Research Paper

Surgical management of extracranial nerve sheath tumours in a tertiary care center

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KEYWORDS
Nerve sheath tumours; Surgical management; Outcome

Abstract  Objective: Excision of the nerve sheath tumours of head and neck is always a challenge to the surgeon because of their close proximity of the neurovascular structures.

Methods: It is a retrospective study contained 13 consecutive patients of nerve sheath tumours involving the head and neck from March 2013 to February 2017 in the department of Otorhinolaryngology and Head Neck Surgery in a tertiary care referral hospital. The retrospective clinical data, diagnostic procedures, surgical approaches and their complications are analyzed after 12 months of surgery.

Results: Total 13 patients, 7 were females and 6 were males. Painless neck swelling was the most common presentation found in 6 and the most common site of origin was the vagus nerve in the parapharyngeal space (6 patients). Transcervical excision of the tumour was performed in 7 patients. Six needed combined surgical approach for the excision of the tumours. Neurovascular injury was detected in 3 patients, and one patient had recurrent disease during the follow-up period.

Conclusions: Excision of the nerve sheath tumours of head and neck is always a challenge to the surgeon because of their close proximity of the neurovascular structure. Although the transcervical approach is commonly practiced for the majority of the tumours, combined approaches can be effectively applied for extensive nerve sheath tumours with the satisfactory clinical outcome.

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Introduction

Nerve sheath tumour including Schwannomas and neurofibroma are the most common benign neoplasm derived from the nerve sheath of both the central and the peripheral nerves. Although they appear throughout the body, head and neck are the most common sites get affected by the tumours, in spite of the fact that schwannoma is more prevalent than the neurofibroma as described by the literature. In the neck, they usually originate from the cranial nerves in the post styloid compartment of the parapharyngeal space (PPS). Schwannomas are solitary, slow growing, painless mass and usually asymptomatic at their initial presentation unless it compresses the adjacent structures. In contrast, neurofibromas are non-encapsulated, soft, pedunculated tumour more often found at the peripheral parts in the body. Later can present as solitary swelling or generalized neurofibromatosis associated with von Recklinghausen’s disease. Incidence of malignant peripheral nerve sheath tumour (MPNST) is very rare and approximately 12% of cases found to be associated with von Recklinghausen’s neurofibromatosis. Clinical presentation depend upon the size and extension of the tumour in different subsites in the head and neck, including tongue, palate, nose, paranasal sinus and anterior skull base. Patients with MPNST can present with rapidly progressive painful lesions and these tumours are usually associated with the worst prognosis. Contrast-enhanced Magnetic resonance imaging (MRI) is the preferred radiological investigation advised for the patients of nerve sheath tumours. T1-weighted MRI reveals an intermediate intensity and in T2-weighted, these tumours appear as a hyperintensity lesions. Besides providing the valuable information regarding the size and extension of the tumour, MRI is very much helpful in distinguishing the malignant from a benign lesion and to evaluate their relationship to the vital neurovascular structures for a proper preoperative planning.

Fine needle aspiration cytology (FNAC) is the definitive investigation practiced in patients suspicious nerve sheath tumour performed through the transcervical route or intraoral route except for the highly vascular tumours. Treatment is mostly by the surgical excision of the tumour and later primarily depends on the proximity of the lesion to the neurovascular structure and the expected postoperative morbidity attributed to surgery. It can be undertaken by a simple transcervical or combined transcervical transparotid in the majority of the cases. Transcervical transmandibular or transcervical transoral approaches can be utilized in selected cases of extensive and recurrent tumours where wider surgical exposure is required.

With the advancement of the endoscopic sinus surgeries, tumours involving the trigeminal nerve and superior laryngeal nerve can be successfully excised with more conservative surgeries. In the present study, we have shared our experience in the surgical management of the nerve sheath tumours of the head and neck in a tertiary care referral center.

Material and method

It is a retrospective study contained 13 patients of nerve sheath tumours involving the head and neck. The study duration was from March 2013 to February 2017 in the Department of Otorhinolaryngology and Head Neck Surgery, All India Institute of Medical Sciences, Bhubaneswar, a tertiary care apex hospital in the eastern zone of India. All diagnosed cases of head-neck neurofibromas/Schwannomas were included in the study. Tumours originating from the peripheral nerves and the eighth cranial nerves were excluded from the study. The medical record was reviewed and the clinical, radiological, and pathological data were analyzed retrogradely. In the clinical data, the symptoms, signs, diagnostic procedures, surgical approaches, and significant intraoperative/postoperative complications were noted. The preoperative MRI findings were analyzed for the size, location, vascularity, and the extension of the tumour. Data obtained were statistically analyzed with SPSS statistics 22 (IBM, Chicago, USA). The average follow-up period was 12 months (range 11–18 months). Patients were reviewed in the tumour clinic in the department at the end of 3, 6, and 12 months period where the clinical and radiological assessment was undertaken to rule out the disease recurrence.

Results

Total 13 patients were included in the study of which 7 were females and 6 were males with age varied from 13 to 52 years [mean (31.91 ± 10.94) years]. The demographic data and the clinical profile of the patients have been demonstrated in the Table 1. Painless neck swelling was the predominant complaint which was observed in 6 patients. Two patients presented with parotid swelling, two had oropharyngeal swelling and one presented with a submandibular swelling. One patient had hoarseness during the initial presentation and one had a cheek swelling (left). Contrast-enhanced MRI was performed in all cases and angiography was done in 2 cases, one of them was a recurrent neurofibroma and another was a vagal schwannoma compressing and splaying of the both carotid arteries at the initial radiological examination. None of the patients had undergone preoperative embolization as there was no definitive feeding vessel identified. Considering the site of origin, 6 patients were found to originate from the vagus nerve in the PPS. The spinal accessory nerve, trigeminal nerve, superior
Table 1  

| No. | Age/sex | Clinical feature | Site of origin | Tumour size (cm) | Surgical approach | Final diagnosis | Complications |
|-----|---------|------------------|----------------|-----------------|------------------|----------------|---------------|
| 1   | 40/M    | Neck swelling    | Left vagus     | 3.3 x 2.8       | Transcervical     | Schwannoma      | None          |
| 2   | 23/F    | Neck swelling    | Right vagus    | 4.5 x 3.4       | Transcervical     | Schwannoma      | None          |
| 3   | 28/M    | Neck swelling    | Right spinal nerve | 4.9 x 3.0   | Transcervical     | Schwannoma      | None          |
| 4   | 31/F    | Parotid swelling | Right vagus    | 5.2 x 7.0       | Transcervical     | Schwannoma      | None          |
| 5   | 18/M    | Neck swelling    | Right vagus    | 5.6 x 4.2       | Transcervical     | Schwannoma      | Marginal mandibular palsy |
| 6   | 47/M    | Lateral pharyngeal bulge | Left sympathetic chain | 4.7 x 3.3 | Transcervical | Schwannoma | None |
| 7   | 52/M    | Neck swelling    | Right vagus    | 5.2 x 4.7       | Transcervical     | Schwannoma      | Direct laryngoscopic injury |
| 8   | 28/F    | Neck swelling    | Left spinal accessory nerve | 4.8 x 4.0 | Endoscopic with transcervical | Schwannoma | None |
| 9   | 38/F    | Parotid swelling | Right superior laryngeal nerve | 1.0 x 1.2 | Endoscopic with transcervical | Schwannoma | None |
| 10  | 28/F    | Neck swelling    | Right brachial plexus | 1.0 x 1.2 | Endoscopic with transcervical | Schwannoma | None |
| 11  | 13/M    | Neck swelling    | Left trigeminal nerve | 5.4 x 4.8 | Endoscopic with transcervical | Schwannoma | None |
| 12  | 37/F    | Cheek swelling   | None           |                 |                  |                |               |
| 13  | 14/F    | Neck swelling    | None           |                 |                  |                |               |

As observed in the present study, the majority of the tumours were schwannom. The mean age of presentation

**Discussion**

Nerve sheath tumours originate from the neural crest from the Schwann cell or sympathoblast, and the specialized neuromesenchymal cells from the neural crest give rise to schwannoma and neurofibroma. They can arise from any of the cranial/spinal nerve in the body except the olfactory and optic nerve as they don’t have the Schwann cells. Schwannomas exclusively arise from the Schwann cells and neurofibromas can have multiple tissues of origin including Schwann cells, perineural cells, and perineural fibroblasts. Although these tumours can affect the various sites in the body, involvement of head and neck require special concern as the majority of the tumours arise from the lower cranial nerves where excision of the tumour is a challenge because of its close proximity to the neurovascular structure. Here we have discussed the clinical findings, diagnosis, treatment, and clinical outcome of the nerve sheath tumours with the different surgical approaches and their respective complications.
was 32 years with a little female predominance (7/13) in contrast to the previous authors where they did not find any preference to any age or sex. Neurofibromas usually present as solitary/multiple lesions involving mostly the peripheral nerves and very rarely the deeper neural structure gets involved. Later is a slow growing, non-encapsulated mass and in 10% cases, it is found to be associated with neurofibromatosis due to the somatic mutation in the NF1 gene located in the chromosome 17. In the neck, the PPS was the predominant site for the nerve sheath tumour accounting for 61.53% of the total patients. In the PPS, all of the tumours were detected in the poststyloid compartment and the majority of the tumours were arising from the vagus nerve and one was arising from the spinal accessory and the other from the cervical sympathetic chain. Of the 13 patients, 5 had very unusual location including the spinal nerve, vallecula, superior laryngeal nerve, brachial plexus, and the trigeminal nerve, affecting one case each. This variation in the incidence of the tumour could be due to the retrospective clinical data. These are very slow growing and painless tumours and often get unnoticed by the patient unless these cause significant pressure symptoms to the adjacent neurovascular structure. Although neck swelling is the most common presentation, patients can have a wide range of symptoms, including swelling in the oral cavity, dysphagia, vocal fold paralysis, and Horner syndrome although the later symptoms are very rare to occur. As described in the present study, 6 patients had painless neck swelling, one patient had Horner’s syndrome and one had hoarseness at the initial presentation. MRI is the preferred over CT scan for radiological assessment of the tumour because it defines superior tissue abnormality and provides adequate information of the extension, invasion, and regional metastasis, required for proper surgical planning. MRI (T1 weighted) of the schwannomas usually reveals a spherical to ovoid homogenous contrast-enhanced soft tissue mass and patients with long standing disease can have non-homogeneous enhancement of the lesion because of the presence of cystic/fatty degeneration. Although MRI plays a vital role in the diagnosis of the disease, the specificity of the later is not very high and site of the origin is not always possible. Hence it is mandatory to have a proper clinicoradiological correlation for the accurate diagnosis of the lesion. Angiography is not routinely advised as the primary investigation for the evaluation of nerve sheath tumours, as described in the present study, one with significant splaying of carotid arteries and another with a recurrent neurofibroma required preoperative angiography to rule out the carotid body tumour as a common differential diagnosis at the same location and to assess the requirement of embolization. FNAC is considered the most definitive investigation advised for almost all patients suspicious for nerve sheath tumour either through an intraoral route or transcervical route. Of 13 patients, FNAC was performed in 10 patients and the diagnostic accuracy was 80%.
One patient was operated for the recurrent neurofibroma was confirmed to be a malignant nerve sheath tumour and another was diagnosed as chronic sialoadenitis in the preoperative period was confirmed to be a schwannoma in the final pathological report. Although cytology is considered as the cornerstone for the diagnosis of the most of the nerve sheath tumours, it is not always conclusive. 23,24 Surgical excision is considered the primary modality of treatment for the nerve sheath tumours. Surgical approach mostly depends upon the size and extension of the tumour in the different anatomical subsites of the head and neck and their proximity to the neurovascular structures especially the tumours in the post styloid compartment. The majority of the PPS nerve sheath tumours can be removed through the transcervical approach as observed in the present study where complete excision of the tumour was achieved in 7 cases. This approach was very much suitable for the cervical nerve sheath tumours with limited oropharyngeal extension. Patients with the extensive extension of tumours may require combined surgical approaches as seen in the present study where two cases have undergone transcervical transtemporal approach and one had undergone transcervical transmandibular through the mandibular swing. Combined intranarial endoscopic and transcervical approach was performed for an extensive infratemporal schwannoma compressing the optic nerve and extending to the cavernous sinus in the left side.

Although combined surgical approaches are very rarely performed in the routine surgical practice, still these are adopted for extensive tumours where more exposure is required than the standard transcervical/transcervical transtemporal approach. 25-27 Endoscopic removal of tumours was successfully achieved in two cases, one was a vallecular schwannoma and other was involving the supraglottis. Complete excision of tumours often ensured in cases of schwannomas because these are well encapsulated tumours in contrast to neurofibromas, where it is always challenging which could be due to the infiltration of the tumours into the surrounding tissues. Fortunately, we did not find any tumour recurrence in 13 months of follow-up in any of our cases which could be due to the small sample size and the short period of follow-up.

Each of the surgical approach has its own complications (Table 2) and the most common being the marginal mandibular injury which can happen to almost all of the external approaches. In the present study, of 10 patients utilizing the transcervical excision, 3 patients had the marginal mandibular palsy in the immediate postoperative period although, one patient gets recovered after 48 h of conservative treatment. It could be because of the intraoperative stretching of the deep cervical fascia during the excision of the submandibular gland, which was performed in the majority of the cases to assess the poststyloid compartment of the PPS. Of the 2 patients undergoing combined transcervical transtemporal approach, one patient had grade III facial palsy in the immediate postoperative period, which could be due to the overstretching of the nerve during its translocation and later gets improved to grade I after 6 weeks with the conservative management. A patient with brachial plexus injury was well managed with active physiotherapy in the postoperative period. Injury to the internal jugular vein was noted in one case, during the exposure of the tumour which was later ligated intraoperatively. Wound infection was detected in one patient after 72 h of surgery, which was managed with conservative treatment. One patient diagnosed as neurofibrosarcoma in the final pathological examination was sent for the oncology department for the chemotherapy (doxorubicin). The patient was asymptomatic till 6 months of follow-up after that he developed a similar swelling over the right side of the neck and the MRI revealed the recurrence of the disease. He had denied for revision surgery and did not follow-up in the oncology clinic.

There were no significant differences in complication rate between the schwannoma and neurofibroma and also no direct correlation was found among the approaches of the surgery for the tumours. The incidence of malignant transformation of the nerve sheath tumour is not clearly demonstrated in the literature, although a study of intracranial nerve sheath tumour has shown the sporadic development of the malignant nerve sheath tumour.28 As shown in the present study, one patient underwent surgery for recurrent neurofibroma was confirmed to be a malignant nerve sheath tumour. Fluorodeoxyglucose-positron emission tomography (FDG-PET) is performed in all cases of MPNST to differentiate it from a benign nerve sheath tumour and has been demonstrated with high specificity.29

Because of the indolent nature and the remote chance of malignant transformation, selected patients can be allowed for the close follow-up, keeping mind the risks and benefits of the surgical treatment because of the high chance of neurovascular injury. In histopathological examination, schwannoma reveal the classical biphasic pattern of cellular characteristics, i.e. Antoni A and Antoni B. Antoni A consists spindle shaped cells with thin and long fibers with palisading arrangement of the nuclei around a central cytoplasmic mass (Verocay bodies). In contrast, Antoni B tends to be less cellular and contain more loosely arranged cells. The recurrence rates after resection are not well documented in the literature. In the present series, there was one recurrence after a mean follow-up of 12 months. The tumours are radio-resistant, and radiation therapy should be reserved for palliative treatment in selective cases where surgical treatment is contraindicated.

**Limitation of the study**

It is a retrospective study with small sample size with a short follow-up period. The purpose of the study was to highlight the management of the nerve sheath tumours with special concern to the surgical approach, complications and the clinical outcome.

**Table 2** Postoperative complications in the study population (n = 13).

| Complications       | n   |
|---------------------|-----|
| Vascular injury     | 1   |
| Neural injury       | 2   |
| Wound infection     | 1   |
| Recurrence          | 1   |
Conclusions

Excision of the nerve sheath tumours of head and neck is always a challenge to the surgeon because of their close proximity of the neurovascular structure. Although FNAC is diagnostic for the nerve sheath tumours, Clinico-radiological correlation is always the mainstay of diagnosis. Although the transcervical approach is commonly practiced for the majority of the tumours, combined approaches can be effectively applied for extensive nerve sheath tumours with the satisfactory clinical outcome.

Compliance with ethical standards

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.

Conflicts of interest

There are no conflicts of interest among the authors.

Research involving the human participants

Informed consent: Written informed consent has been taken from each patient prior to the surgery and same has been informed to the institute reviewer board.

No part of the body has been demonstrated in the case report without the permission of the concerned patient.

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