

Original Article

Head and Neck Nerve Sheath Tumors: A 10-Year Evaluation in Iran

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ABSTRACT

Background and Objective: Approximately 45% of benign peripheral nerve sheath tumors (PNST) occur in head and neck regions. Malignant peripheral nerve sheath tumors (MPNST) are less common and only 815%- of these tumors occur in head and neck region. In this study, we aimed at evaluating the demographic characteristics, pathologic findings, locations, main clinical presentations and family history of these tumors in head and neck regions.

Patients and Methods: In a cross sectional and retrospective study, we analyzed head and neck peripheral nerve sheath tumors in patients who referred to Amir-Aalam Hospital affiliated to Tehran University of Medical Sciences for 10 years (1996 to 2005).

Results: There were 113 cases with a kind of nerve sheath tumor; the female/male ratio was 1.09. The frequency of schwannoma, neurofibroma and MPNST was 66.37%, 30.1% and 3.53%, respectively including 39 cases of acoustic neuroma, 18 cases of cervical nerve trunk involvement, 12 cases of facial nerve involvement, 11 cases of the 10th cranial nerve PNST and only one case of cervical sympathetic nerve involvement.

Conclusion: Schwannomas are the most common types of PNSTs seen in head and neck regions, and acoustic neuroma comprises the majority of them. About 25- 45% of extracranial schwannomas also occur in head and neck regions that usually present a slow growing of the mass lesion and various mass effects. Although neurofibroma and MPNSTs are less common, they must be kept in mind as differential diagnoses of other benign and malignant lesions in this region.

Key words: Nerve Sheath Tumors, Head Neoplasms, Neck Neoplasms, Iran

Introduction

Approximately 45% of benign peripheral nerve sheath tumors (PNST) including schwannoma (intra- or extra-cranial) and neurofibroma occur in head and neck regions (1). Schwannoma and neurofibroma are benign tumors that arise from nerve sheath structures.

An interesting report suggested that some nerve sheath tumors may contain histologically clear components of both Schwannoma and neurofibroma within the same specimen; that may explain the confusion about making an accurate histological diagnosis in the past (2). Neurofibroma may be solitary or multiple, sporadic or in association with

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neurofibromatosis (NF) I or II syndromes.

Malignant peripheral nerve sheath tumors (MPNST) are less common and only 8-15% of these tumors occur in head and neck regions (3). MPNST never arises from the malignant transformation of schwannoma; they may be associated with neurofibromatosis or occur in a sporadic manner (4).

In this study, we aimed at the retrospective evaluation of the demographic characteristics, pathologic findings, location, main clinical presentations and family history of these tumors in head and neck regions.

Materials and Methods

In a cross sectional and retrospective study, we analyzed head and neck peripheral nerve sheath tumors for 10 years (1996 to 2005). The Ethic Committee of our hospital gave approval for the study. The cases were patients who referred to Amir-Aalam Hospital affiliated to Tehran University of Medical Sciences for a head and neck mass surgery and a histopathologic diagnosis of a nerve sheath

tumor. Amir-Aalam Hospital admits patients from all region of Iran, representing a wide spectrum of socioeconomic levels.

The required clinical and pathologic data of the patients include their age, gender, family history, tumor pathology, tumor location, the involved nerve and the main clinical presentations, which were collected from the hospital archive. The Data were analyzed using the computer software package SPSS 9.0 (SPSS, Chicago, IL). All P values were two tailed, with statistical significant defined by $P \leq 0.05$.

Results

During 10 years, there were a total of 113 cases with a kind of nerve sheath tumor, the F/M ratio was 1.09 and the age range was between 13 and 78 yr (mean: 48.5). The frequency of schwannoma, neurofibroma and MPNST was 66.37%, 30.1% and 3.53%, respectively. Table 1 shows the age and sex distribution of each tumor type.

Nine cases (7.96%) had a positive family history of PNST, seven of which presented neurofibroma.

Table1: Age and sex distribution of each tumor type

Tumor type	SEX (No)		AGE (No)		
	Female	Male	<20	20-39	≥40
Schwannoma	44	31	10	33	32
Neurofibroma	13	21	11	12	11
MPNST*	2	2	0	3	1
Total	59	54	21	48	44

MPNST: Malignant peripheral nerve sheath tumors

Table 2 shows the frequency of each PNST according to the specific nerve involved in head and neck regions.

Table 2:The number of each tumor type according to the specific involved nerve in head and neck regions

Tumor type	Acoustic nerve	Cervical nerve plexus	Facial nerve	Vagus nerve	Sympathetic plexus	Unknown
Schwannoma	39	7	7	8	1	13
Neurofibroma	0	10	4	1	0	19
MPNST*	0	1	1	2	0	0
Total	39	18	12	11	1	32

MPNST: Malignant peripheral nerve sheath tumors

Table 3 shows the frequency of each PNST according to the exact anatomic site in head and neck regions.

Table 3: The number of each tumor type according to the exact anatomic site in head and neck regions

Tumor type	Intracranial	Lateral neck	Nasopharynx and Sinuses	Scalp and Facial soft tissue	Oropharynx	Parotid gland	Parapharyngeal space	Larynx	External ear
Schwannoma	45	7	7	2	5	2	4	3	0
Neurofibroma	0	6	7	8	6	5	1	0	1
MPNST*	0	1	0	0	0	1	2	0	0
Total	45	14	14	10	11	8	7	3	1

MPNST: Malignant peripheral nerve sheath tumors

Among the 39 cases of acoustic neuroma, 10.26% were below 20 years, 33.33% were 20-40 yr old and 56.41% were older than 40 yrs. In addition, the F/M ratio was 25/14 and the tumor location was intracranial. The most frequent patient symptoms were sensory neural hearing loss (92.3%), tinnitus (82%) and vertigo (28.2%). The other infrequent symptoms were pain, ataxia and seizure. As it is obvious, all tumors with the 8th cranial nerve involvement were schwannoma.

Cervical nerve trunk involvement was seen in 18 cases. Most of these cases (50%) were above 40 years old and the F/M was 5/13. All patients had a lateral neck mass lesion. Most of these tumors were neurofibroma.

There were 12 cases with facial nerve involvement, 50% were below 20 years old, 41.66% were 20-40 yrs old and 8.33% were older than 40 yrs. Furthermore, the F/M ratio was 5/7; 58.33% was extratemporal, 33.33% was intratemporal and 8.33% was intracranial. Most patients presented a mass on their parotid region. The other presenting symptoms were hemifacial palsy and hearing loss. Schwannoma was the preferred pathology.

Eleven cases existed with the 10th cranial nerve PNST, 72.72% were 20-40 years old and the F/M ratio was 7/4. Hoarseness (54.5%), mandibular angle mass (36.3%), pharyngeal mass, dysphagia and dyspnea were the most frequent symptoms. Besides, schwannoma was the most prevalent pathologic feature. All of the seven tumors located in the parapharyngeal space originated from the 10th cranial nerve.

Only one case was found with cervical sympathetic nerve involvement. She was 37 years old, presented with a lateral neck mass and the histopathologic diagnosis was schwannoma.

In 32 cases, the exact involved nerve could not be truly specified. According to the position, these tumors were located in Nasopharynx and Sinuses (14 cases), oropharynx (11 cases), Scalp and Facial soft tissue (5 cases), Parotid gland (1 case) and the external ear (1 case).

The clinical presentations in 14 cases of PNST in the sinonasal region were nasal obstructive mass (78.5%), proptosis (35.7%), epistaxis and pain. The Maxillary sinus was the only involved sinus.

Discussion

PNSTs of head and neck regions comprise a very small percent but an important section of all neoplastic lesions in this area. They are a complex array of both benign and malignant tumors including neurofibroma, schwannoma and MPNST. On the other hand, head and neck regions are by far the most common sites for schwannoma (5). PNSTs are of great interest because of the disturbance in the involved nerve function, their cosmetic effects and differential diagnosis with other benign and malignant tumors in the head and neck territory. Besides, operative procedures are of great importance in the complete and safe removal of tumors that are usually located adjacent to a nerve bundle.

PNSTs may arise from any peripheral, cranial and autonomic nerve in head and neck (5). Das Gupta *et al.* (6), in their series of 303 benign nerve sheath tumors found that 44.87% of the tumors occurred in head and neck regions. In the neck, they may arise medially or laterally. Medially, they arise from the last four cranial nerves (glossopharyngeal, vagus, accessory and hypoglossal) or the sympathetic chain. Laterally, they arise from cutaneous or muscular branches of the cervical plexus or the brachial plexus (7).

Neurofibroma is less prevalent than schwannoma in this region but similar to schwannoma, it can occur in different parts of head and neck, including the tongue, nasopharynx, larynx, vagus, buccal area and the salivary gland. Neurofibromas may be solitary or multiple (i.e. neurofibromatosis type 1 and 2).

Malignant peripheral nerve sheath tumors account for only 5% of all soft tissue sarcomas with a preference for extremities (3, 8, 9). Only 8 to 15% of these tumors arise from head and neck regions (3, 9).

It is now felt that Schwann cells are the common precursor for most nerve sheath tumors (5). Twenty-five to 45% of all extracranial schwannomas occur in head and neck regions (10). The usual presenting symptom is the gradual enlargement of the neck mass. Pain and neurologic deficit are ominous signs and are suggestive of malignancy (11).

There are several case reports about head and neck schwannoma, for example, in the tongue (12, 13), cervical sympathetic chain (14, 15), sinonasal tract and nasopharynx (1, 16) as well as the parotid gland (17).

The preoperative diagnosis of PNSTs and the consideration of differential diagnoses are extremely important if one is to warn the patient about possible neurological sequelae post-operatively, i.e. Horner's syndrome, vocal palsy, etc. Obviously, for benign lesions, all efforts must be made to preserve the nerve of origin (5).

For each region, other more common benign and malignant differential diagnoses must be considered. Some examples include carotid body tumor, lymphoma and other soft tissue tumors in the neck region, deep lobe parotid tumors in the territory of facial nerves and meningioma in the place of acoustic neuroma.

Fine needle aspiration, while conclusive in many cases of neck masses, is much less valuable for compact neural tumors (15). Imaging studies play a central role in the diagnosis of head and neck PNSTs; it may show the nerve of origin; besides, schwannoma, in general, is more hypodense in comparison to muscles in CT scans without contrast media. Other studies such as MRI or angiography may be needed to rule out other differential diagnoses (15-18).

Classically, a neurofibroma will be intimately associated with the nerve of origin, while a schwannoma will arise eccentrically and thus will be more amenable to the surgical sparing of the nerve (15, 19). However, this strictly depends on the involved nerve, for example, Hood *et al.* were not able to dissect the tumor from the sympathetic chain

in a series of cervical sympathetic chain schwannoma and due to the intimate involvement of the nerve, they required the resection of a segment of the sympathetic chain (15).

From the pathologic point of view, schwannomas demonstrate two distinct areas of cellularity. Antoni A regions are comprised of more densely arranged spindle cells with specific areas of palisading nuclei known as Verocay bodies. Antoni B regions tend to be more hypocellular, with a more loose and disorderly arrangement. Microscopically, neurofibromas are formed by a combined proliferation of all the elements of a peripheral nerve: axons, Schwann's cells, fibroblasts, and (in the plexiform type) perineurial cells. The nuclei of the tumor cells of neurofibroma show a typical fascicular pattern of growth and serpentine shape. Malignant peripheral nerve sheath tumor (MPNST) show marked hypercellularity and a high mitotic activity in the absence of significant pleomorphism. The plump and almost the epithelioid appearance of the cells surrounding the vessels is a common feature in this tumor type (20, 21).

Conclusion

Schwannomas are the most common types of PNSTs seen in head and neck regions, and acoustic neuroma includes the majority of them. Excluding this intracranial tumor, between 25-45% of extracranial schwannomas also occur in head and neck regions that are usually presented with a slow growing of the mass lesion and various mass effects. Although Neurofibroma and MPNST are less common, they must be kept in mind as differential diagnoses of other benign and malignant lesions in this region.

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