Extracranial non-vestibular head and neck schwannomas

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ABSTRACT

Objectives: To retrospectively describe our 10-year experience with extracranial non-vestibular head and neck schwannomas by presenting their clinical features, diagnostic methods, surgical decisions, and treatment outcomes.

Methods: This is a retrospective study conducted at the Department of Otolaryngology, Head and Neck Surgery, Shanghai Jiao Tong University Affiliated Shanghai First People's Hospital, Shanghai, China. The medical records of 46 patients diagnosed with schwannoma in the extracranial head and neck region as confirmed on paraffin-embedded sections from January 2003 to December 2012 were reviewed.

Results: All tumors were benign, and 52% presented as asymptomatic palpable solitary masses. Compressive symptoms, which can represent meaningful indicators of the nerve of origin were commonly noted. The most common nerve of origin was the brachial plexus (n=13, 28.3%).

Conclusion: While postoperative histopathologic examination is still the gold standard, fine needle aspiration cytology, CT scan, and magnetic resonance imaging may be useful in the diagnosis of schwannomas. As schwannomas are radioresistant, and as, despite their benign nature, can cause severe secondary symptoms, the best treatment of choice is complete excision with preservation of functions.

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Neurogenic tumors of the head and neck are relatively rare, and provide an interesting topic in terms of the clinical manifestation, diagnosis, and treatment. It comprise a series of neoplasms, including schwannomas, neurofibromas, and neuroepitheliomas, among others. Schwannomas are solitary, encapsulated, slow-growing, benign tumors arising from the Schwann cells of the peripheral, cranial, and autonomic nerves. Almost 25-45% of schwannomas are located in the head and neck region.[1] The clinical signs and symptoms vary according to the size and location of the tumor, and the nerve of origin. However, these clinical features are basically meaningless to the definite diagnosis.[2] Currently, although preoperative CT and magnetic resonance imaging (MRI) may provide information regarding the diagnosis of schwannoma, it can only be confirmed by postoperative histopathologic examination.[2] The preferred curative method is complete surgical resection,[2] although it is generally difficult to preserve the function of the affected nerve, owing to the nerve fascicles expanding thinly and randomly on the tumor's surface. Here, we describe 46 patients with extracranial head and neck schwannomas treated at our hospital during a 10-year period, to provide important information pertaining to the presentation, diagnosis, and management of this tumor.

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Methods. This is a retrospective study conducted at the Department of Otolaryngology, Head and Neck Surgery, Shanghai Jiao Tong University Affiliated Shanghai First People's Hospital, Shanghai, China. The medical records of 46 patients diagnosed with schwannoma in the extracranial head and neck region as confirmed on paraffin-embedded sections from January 2003 to December 2012 were reviewed. Patients with tumors arising intracranially, or from the trigeminal nerve were excluded from this study. Data collected included patients' gender and age, clinical signs and symptoms, tumor size, nerve of origin, anatomical location of the tumor, diagnostic methods, surgical decisions, histopathological findings, and treatment outcomes. All procedures carried out in this study complied with the ethical standards of the Shanghai Jiao Tong University Affiliated Shanghai First People's Hospital guidelines on human experimentation and with the Helsinki Declaration. Differences between the groups was assessed using the Chi-square test, with $p<0.05$ considered statistically significant.

Results. A total of 46 patients were identified and included in our analysis. Patients' ages ranged from 18-80 years (mean age at surgery - 47.6 years; median age - 45.5 years), and there was a slight female predominance (female: male ratio was 26:20 [1.3:1]; **Table 1**). All cases were benign, and the mean size of the tumors was 4.5 cm (range; 2-15 cm). The schwannomas in our cases were distributed in 4 anatomical sites as

follows: 42 (91.3%) were located in the neck region, one (2.2%) in the skull base, one (2.2%) in the oral cavity, and 2 (4.3%) in the parotid region. Eight cases of neck Schwannoma were found in the parapharyngeal space (**Table 2**). Most schwannomas presented as a symptomless isolated neck mass (n=28, 61%); 14 patients (30%) patients experienced neurological symptoms (that is, distal numbness of the hands or fingers, and Horner's syndrome), and 4 (9%) patients presented with pain, or obstructive symptoms. To facilitate the diagnosis and to gain as much information as possible, we performed preoperative imaging studies. However, the results of the preoperative studies were not as reliable as we had expected. Thirty patients underwent preoperative CT, while only 9 (30%) CT results suggested that the mass might be schwannoma. Similarly, MRI was performed in 19 patients, and only 9 of the MRI scans suggested that the masses were schwannomas. Twenty-three patients underwent preoperative ultrasound imaging of the mass, and fine needle aspiration cytology (FNAC) was carried out for tumors located at the superficial neck (n=6). In 2 (33.3%) of these cases, FNAC confirmed, or suggested the diagnosis of schwannoma, whereas 3 (50%) cases showed inconclusive results, and one (16.7%) case provided a diagnosis other than schwannoma. There were no significant differences in the diagnostic yield among these 3 diagnostic methods (p=0.305). It was possible to determine the nerve of origin in 24 patients (52%) preoperatively with 13 involving the brachial plexus, 4 involving the vagus nerve, 2 involving the facial nerve, 2 involving the sympathetic chain, 2 involving the cervical plexus, and one case involving the hypoglossal nerve. In 22 (48%) cases, the tumors were not arising from any major nerve (**Table 3**).

Table 1 - Age and gender distribution among patients included in a study in China (n=46).

Age, years	Males	Females
11-20	1	1
21-30	1	4
31-40	7	4
41-50	3	4
51-60	2	5
61-70	5	5
71-80	1	3
Total	**20**	**26**

Table 2 - Anatomical sites of extracranial head and neck schwannomas among patients included in a study in China (n=46).

Location	n (%)
Neck	42 (91.3)
Superficial neck	34 (73.9)
Parapharyngeal space	8 (17.4)
Oral cavity	1 (2.2)
Skull base	1 (2.2)
Parotid gland	2 (4.3)
Total	**46 (100)**

Table 3 - The nerve of origin of 46 extracranial head and neck schwannomas among patients included in a study in China (n=46).

Nerve of origin	n (%)
Brachial plexus	13 (28.3)
Vagus	4 (8.7)
Sympathetic chain	2 (4.4)
Facial nerve	2 (4.4)
Cervical plexus	2 (4.4)
Hypoglossal nerve	1 (2.2)
Unidentified	22 (47.8)
Total	**46 (100)**

In all cases, the tumors were resected completely. Most of the tumors (n=42, 92%) were resected via the transcervical approach. All parotid schwannomas derived from the facial nerve (n=2) were resected using at least superficial parotidectomy. Out of all 46 tumors, 44 (95.7%) were resected with intracapsular enucleation, and 2 (4.3%) were removed with the division of the nerve of origin. All cases were confirmed as schwannomas postoperatively by histopathological examination. Postoperative neurological sequelae separate from, or worse than those reported preoperatively were present in 20 (43.5%) patients. Of these, 18 patients underwent intracapsular enucleation, and 2 patients underwent complete resection of the affected nerve. The mean follow-up period was 41.9 months (range; 12-96 months). During this period, there were no cases of local recurrence in patients who underwent tumor resection with division of the nerve of origin, or intracapsular enucleation. During the clinical follow-up, one patient who was 80 years old at the time of operation, died due to an unrelated disease 5 years post-surgery. For the 14 patients who presented with neurological symptoms preoperatively, and the 2 patients who underwent tumor resection with division of the nerve of origin, the symptoms remained and never fully recovered during the follow-up period, whereas the 4 patients who underwent intracapsular enucleation experienced transient deficit, and recovered within 6 months postoperatively.

Discussion. Schwannomas originate from perineural Schwann cells and grow extrinsic to their parent nerve fascicles; they can occur along both sympathetic and somatic nerves in the body, with the exception of the olfactory and optic nerves, as these lack Schwann cells.[3] Due to the tertiary neurosurgical or otolaryngological treatment required for trigeminal and vestibular schwannoma patients, we only studied cases of extracranial non-trigeminal and non-vestibular schwannomas in this study. Accordingly, this study aimed to illustrate the clinical characteristics, diagnoses, treatments, and outcomes of extracranial non-vestibular schwannomas.

In the present study, schwannomas displayed a slight female predilection. In terms of patient age, schwannomas are most commonly reported in patients aged between 30 and 60 years. In the early stage, there are no specific symptoms or signs associated with schwannomas, and most patients present with an asymptomatic palpable solitary mass. Conversely, in the late stage, patients may endure physical discomfort, including neurologic deficits or obstructive symptoms.[4] In our cases, the symptoms included hoarseness, radiating pain of the upper limb, and painless swelling of the neck, among others. In general, the symptoms of neurologic deficit are important for the diagnosis of Schwannoma, especially for diagnosing the nerve of origin. Our results instead agreed with those of Yafit et al,[1] who reported that the most common nerve of origin is the brachial plexus. A preoperative diagnosis of schwannoma is difficult, and the differential diagnoses are diverse, including thyroid nodule, enlarged lymph node, paragangliomas, thyroglossal cyst, or tumor metastasis. To differentiate between schwannoma and these conditions, CT was performed in 30 patients in our study. On CT scans, small schwannomas are considered as homogenous, enhancing masses.[5] When the size is large (>3 cm), the tumor is often heterogeneous, with randomly distributed areas of low attenuation observed, surrounded by a peripheral enhancing ring. In general, cystic elements may be observed.[5] On MRI T1-weighted images, schwannomas show a low signal intensity ranging from the brain to the muscles, which can either be homogeneous or heterogeneous.[3] On T2-weighted images, schwannomas have a higher signal intensity than the cerebrospinal fluid, and the signal may be either heterogeneous or homogeneous.[3] After gadolinium injection, they are usually well enhanced.[6] In terms of the CT and MRI findings in our cases, the nerve of origin could be visualized in some patients upon MRI, but in none upon CT. Further, while FNAC remains a useful diagnostic method for head and neck masses, in our series, the diagnosis was inconclusive in approximately 50% of the FNAC samples. In these schwannomas, samples obtained were not enough to make a specific diagnosis. Nonetheless, metastatic or primary tumors can be excluded, and the presence of benign tissue can be suggested using this technique.[3] Moreover, in some cases, the FNAC procedure causes pain, and this was another reason for there being relatively few patients undergoing FNAC in our study.

Although CT, MRI, and FNAC may be somewhat helpful in distinguishing schwannomas from other tumors,[5] postoperative histopathologic examination is still the gold standard, with the presence of a clear capsule, Antoni A and/or B areas, and a positive reaction for S-100 protein considered characteristic histopathological features of schwannomas.[7] The Antoni A region can be described as a densely packed pattern of spindle cells, with frequent nuclear palisading arrangements, whereas the Antoni B region consists of loosely arranged spindle cells, with vacuoles and spindle-shaped nuclei prone to degeneration, hemorrhage, and cyst formation.[5] In both patterns, the tumor cytoplasm is elongated and the nuclei are regular and oval.[3] As schwannomas are both benign and radioresistant, complete surgical excision of the tumors by the appropriate approach is considered the standard curative treatment.[1] However, owing to the potential neurological dysfunction associated with the surgery, conservative methods should be considered.[1] In our study, most patients underwent complete surgical excision despite having symptomless isolated neck masses. Moreover, although there were no cases of recurrence in this study, it is also a controversial issue whether benign tumors can become malignant.[4] Whatever the chosen treatment approach is, the nerves are likely to be influenced.[1] Of note, previous reports have shown that significant recurrence did not occur during long-term follow-up in patients who received conservative treatment.[6] Hence, due to the benign nature of this tumor, the above methods should be compared and assessed in the future in order to establish the optimal treatment for allowing preservation of nerve functions. However, it is still necessary to inform the patients of the possibility of neurological sequelae before the operation.[3] Many studies have demonstrated the importance of preserving the nerve, while the preservation of structure does not always ensure functional preservation; thus, postoperative rehabilitation and neural reconstruction are highly

important.[6] Close observation and follow-up is another option, especially in selected cases of small incidental tumors at high risk of postoperative complications. Because of the multiple symptoms, wide range in location, size of the tumors, and rarity of the disease, establishing the optimal treatment is challenging. The limitation of this study is the small sample size, which weakens the stringency of results.

In conclusion, preoperative suspicion and awareness of the possibility of schwannoma are very important in making a proper diagnosis. Adequate imaging studies should be performed to increase the diagnostic rate preoperatively, and owing to the benign nature of this disease, the patients' symptoms and willingness to undergo surgery should be taken into consideration when choosing the appropriate treatment modality.

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