axillary nerve. Patient received regular physiotherapy. The left sided deltoid muscle weakness improved significantly by the end of 3rd month. Patient was followed up for 2 years and there was no sign of recurrence of the tumour.

Case 4

A 50 year old man, presented with an asymptomatic swelling in the right upper lateral neck since past 5 years. On examination, there was a solitary, non-tender, non-pulsatile, firm mass in the right carotid triangle area measuring about 6 cm × 4 cm with margins well defined all around (Fig. 9). Cranial nerve examination was normal. Fine needle aspiration cytology study was non-diagnostic. Contrast enhanced Computed Tomographic scan revealed an isodense mass in the right carotid space with non-homogenous enhancement measuring 7.2 × 5.5 × 3.1 cm with central hypodense area pushing the carotid artery anteromedially and compressing the internal jugular vein with dilatation of proximal part of vein. CECT suggested of a cystic nerve sheath tumour (Fig. 10). Magnetic resonance (MR) angiography was done which showed a well-defined space occupying lesion in the right carotid space displacing right common carotid artery and carotid artery bifurcation anteriorly and internal jugular vein anterolaterally suggestive of vagal schwannoma. Patient was taken up for debulk operation with the preservation of

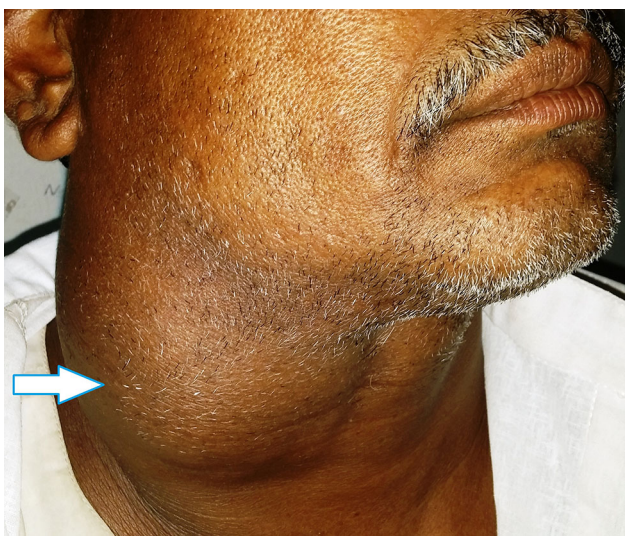

Fig. 9 Pre-operative photograph of the patient with *arrow* pointing at cervical sympathetic chain schwannoma

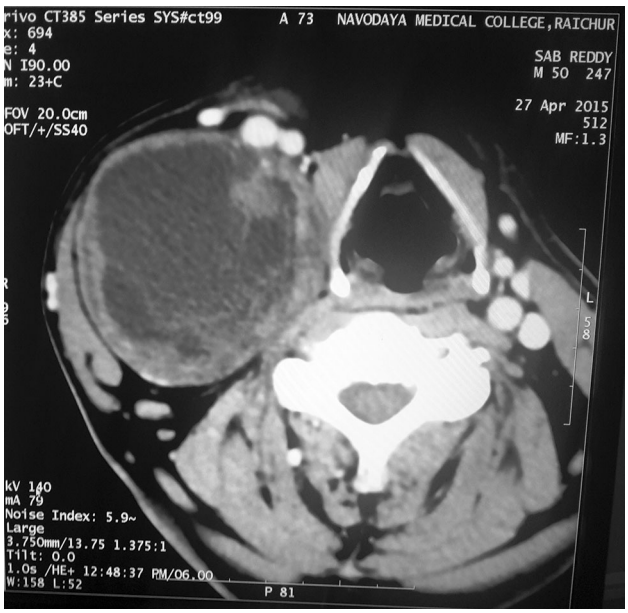

Fig. 10 Contrast enhanced computed tomography scan shows isodense mass in right carotid space with non homogenous enhancement

NOO through a transcervical approach. Right sternocleidomastoid was retracted and carotid sheath was opened. A small nerve twig was attached to the tumour in its inferior aspects and vagal nerve was found to lie separate from the tumour and was intact. The tumour was found to be well-encapsulated and was excised by debulking with preservation of NOO method. Postoperatively, patient developed mild features of Horner's syndrome, which took 6 months' time to improve. Histopathological study was suggestive of a schwannoma. Since the patient developed mild features of Horners' syndrome, the tumour was probably arising from the cervical sympathetic chain. Patient was followed up for 2 years and there was no sign of recurrence of the tumour.

Discussion

Schwannoma was first identified by Virchow in 1908 and later reported by Verocay in 1910 [5]. Since that time, they have been called neurilemmomas, solitary nerve sheath tumours, perineural fibroblast tumours and, most recently, schwannomas, according to the World Health Organization (WHO) classification [6]. Schwannomas are uncommon nerve sheath neoplasms that may originate from any peripheral, cranial or autonomic nerves of the body with the exception of the olfactory and the optic nerves [7]. In head and neck region they most commonly arise from the parapharyngeal space. The most common nerves of origin are the vagus and the cervical sympathetic chain [8].

Neurogenic tumours of head and neck arise from neural crest cells which differentiate into nerve sheath cells and sympathoblast. Schwann cell is the parent cell of both schwannomas and neurofibroma. Neurofibroma also has an origin from the perineurium and is thus linked inseparably from the nerve of origin [9]. Schwannomas grow longitudinally along the length of the nerve assuming a fusiform appearance without compromising the morphological and functional integrity of the nerve and can be separated surgically from their nerve of origin. Distinction between schwannomas and neurofibroma can be made microscopically [10].

Symptomatology

Schwannomas are almost always diagnostic problems because their history and clinical examination are nonspecific and deceptive [7]. According to the literature [11–14], schwannomas are equally distributed between the genders, however, a more prevalence in males (3:1) was found by the present study. In terms of age, the greatest incidence in our study was in second decade. But other authors have described it between the third and fifth decades [14–18].

In cases unassociated with neurofibromatosis, schwannomas are seen clinically as solitary slow-developing lesions that show symptoms only when large areas have been affected [19]. A similar study by Liu et al. [4] also reveals that most of the tumours (60 %) presented themselves as an asymptomatic palpable mass. In our study also, the patients came to us as asymptomatic palpable mass in the neck.

Pre-operative Investigations

Fine-needle aspiration biopsy has drawbacks in terms of accuracy, as described by Zbaren and Becker [17], and does not constitute an effective means of preoperative diagnosis. In a similar study by Liu et al. [4], the results of FNAC were inconclusive in 50 % of cases, suggested the diagnosis of schwannoma in 20 % of cases and suggested a diagnosis other than schwannoma in 10 % of their cases. In our study, FNAC was non-diagnostic in 3 (75 %) of our cases and suggested a diagnosis other than schwannoma in 1 (25 %) case. Ultrasonography was done in 2 of our cases, but it was non-diagnostic again. On noncontrast CT, it was reported that schwannomas were typically hypodense versus muscle; with contrast, these lesions tended to show some peripheral enhancement [20]. No case (0 %) in our study was able to suggest the diagnosis of schwannoma by CT and clinical features. On the other hand, MRI consistently identifies these lesions on both T1- and T2-weighted imaging. T1-weighted images display low signal intensity,

and T2-weighted images show high intensity [20–22]. Hirano et al. [23] also reported that MRI was especially useful for the diagnosis of schwannomas. The relationship between the schwannoma and its nerve of origin can be better appreciated with MRI than CT. In addition, MRI appears to be the investigation of choice for diagnosis and identification of nerve of origin (NOO). In a study by Ryuji et al. [24], twenty cases (80 %) suggested the diagnosis of schwannoma by MRI investigation. These results indicate that MRI is most sensitive and specific in the diagnosis of schwannoma [21]. We cannot comment on the significance of MRI as it was not done in 2 of our cases due to non-affordability by the patient. In the remaining two cases, MRI was diagnostic of Schwannoma, but the Nerve of Origin was accurately localized in only one case.

Surgery

Most of the head and neck schwannomas can be resected by transcervical approach. The different techniques include Maxillary swing, combined craniofacial approaches, intracapsular enucleation and debulk operation with the preservation of NOO [4]. The preferred method of removing a schwannoma is intracapsular enucleation. Complications are usually transient and in most cases do not require treatment [24]. According to the study by Valentino et al., intracapsular enucleation while preserving the nerve fibers preserved its function by more than 30 % when compared to complete tumour resection [14]. In the study by Liu et al. [4], the rates of nerve palsy after operation were 100, 67 and 50 % for resection with division of NOO, intracapsular enucleation and debulk operation, respectively. All our cases were operated by debulk operation with the preservation of NOO. However, we noticed neurological deficit during post-operative period in 3 (75 %) cases. Two of these three cases recovered completely by 6 months while one case did not improve even by the end of 2 years follow up.

Nerve of Origin

The low incidence of schwannomas is also reflected when the anatomical sites involved are considered. In the case of the brachial plexus, 147 cases had been published up to 1987 [18]. The cranial pairs most affected by neurilemmomas are the ninth, seventh, eleventh, fifth and fourth, in order of frequency [25]. Up to 1989 approximately 70 cases of the vagus nerve schwannomas had been described in the literature [13]; the first was described by Sekiguichi in 1926. Schwannomas of the sympathetic chain are even rarer and only 14 cases had been described up to 1997 [15]. Other infrequent sites include the lingual and recurrent laryngeal nerves, cervical plexus, nasal and paranasal regions [26], facial nerve [26], posterior pharyngeal wall [27], larynx [12], thyroid gland [28] and other regions.

In our study, the NOO was mainly determined by the postoperative neurological deficit and suggested by the intraoperative findings. According to Liu et al. [4], the concordance rate of the identification of the NOO from intraoperative findings and postoperative neurological deficit was 87 %, which means that the NOOs determined by the intraoperative findings was the same as those determined by the postoperative neurological deficit. In our case, the rate was 75 %. In our study, there was one case each involving lingual nerve, vagus nerve, cervical sympathetic chain and axillary nerve.

Long Term Outcome

We followed up our cases for 2 years and there was no local recurrence. However, in the study by Liu et al. [4], in one patient who had debulk operation, follow-up MRI showed slow progression of the tumour.

Both the incidence of malignant schwannoma and the rate of malignant transformation from benign schwannoma are not available in the literature [4]. A review on intracranial malignant peripheral nerve sheath tumour (MPNST) suggested that most of the MPNSTs are developed sporadically rather than transforming from benign tumours [29]. Current knowledge also showed that most MPNSTs are associated with neurofibromatosis type 1 [30, 31]. From these circumferential evidences, the rate of malignant transformation can be considered low in patients with solitary extracranial head and neck schwannoma. Because of the indolent nature and the remote chance of malignant transformation, an observational approach is often possible. The decision of operation should, therefore, be based on the balance between the risk and benefit of the surgery, i.e. the severity of preoperative symptomatology and the anticipated postoperative neurological deficit [4].

The operation of schwannoma arising from major nerve can render lifelong morbidity to the patients. An accurate preoperative diagnosis with the identification of the NOO, therefore, allows patients to make an informed decision on whether to undergo operation or observation. Also, the treatment of postoperative nerve palsy should be regarded as a part of the management of schwannoma. As the sympathetic trunk and the vagus nerve are the common NOOs, corrective procedures, e.g. levator operation and medialization of vocal cord should be explained to the patients before operation [4].

Conclusion

Schwannomas are derived from Schwann cells. They are usually solitary lesions, except in Von Recklinghausen's disease. Pain and neurological symptoms are uncommon, and become evident only at advanced stages. They are generally benign, and rarely recur. An accurate preoperative workup with the identification of NOO is very important not only for a correct diagnosis, but also for surgical planning and informing the patient about the possible complications. This allows the patient to make an informed decision on whether to undergo operation or observation.

Compliance with Ethical Standards

Conflict of interest None.

Ethical Approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed Consent Informed consent was obtained from all individual participants included in the study.

References

1. Kumar AB, Rajan P (2004) Schwannoma of the tongue- a case report. Calicut Med J 2(2):e4
2. Ducatman BS, Scheithauer BW, Piepgras DG, Reiman HM, Ilstrup DM (1986) Malignant peripheral nerve sheath tumours: a clinicopathologic study of 120 cases. Cancer 57(10):2006–2021
3. Colreavy MP, Lacy PD, Hughes J et al (2000) Head and neck schwannomas—a 10 year review. J Laryngol Otol 114(2):119–124
4. Liu H-L et al (2011) Extracranial head and neck schwannomas: a study of the nerve of origin. Eur Arch Otorhinolaryngol 268:1343–1347
5. Katti TV, Anantharao AS, Punyashetty KB (2014) Lingual schwannoma in posterior 1/3 of tongue—a rare presentation. J Evid Based Med Healthc 1(7):529–532
6. Jain S, Houseknecht K, Rojiani AM (2008) Management of nerve sheath tumours arising in the sympathetic chain. Cancer Control 15(4):352–357
7. Gavin CW, Khee-Chee S, Dennis TH (2007) Extracranial non vestibular head and neck schwannomas: a 10 years' experience. Ann Acad Med Singap 36(4):233–240
8. Bhandary SK, Vaidyanathan V, Bhat V, Biniyam K, Shenoy S (2011) Neck schwannoma masquerading as a carotid body tumour. J Clin Diagn Res 5(3):654–656. Available from http://www.jcdr.net/back_issues
9. Watkinson JC, Gaze MN, Wilson JA (eds) (2000) Benign neck disease. In: Stell and Maran's head and neck surgery, 4th edn. Butterworth Heinemann, Oxford, pp 193–194
10. Awasti SK, Dutta A (2011) Cervical sympathetic chain schwannomas: a case report. Indian J Otolaryngol Head Neck Surg 63(3):292–294
11. Feany MB, Anthony DC, Fletcher CD (1998) Nerve sheath tumours with hybrid features of neurofibroma and schwannoma: a conceptual challenge. Histopathology 32(5):405–410
12. Ingels K, Vermeesch H, Verhoye C, De Potter C (1996) Schwannoma of the larynx: a case report. J Laryngol Otol 110(3):294–296
13. Martins AS, Tincani AJ, Velloso LA (1990) Schwannomas de nervo vago: revisão da literatura e relato de três casos [Schwannoma of the vagus nerve: presentation of three cases and literature review]. Rev Col Bras Cir 17(6):145–148
14. Valentino J, Boggess MA, Ellis JL, Hester TO, Jones RO (1998) Expected neurologic outcomes for surgical treatment of cervical neurilemmomas. Laryngoscope 108(7):1009–1013
15. Ganesan S, Harar RP, Owen RA, Dawkins RC, Prior AJ (1997) Horner's syndrome: a rare presentation of cervical sympathetic chain schwannoma. J Laryngol Otol 111(5):493–495
16. Sharaki MM, Talaat M, Hamam SM (1982) Schwannoma of the neck. Clin Otolaryngol Allied Sci 7(4):245–251
17. Zbaren P, Becker M (1996) Schwannoma of the brachial plexus. Ann Otol Rhinol Laryngol 105(9):748–750
18. Seppälä MT, Sainio MA, Haltia MJ, Kinnunen JJ, Setala KH, Jaaskelainen JE (1998) Multiple schwannomas: Schwannomatosis or neurofibromatosis type 2? J Neurosurg 89(1):36–41
19. Langner Erwin et al (2007) Schwannomas in the head and neck: retrospective analysis of 21 patients and review of the literature. Sao Paulo Med J 125(4):220–222
20. Anil G, Tan TY (2010) Imaging characteristics of schwannoma of the cervical sympathetic chain: a review of 12 cases. Am J Neuroradiol 31(8):1408–1412
21. Kami YN, Chikui T, Okamura K et al (2012) Imaging findings of neurogenic tumours in the head and neck region. Dentomaxillofacial Radiol 41:18–23
22. Tomita T, Ozawa H, Sakamoto K, Ogawa K, Kameyama K, Fujii M (2009) Diagnosis and management of cervical sympathetic chain schwannoma: a review of 9 cases. Acta Otolaryngol 129(3):324–329
23. Hirano S, Kitamura H, Miyata K et al (1994) Extracranial neurinomas of head and neck. Jibiinkouka Rinsyo 87:253–257 **(Japanese)**
24. Ryuji Y et al (2013) Diagnosis and management of extracranial head and neck schwannomas: a review of 27 cases. Int J Otolaryngol 1–5. doi:10.1155/2013/973045
25. MacCollin M, Woodfin W, Kronn D, Short MP (1996) Schwannomatosis: a clinical and pathological study. Neurology 46(4):1072–1079
26. Hajjaj M, Linthicum FH Jr (1996) Facial nerve schwannoma: nerve fibre dissemination. J Laryngol Otol 110(7):632–633
27. Haraguchi H, Ohgaki T, Hentona H, Komatsuzaki A (1996) Schwannoma of the posterior pharyngeal wall: a case report. J Laryngol Otol 110(2):170–171
28. Sugita R, Nomura T, Yuda F (1998) Primary schwannoma of the thyroid gland: CT findings. AJR Am J Roentgenol 171(2):528–529
29. Ziadi A, Saliba I (2010) Malignant peripheral nerve sheath tumour of intracranial nerves: a case series review. Auris Nasus Larynx 37:539–545
30. Tucker T, Wolkenstein P, Revuz J, Zeller J, Friedman JM (2005) Association between benign and malignant peripheral nerve sheath tumours in NF1. Neurology 65:205–211
31. Pasmant E, Masliah-Planchon J, Lévy P, Laurendeau I, Ortonne N, Parfait B, Valeyrie-Allanore L, Leroy K, Wolkenstein P, Vidaud M, Vidaud D, Bièche I (2011) Identification of genes potentially involved in the increased risk of malignancy in NF1-microdeleted patients. Mol Med 17(1–2): 79–87.