Head and Neck Schwannomas: 20-Year Experience of a Single Institution Excluding Cutaneous and Acoustic Sites

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Abstract While head and neck sites comprise the most common location of schwannomas, clinicopathologic data regarding those tumors occurring in non-acoustic and non-cutaneous locations are relatively sparse. In this study, therefore, we sought to examine retrospectively the clinical and pathologic features of head and neck schwannomas excised at our institution over a 20-year period. During this period, we identified a total cohort of 85 patients, which included 36 males (42.4 %) and 49 females with average age of 41.3 years, the majority of which presented asymptotically with a mass. Localized symptoms were, however, associated with all of the schwannomas that arose in the oral cavity and larynx, while tumors within or adjacent to bone were often associated with neurologic complaints (7 of 15 such tumors [46.7 %]). Clinical follow-up data was available in 86.4 % of all cases and demonstrated no recurrences or mortality. Pathologically, the microscopic features were characteristic of those well-described for schwannomas in other sites, including alternating Antoni A and B areas and the presence of degenerative changes. Tumor encapsulation, however, was variable and was completely absent in schwannomas of the nasal cavity, paranasal sinuses, and larynx. Additionally, a significant minority of the tumors (28.2 %) exhibited foci that resembled neurofibroma. Non-acoustic, non-cutaneous schwannomas of the head and neck appear to have clinicopathologic features similar to their soft tissue counterparts with some subsite variation in presentation and/or microscopic features.

Keywords Schwannoma · Sinonasal · Nerve sheath tumor

Introduction

Schwannomas are benign neoplasms that arise from the Schwann cells of peripheral nerves. They occur throughout the body, and early data that considered schwannomas and neurofibromas together demonstrated the head and neck to be the most common region of the body in which these tumors arise [1]. More recent studies that have separated these entities have shown schwannomas to be the most common benign nerve sheath tumor of the head and neck [2, 3]. The clinical features of head and neck schwannomas have been documented in numerous short series [2–6] and case reports, including specific subsites such as the tongue [7–10], palate and other intraoral locations [11–16], and nose, paranasal sinuses, and anterior skull base [17–28]. Pathologic details of schwannomas arising in the head and neck are, however, sparsely documented in the literature. The aim of the present study was, therefore, to examine more thoroughly the clinicopathologic characteristics of head and neck schwannomas in a relatively large series collected at a single institution.

Methods

The surgical pathology files of our institution were searched to identify schwannomas excised from head and neck sites over the 20-year period 1995–2014; cutaneous and acoustic tumors were excluded from the study. The medical records of the corresponding patients were reviewed to document age at presentation, gender, clinical presentation, and outcome after

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Table 1 Summary of clinical features of head and neck schwannomas

Site (% of tumors)	Male:female (% male)	Average age (range) (years)	Presentation	Cases with follow-up	Average follow-up (range) (years)	Percent with NED
Nasal cavity (4.7)	1:3 (25.0)	43.1 (18.6–65.3)	Local symptoms—4/4 (100 %) Neurologic symptoms—0 Pain—0 Mass/asymptomatic—0	4/4 (100 %)	2.4 (0.2–4.3)	100
Paranasal sinuses (3.5)	2:1 (66.7)	62.3 (46.0–78.6)	Local symptoms—1/3 (33.3 %) Neurologic symptoms—1/3 (33.3 %) Pain—0 Mass/asymptomatic—0	3/3 (100 %)	0.7 (0.1–1.3)	100
Oral cavity (9.3)	3:5 (37.5)	21.6 (7.1–33.8)	Mass/asymptomatic—1/3 (33.3 %) Local symptoms—0 Neurologic symptoms—0 Pain—3/9 (33.3 %) Mass/asymptomatic—4/9 (44.4 %) Unknown—2/9 (22.2 %)	7/9 (77.8 %)	1.8 (0–10.3)	100
Parotid gland (5.8)	2:3 (40.0)	46.9 (37.0–67.8)	Local symptoms—0 Neurologic symptoms—0 Pain—0 Mass/asymptomatic—5/5 (100 %)	5/5 (100 %)	1.2 (0.3–3.2)	100
Larynx (2.3)	0:2 (0)	43.5 (21.3–65.6)	Local symptoms—2/2 (100 %) Neurologic symptoms—0 Pain—0 Mass/asymptomatic—0	2/2 (100 %)	1.0 (0–1.9)	100
Parapharyngeal space (9.3)	4:4 (50.0)	49.4 (25.7–62.7)	Local symptoms—1/8 (12.5 %) Neurologic symptoms—0 Pain—0 Mass/asymptomatic—7/8 (87.5 %)	8/8 (100 %)	2.9 (0.6–6.0)	100
Soft tissue (47.7)	21:20 (51.2)	40.5 (8.2–73.4)	Local symptoms—4/42 (9.5 %) Neurologic symptoms—2/42 (4.8 %) Pain—3/42 (7.1 %) Mass/asymptomatic—30/42 (71.4 %) Unknown—3/42 (7.1 %)	33/41 (80.5 %)	1.7 (0–13.4)	100
Intra/juxtaosseous (17.4)	3:12 (20.0)	42.6 (14.2–63.7)	Local symptoms—4/15 (26.7 %) Neurologic symptoms—7/15 (46.7 %) Pain—2/15 (13.3 %) Mass/asymptomatic—2/15 (13.3 %)	14/15 (93.3 %)	4.2 (0.1–10.4)	100

excision. Presenting signs and symptoms were categorized as asymptomatic or detection of a mass, pain, neurologic manifestations, or local effects due to the site of the tumor. The outcome of disease was determined from a physician's note or radiology report, whichever was more recent.

All identified cases for which slides were available were retrospectively reviewed, and the following features were evaluated: presence or absence of Antoni A and B areas, Verocay bodies, hyalinized vessels, and degenerative features (ancient change). Additionally, encapsulation of tumors, the presence of neurofibroma-like foci, and whether the tumors fulfilled criteria for one of the recognized variants of schwannoma were documented. The sizes of the schwannomas were obtained from the gross pathology reports, as was the head and neck subsite in which the tumor was located; the location was clarified by the corresponding operative report when necessary.

Results

A total of 85 patients had histologically confirmed head and neck schwannomas excised over the study period (Table 1). Across all subsites, these patients included 36

males (42.4 %) and 49 females with an average age of 41.3 years. The patients included 5 with a documented history of neurofibromatosis type 2 (NF2); 3 of the NF2 patients each had 2 non-acoustic, non-cutaneous head and neck schwannomas excised, yielding a total of 88 tumors. Note that the number of patients presented in Table 1 sums to 86 because 1 of these latter NF2 patients had tumors in 2 subsites, while the other 2 NF2 patients had separate schwannomas in the same regions. The majority of the total cohort of tumors (55.7 %) was asymptomatic, presenting as a mass that was detected clinically or incidentally on radiologic imaging performed for another reason. Of note, however, all of the schwannomas within the oral cavity or larynx manifested with localized symptoms, and neurologic complaints were common among the intra- and juxtaosseous tumors. Outcome information was available for 76 of the tumors (86.4 %); all patients were alive and free from recurrence at last follow-up.

Regarding pathologic features, 85 tumors had slides available for review (96.6 %), including 4 cases in the nasal cavity, 3 cases in the paranasal sinuses, 9 cases in the oral cavity, 5 cases in the parotid gland, 1 case in the larynx, 8 cases in the parapharyngeal space, 40 cases in the

Table 2 Summary of pathologic features of head and neck schwannomas

	Average size (cm)	Growth pattern (%)	Verocay bodies (%)	Hyalinized vessels (%)	Ancient change (%)	Encapsulated (%)	NF-like areas (%)
Nasal cavity	4.8	Antoni A dominant—75 Antoni B dominant—25 Equal—0	50.0	75.0	100	0	25.0
Paranasal sinuses	3.0	Antoni A dominant—66.7 Antoni B dominant—0 Equal—33.3	33.3	100	100	0	33.3
Oral cavity	1.7	Antoni A dominant—100 Antoni B dominant—0 Equal—0	100	44.4	44.4	66.7	22.2
Parotid gland	3.0	Antoni A dominant—100 Antoni B dominant—0 Equal—0	100	100	100	80.0	20.0
Larynx	5.5	Antoni A dominant—100 Antoni B dominant—0 Equal—0	100	100	100	0	100
Parapharyngeal space	4.4	Antoni A dominant—50.0 Antoni B dominant—25.0 Equal—25.0	75.0	100	100	100	0
Soft tissue	3.7	Antoni A dominant—52.5 Antoni B dominant—20.0 Equal—27.5	92.5	80	92.5	90	37.5
Intra- or juxtaosseous	2.6	Antoni A dominant—66.7 Antoni B dominant—33.3 Equal—0	66.7	73.3	93.3	40.0	20.0

soft tissue of the neck, and 15 cases in or adjacent to bone (Table 2). Upon microscopic examination, the majority of tumors were relatively cellular with Antoni A foci predominating over Antoni B areas in each of the subsites. Other characteristic features of schwannomas, including hyalinized vessels and ancient/degenerative changes, were noted for a subset of tumors in each location, but such findings were least common among tumors arising in the oral cavity. Among recognized variants of schwannoma, only the plexiform variety was identified in our study ($n = 4$); the plexiform tumors included 3 cases in 2 different patients with NF2 and 1 sporadic example (Fig. 1a). Encapsulation of the tumors was variable across subsites and was absent in all schwannomas that arose in the nasal cavity, paranasal sinuses, and larynx (Fig. 1b, Fig. 2). Not uncommonly the schwannomas contained foci reminiscent of neurofibroma (NF) (Fig. 1c), being observed in 28.2 % of all cases. Ulceration of the overlying skin or mucosa, in contrast, was rare and identified in only three cases, although this feature was noted in 2 of 9 tumors located in the oral cavity. Similarly, erosion into bone was only infrequently identified ($n = 5$, 5.7 % of total) but occurred in 2 of 4 nasal cavity schwannomas. Six of the tumors were noted to contain pseudoglandular spaces, including 3 of the 5 cases that occurred within the parotid gland (Fig. 1d).

Other infrequent microscopic features included the presence of Gamna-Gandy bodies and adipose tissue in 1 case each.

Discussion

Schwannomas are benign neoplasms that exhibit differentiation toward the Schwann cells that produce the myelin sheath of peripheral nerves. These tumors have been documented at sites throughout the body, including the head and neck, an anatomic region in which they are particularly common. Despite the frequency with which schwannomas occur in head and neck locations, clinicopathologic details in these sites are limited. In the current study, we describe the clinical and pathologic findings of a large series of head and neck schwannomas collected at our institution over a 20-year period, excluding those arising in the skin and from the eighth cranial nerve (acoustic schwannoma, acoustic neuroma).

Clinically, the schwannomas in our series were largely tumors of adults, the average age being in the fifth decade for all of the subsites except the oral cavity. In the oral cavity, there were 3 cases in children and adolescents, but the small number of tumors arising in young persons in our

Fig. 1 Plexiform schwannoma from a patient with neurofibromatosis type 2 (a). A schwannoma of the nasal cavity demonstrating lack of encapsulation (b). Schwannoma with neurofibroma-like foci (*right side*), characterized by coarse collagen bundles in a myxoid

background (c). A prominent pseudoglandular space within a schwannoma of the parotid gland with condensation of tumor cells at the periphery of the space (*upper left*) (d)

Fig. 2 An intranasal schwannoma, which grew as a polypoid mass (a) and lacked a capsule when sectioned (b). A capsule is grossly apparent in this schwannoma excised from a parotid gland (c)

series (9 tumors in 8 patients) does not allow us to conclude whether there is a proclivity for oral schwannomas in young patients. The most common presentation was that of an asymptomatic mass, which was often noted only incidentally by radiologic imaging performed for another indication. Tumors in the oral cavity and larynx, however, uniformly manifested localized symptoms. This observation is congruent with at least one previous report [8], in which tumors located in the base of tongue were more likely symptomatic than those arising in the oral tongue.

Neurologic complaints were similarly common among tumors that arose within or adjacent to bone, suggesting that confinement of growth of the mass may lead to such complaints. The frequent occurrence of clinical complaints when schwannomas arise in these mucosal and bony sites should not, therefore, be misinterpreted as an unequivocal indication of malignancy. Outcome data was available for the majority of the schwannomas in our series. All patients for whom outcome was documented were free from recurrence at last follow-up, in accordance with most previous reports, including a systematic review of 94 schwannomas of the sinonasal tract and anterior skull base in which there were only 3 recurrences [28]. One study that compared the behavior of oral and maxillofacial schwannomas to those arising in the neck, however, found that one-third of the former tumors recurred, perhaps due to incomplete excision as a result of anatomic constraints [29].

Intratumoral foci that resembled NF were observed in each of the subsites except the parapharyngeal space. These foci were present in schwannomas from both patients with documented NF2 (6 of 8 such tumors) and those without this condition. A retrospective review of tumors with hybrid features of schwannoma and NF documented 9 such cases, which were located within the extremities and trunk [30]. In this study, the hybrid neoplasms consisted of NF with central, demarcated schwannomatous nodules, the latter regions manifesting typical features such as hyalinized vessels, degenerative atypia, and more diffuse S-100 protein immunohistochemical staining. A case report of a hybrid neoplasm arising from the facial nerve has also been published [31]. Our study appears to be the first to describe the prevalence of NF-like foci among a series of head and neck schwannomas and demonstrates that such hybrid features are relatively common in this region among patients with and without NF2. Of note, a recent review of 12 NFs arising in the sinonasal tract also included 1 tumor with features of both NF and schwannoma, leading the authors to emphasize that schwannoma, NF, and perineurioma have overlapping morphologies and may blend together [32]. Recognition of the occurrence of neoplasms that combine features of schwannoma and NF is important so as not to overdiagnose malignant degeneration of the latter [30]. Furthermore, the significant rate of occurrence of such hybrid features in sporadic tumors implies that clinical investigation for other features of neurofibromatosis is not necessarily required.

Schwannomas are generally encapsulated neoplasms, but a subset of schwannomas in our series lacked a capsule, including all of those arising in the nasal cavity, paranasal sinuses, and larynx. This observation corroborates the findings in other series of sinonasal schwannomas, in which only 1 of 18 total tumors was encapsulated [19, 20,

22–24]. The reason(s) for absence of a capsule in such mucosal sites is unknown but has been hypothesized to be related to origin from autonomic nerves, which lack an epineurium, or biologic peculiarities of the submucosal setting [22]. Regardless of the underlying mechanism, lack of encapsulation appears to be a nearly constant feature of mucosal-based schwannomas, a histologic feature that differs from their soft tissue counterparts.

Based on our large series of head and neck schwannomas, these neoplasms appear to share many clinical features with those that arise in soft tissue, including typical occurrence in adults as an asymptomatic mass. Tumors within the oral cavity or larynx or in relation to bones of the head and neck are, however, often associated with symptoms. Nonetheless, clinical outcome is excellent, with all patients in our study being free of disease at last examination. The majority of tumors we examined demonstrated pathologic features characteristic of schwannomas at other sites, although mucosal location was invariably associated with lack of encapsulation. The presence of NF-like foci was also commonly observed and should not be mistaken for malignant transformation when associated with degenerative features.

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