# The Prognosis and Recurrence of Head and Neck Schwannomas : An 8-year Retrospective Study

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## Abstract

Introduction: We studied a series of head and neck schwannoma (also known as neurilemmoma, neurinoma, neurolemmoma, and Schwann cell tumor) treated during an eight-year period. Focus was given to analyzing location, symptoms, nerve of origin, postoperative resolution, and recurrence. The objective of this article is to provide the patient and relative healthcares some reference suggestions.

Materials and Methods: Clinical records of 69 head and neck schwannoma patients treated in Taichung Veterans General Hospital since January 2003 to December 2010 were retrospectively examined. We surveyed the tumor location, the symptoms, nerve of origin, postoperative nerve injury symptoms, resolution, and recurrence after resection of head and neck schwannomas with characteristics statistics and comparison analysis.

Results: In this study, patients' gender was approximately 55% female and 45% male. They ranged from 12 to 83 years of age with the mean age of diagnosis at 47.84 years. In this study, lesions were diagnosed in the head region of 53 patients (including 5 oral cavities, 32 brain cerebellopontine angles, 11 brain other than cerebellopontine angles, 2 eyes, and 3 scalps), as well as lesions in the neck area of 16 additional patients. The most common observation was found along the acoustic nerve and diagnosed as cerebellopontine angle (C-P angle) syndrome. Resolution rates were 59.4% after resection of schwannomas. According to protect the function of origin nerve, we would not remove those tumors completely. Among these was one case of neurofibromatosis type 2—an inherited disease. For recurrent intracranial schwannomas, 4 of 6 cases were C-P angle schwannomas. Intracranial schwannomas showed a recurrence rate of 7.2% which was higher than the extracranial schwannomas recurrence rate of 1.4%.

Discussion: Schwannoma is a slow-growing tumor, its involucrum is complete, the boundary is clear, and the delayed appearance of symptoms may result in late diagnosis and treatment. In general, the choice of surgical approaches often depends upon location, but the best outcomes normally resulted from surgical intervention. Nevertheless, once intracranial schwannomas recur, then successive treatments of Gamma Knife radiosurgeries were performed in our hospital.

**Key words**: Head and neck schwannomas, Nerve of origin, Recurrence rate.

## Introduction

Schwannomas of the head and neck are abnormal nerve sheath neoplasms that may originate from any cranial, peripheral or autonomic nerves. It is also called neurilemmomas or neurinomas, which are slow-growing, benign, and encapsulated. Schwannoma can be distinguished from neurofibroma which can also attack the nerve fibers. While neurofibroma involves entwining of the tumor with the parental nerve fascicles, by contrast, schwannoma grows extrinsic to the nerve fibers. Schwannomas may appear similar to infection or tumor metastasis in the head and neck area. Previous research indicates that about 25-45% of cranial schwannomas arise anywhere along the neuron axon from the skull base or spinal column to the skin, mucosal, or end organ structures of the head and neck area[1, 2]. Only 1% of schwannomas come from an intraoral origin, such as a predilection for the tongue, followed by the palate, buccal mucosa, floor of the mouth, and mandible[3]. Malignant change in head and neck schwannomas is rare with the occurrence normally less than 14%[4].

A definitive diagnosis can only be concluded based on the histological and immunohistochemical findings[4,7]. Schwannoma which originates from the Schwann cell, is almost exclusively comprised of elongated spindle cells with long, oval nuclei and indistinct cell membranes. It is often arrayed in a palisade pattern (Antoni A regions) or edematous regions with loosely arranged cells in a myxoid matrix prone to degeneration (Antoni B regions)[5,6]. The treatment for Schwannomas is typically surgical excision, however, when Schwannomas occurs in the brain, the treatment can be surgical excision or Gamma Knife radiosurgical treatment for small tumors. This more conservative approach is taken to avoid neurological decline that can result from brain surgery. In our hospital, the treatment policy for brain tumors asserts that the surgeon shall remove the tumor conservatively in order to preserve as much brain function as possible for tumor sizes larger than 4 cm. Any smaller or residual tumors will be removed by Gamma Knife radiosurgery.

## Materials and Method

Clinical records of 69 patients with schwannomas treated in our hospital from January 2003 to December 2010 were retrospectively reviewed from the medical records of the Taichung Veterans General Hospital, the largest tertiary referral center in central part of Taiwan. Data collected from patients included sex, age, pathology report, magnetic resonance imaging (MRI) or computed tomography (CT) scan examinations to facilitate diagnosis and relative anatomy, the tumor location, ailments and symptoms, nerve of origin, and possible postoperative nerve injury. We also focused on resolution, and recurrence after resection of head and neck schwannomas.

## Results

From our research of head and neck Schwannomas, the age range of the patients' studies was 12 to 83 years with the mean age of diagnoses at 47.84 years (the mean ages were 49 for males and 46.8 for female). It should be noted that females represented the majority (55%) in Table 1. There were 53 cases (41.5% male) that showed a predilection for the head region and

Table 1. Statistical description of head and neck schwannomas patients

| Age at diagnosis distributions | | | Tumor location | | |
|---|---|---|---|---|---|
| Age | Male | Female | Location | Male | Female |
| 11-20 | 0 | 4 | Head | | |
| 21-30 | 3 | 1 | Oral cavity | 2 | 3 |
| 31-40 | 5 | 5 | Brain (C-P angle) | 14 | 18 |
| 41-50 | 10 | 7 | Brain(others) | 6 | 5 |
| 51-60 | 6 | 16 | Eye | 0 | 2 |
| 61-70 | 5 | 4 | Scalp | 0 | 3 |
| 71以上 | 2 | 1 | | | |
| Total | 31 | 38 | **Total** | **22** | **31** |
| Year of surgery | | | Neck | | |
| Year | male | female | Location | Male | Female |
| 2003 | -- | -- | Pharynx | 1 | 1 |
| 2004 | 3 | 2 | Submandibular gland | 1 | 1 |
| 2005 | 6 | 2 | Spinal cord | 3 | 4 |
| 2006 | 3 | 5 | Neck soft tissue | 4 | 1 |
| 2007 | 7 | 7 | **Total** | **9** | **7** |
| 2008 | 4 | 3 | | | |
| 2009 | 6 | 13 | | | |
| 2010 | 2 | 6 | | | |

16 cases (56.2% male) in the neck region. This neoplasm is usually a solitary lesion, and can be multiple when linked with neurofibromatosis.

Within the population that was studied for head and neck Schwannomas, the most common sites observed were the brain (consisted of 20 males and 23 females), nerves of origin such as the VIII acoustic nerve (34 patients), followed by the cervical plexus (10 patients), the trigeminal nerve (8 patients), and the vagus nerve (5 patients). In our study, the nerve of origin at C-P angle schwannomas attacked to acoustic nerve most frequently. Table 2 illustrates that the resolution rate was 59.4% after resection of schwannomas. Where resolution is defined as a patient without any tumors from post-operative MRI or CT scans after 3 months or longer. In this research we observed a recurrence rate of 8.6% including 6 recurrent cases and only one scalp tumor recurrent case. From the five cases of intracranial schwannomas, 4 cases were C-P angle schwannomas and a single case was schwannoma that originated from the oculomotor nerve. Obviously, intracranial schwannomas had a higher recurrence rate of 8.6% than the extracranial schwannomas recurrence rate of 1.4%. The tongue mass was reported in 3 (60%) patients, making it the most common of the 5 oral cavity schwannomas

Table 3 describes the six recurrent cases. The term “recurrence” as defined in this paper indicates that such was recorded either on the admissions notes or the operation notes from the surgeon. It shows time intervals from the operation date to the recurrent date vary from 2 to 36 months for head and neck schwannomas. There was only one unhealed young female patient that had bilateral acoustic neuroma,

Table 2. Characteristic description of head and neck schwannomas patients

| Location | Presenting signs and symptoms | Nerve of origin | No. of cases |
|---|---|---|---|
| *Head (53 patients)* | | | |
| 1. Oral cavity:<br>Tongue<br>ENT (1)<br>DENT(2) | <br><br>Tongue mass<br>Unhealed tongue ulcer | <br><br>CN V<br>CN V | <br><br>2<br>1 |
| Mandibular body<br>DENT(1) | <br>Jaw bone mass | <br>CN V | 1 |
| Buccal mucosa<br>DENT(1) | <br>Buccal mucosa mass | <br>CN V | 1 |
| 2. Brain:<br>C-P angle<br>NS(32) | Progressive blurred vision | CN III | 1 |
| | Progressive hearing impairment | CN VIII | 13 |
| | Hearing impairment & facial numbness | CN VIII (CN VII involved) | 4 |
| | Tinnitus | CN VIII (CN VII involved) | 1 |
| | Tinnitus | CN VIII | 2 |
| | Unstable gait | CN VIII | 4 |
| | Headache | CN VIII | 2 |
| | Dizziness & inttermettant vomit | CN VIII | 4 |
| | Deviation of tongue | CN XII | 1 |
| Others<br>NS(11) | Diplopia | CN II | 1 |
| | Diplopia | CN III | 1 |
| | Facial numbness | CN V | 1 |
| | Blurred vision | CN VII | 1 |
| | Vertigo, tinnitus | CN VII | 1 |
| | Hearing impairment | CN VIII | 2 |
| | Unsteady gait | CN VIII(CN VII involved) | 1 |
| | Facial numbness | CN VIII(CN VII involved) | 1 |
| | Blurred vision | CN X | 1 |
| | Body numbness | Cervical plexus | 1 |
| 3. Eye<br>NS(2) | Progressive blindness | CN II | 1 |
| | Diplopia | CN II | 1 |
| 4. Scalp<br>NS(2)& PS(1) | <br>Scalp mass | <br>Cervical plexus | 3 |
| *Neck (16 patients)* | | | |
| 1. Pharynx<br>ENT(2) | Neck mass | CN X | 1 |
| | Neck mass | Cervical plexus | 1 |
| 2. Submandibular gland<br>ENT(2) | <br>Neck mass | <br>CN V | 2 |
| 3. Spinal cord<br>NS(7) | Neck pain | Cervical plexus | 2 |
| | Weakness & numbness of limb | Cervical plexus | 1 |
| | Nausea & vomit | Cervical plexus | 1 |
| | Neck pain | Brachial plexus | 2 |
| | Leg weakness | Brachial plexus | 1 |
| 4. Neck soft tissue<br>ENT(5) | Neck mass | CN X | 3 |
| | Neck mass | Cervical plexus | 1 |
| | Neck mass | Cervical sympathetic chain | 1 |

Abbreviation: ENT (Ear, Nose and Throat Department), DENT (Dentistry Department), NS (Neurosurgery Department), PS (Plastic Surgery Department), CN (cranial nerve and sequential numbers in medical nerve term after it).

Table 3. Seven cases of Patient's recurrent date and recurrent side of head and neck schwannomas

| Age & Sex | 1st operation date | 1st diagnosis & treatment | Nerve of origin | Recurrent date (time interval) | Recurrent diagnosis & treatment |
|---|---|---|---|---|---|
| 44 Female Lin, MW | 2007/06/27 | Left C-P angle neurilemmoma, s/p left retromastoid suboccipital craniotomy with removal of tumor. | VIII acoustic nerve (VII facial nerve was compressed) | 2010/03/25 (31 months) | Recurrent neurilemmoma in the left IAC, s/p Gamma Knife radiosurgery. |
| 41 Male Shiao, W T | 2007/04/26 | Right C-P angle neurilemmoma, s/p right retromastoid suboccipital craniectomy with removal of tumor. | VIII acoustic nerve | 2009/06/22 (26 months) | Vestibular schwannoma over right C-P angle with recurrence ,s/p Gamma Knife radiosurgery |
| 52 Male Huang, SK | 2004/02/09 | left C-P angle, s/p left retromastoid suboccipital craniectomy with partial removal of tumor. | VII facial nerve, VIII acoustic nerve | 2004/4/28 (2 months) | Vestibular left C-P angle, s/p craniotomy with removal of tumor |
| 41 Female Xu, LQ | 2004/11/05 | left C-P angle, s/p Left suboccipital retromastoid craniotomy with removal of tumor. | VIII acoustic nerve | 2008/11/06 (36 months) | Recurrent left acoustic neurilemmoma, s/p Gamma Knife radiosurgery |
| 28 Male Lin,YS | 2007/08/20 | Left oculomotor nerve neurilemmoma, s/p left F-T craniotomy (keyhole approach) and removal of tumor | III oculomotor nerve | 2008/05/01 (8 months) | Recurrent neurilemmoma, left cavernous sinus, s/p craniotomy. |
| 18 Female Huang, SY | 2007/01/04 | Neurofibromatosis type 2 with bilateral acoustic neuroma, s/p gamma knife radiosurgery | VIII acoustic nerve | 2009/05/08 (28 months) | Left acoustic neuroma, s/p gamma knife. |

Abbreviation: s/p ( post-status), F-T (frontotemporal).

schwannoma over her scalp. By the way, her disease is often inherited due to a mutation of the NF2 gene.

Since schwannomas usually displace the nerve of origin as they grow, postoperative neural function can often be an achievable goal. Our study found that 15 patients (16.3%) were without any postoperative nerve injury, symptom, or sign. Table 4 displays the nerve injury symptom or sign after resection of schwannoma including multiple symptoms or signs. The most common sequelae after resection of head and neck schwannomas post-operative were facial numbness of 7 cases (10.0%). None of the schwannomas were malignant.

## Discussion

After resection of schwannomas, resolution rates were 59.4% and recurrence rates were 8.6% based on our analyses. Intracranial schwannomas showed a recurrence rate of 7.2% which was higher than the extracranial schwannomas recurrence rate of 1.4%.

The singular recurrent case was neurofibromatosis type 2 (defined as "Multiple Inherited Schwannomas, Meningiomas, and Ependymomas")—an inherited disease caused by mutations of the "Merlin" gene, NF2 in Chromosome 22. This gene can hasten the development of symmetric, non-malignant brain tumours in the region of the cranial nerve VIII. In this particular case, it was the auditory-vestibular nerve which transmits sensory information from the inner ear to the brain.

Sanna et al.[3] find the results and demonstrate the tumor size need not play an important role in the adoption for removal. On the contrary, the authors experience acquired over the years has revealed that large and giant tumors may be treated without sacrificing the cranial nerves and without dealing with problems of partial resection and the possible risks associated with a second procedure to remove the residual tumor[9,10]. Death after the giant acoustic neuromas surgery is generally dependent on occlusion of the anteroinferior cerebellar artery or brain stem trauma caused by surgical manipulations or perioperative bleeding[11]. The most serious postoperative complication in acoustic neuromas surgery is the development of hematoma in cerebellopontine angle. Because it causes a rapid worsening of the patient's state of consciousness, it is necessary to keep the patient under constant neurologic monitoring for the first 24 to 48 hours. Usually we awaken the patient to assess the level of consciousness and remove the endotracheal tube as soon as the operation is over.

In general, the rate of conservation of hearing in tumors larger than 2 cm is very low[12]. The retrosigmoid approach (RSA) is preferred by many neurosurgeons for the removal of acoustic neuromas surgery of any size. RSA points out there is only 4% of patients with tumors larger than 2 cm did success in hearing preservation but Ebersold et al.[13] did not find any success in tumors larger than 4 cm. A report from House Ear Institute mentions a higher incidence of meningitis in large tumors, probably because during operation on large tumors the meninges are exposed for a long time[14,15]. For this reason, Sanna et al.[3] usually administer postoperative antibiotic therapy (piperacillin, 2 g every 4 hours) for more than 48 hours if the intervention lasts longer than 8 hours.

Once intracranial schwannomas recur, usually the subsequent treatment is Gamma Knife

Table 4. Postoperative sequelae and resolution after resection of schwannoma

| Location | Preoperativ signs and symptoms | Post operative sequelae | Resolution* |
|---|---|---|---|
| *Head (53 patients)* | | | |
| 1.Oral cavity: | | | |
| Tongue | | | |
| CN V | Tongue mass(2) | | 2/2 |
| CN V | Unhealed tongue ulcer(1) | | 1/1 |
| Mandibular body | | | |
| CN V | Jaw bone mass | Operation site numbness | 0/1 |
| Buccal mucosa | | | |
| CN V | Buccal mucosa mass | | 1/1 |
| 2. Brain: | | | |
| C-P angle | | | |
| CN III | Progressive blurred vision(1) | Facial numbness(1) | 1/1 |
| CN VIII | Progressive hearing impairment(13) | Facial numbness(1)<br>Deafness(1) | 5/13 |
| CN VIII (CN VII involved) | Hearing impairment & facial numbness(4) | Facial numbness(1) | 2/4 |
| CN VIII (CN VII involved) | Tinnitus(1) | | 0/1 |
| CN VIII | Tinnitus(2) | | 1/2 |
| CN VIII | Unstable gait(4) | Facial palsy(1) | 2/4 |
| CN VIII | Headache(2) | Facial palsy(1)<br>Facial numbness(1) | 1/2 |
| CN VIII | Dizziness & inttermettant vomit(4) | | 3/4 |
| CN XII | Deviation of tongue(1) | | 0/1 |
| Others | | | |
| CN II | Diplopia(1) | Facial numbness(1) | 0/1 |
| CN III | Diplopia(1) | | 1/1 |
| CN V | Facial numbness(1) | | 0/1 |
| CN VII | Blurred vision(1) | | 1/1 |
| CN VII | Vertigo, tinnitus(1) | | 1/1 |
| CN VIII | Hearing impairment(2) | Facial numbness(1) | 0/2 |
| CN VIII (CN VII involved) | Unsteady gait(1) | | 1/1 |
| CN VIII (CN VII involved) | Facial numbness(1) | | 0/1 |
| CN X | Blurred vision(1) | | 1/1 |
| Cervical plexus | Body numbness(1) | | 1/1 |
| 3. Eye | | | |
| CN II | Progressive blindness(1) | | 0/1 |
| CN II | Diplopia(1) | | 0/1 |
| 4. Scalp | | | |
| Cervical plexus | Scalp mass(3) | | 3/3 |

Table 4. Postoperative sequelae and resolution after resection of schwannoma (Continue)

| *Neck (16 patients)* | | | |
|---|---|---|---|
| 1. Pharynx | | | |
| CN X | Neck mass(1) | Pain when stimulated | 1/1 |
| Cervical plexus | Neck mass(1) | Neck numbness(1) | 1/1 |
| 2. Submandibular gland | | | |
| CN V | Neck mass(2) | | 2/2 |
| 3. Spinal cord | | | |
| Cervical plexus | Neck pain(2) | Scalp numbness(1) | 1/2 |
| Cervical plexus | Weakness & numbness of left limb(1) | | 1/1 |
| Cervical plexus | Nausea & vomit(1) | Palm numbness(1) | 1/1 |
| Brachial plexus | Neck pain(2) | | 1/2 |
| Brachial plexus | Leg weakness(1) | | 1/1 |
| 4. Neck soft tissue | | | |
| CN X | Neck mass(3) | Palm numbness(1) | 3/3 |
| Cervical plexus | Neck mass(1) | | 1/1 |
| CSC | Neck mass(1) | | 1/1 |

Abbreviation: CSC (cervical sympathetic chain).
Resolution definition: No tumor can be found at Post-operative MRI or CT scan follow-up (after 3 months).
Resolution*: resolution number of cases/all number of cases.

radiosurgery which we perform in our hospital. From our study, the most common sequelae after resection of head and neck schwannomas post-operative were facial numbness. We believe further investigation should be conducted to determine if any causes can be identified for the higher recurrence rates in brain schwannomas as compared to other sites.

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# 台中榮總頭頸部神經鞘瘤預後及復發分析：8年回溯性研究

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## 摘 要

緒論：本研究針對八年來中部某醫學中心所發生頭頸部神經鞘瘤(稱之史旺氏瘤等)之病患所產生部位、癥狀、所在神經叢、術後復元情形以及復發率加以研究。

材料與方法：本文在於探討台中榮民總醫院自2003年1月至2010年12月間回顧其診療紀錄完整之69名頭頸部神經鞘瘤患者。作者利用統計特性分析及比較分析法針對其腫瘤所在位置、不同的病癥、神經鞘瘤神經元起始來源，進而對於術後觀察神經受損情形至痊癒經歷期間以及腫瘤割除再復發之可能。旨在提供頭頸部神經鞘瘤病患及醫護相關人員參考與建議。

結果：本研究發現男性病患約佔45%，相較於女性55%略少，而年齡層由12歲至83歲分布甚廣，平均年齡爲47.84歲。在69位患者中53位診斷腫瘤部位有5位在口腔，32位在小腦橋腦角，2位在眼部以及3位在頭皮部位，此外16位患者則腫瘤長在頸部，其中最常見者爲聽覺神經鞘瘤。術後治療率約略達59.4%。此外，在6例顱內神經鞘瘤中有4例爲小腦橋腦角，基於這些顱內神經鞘瘤復發率佔8.6%，維護腫瘤所在的神經功能，一些腫瘤無法切除乾淨，因此研究中發現顱內神經鞘瘤復發率爲7.2%高於顱外復發率1.4%。

討論：本篇研究中發現術後治療率達59.4%，顱內神經鞘瘤復發率高於顱外復發率。神經鞘瘤成長緩慢，不僅包膜完整且邊界清楚，由於此類腫瘤成長不明顯導致病患就醫亦不積極。一般而言，依照其腫瘤所在位置選擇治療方式，在無妨礙神經功能之虞，通常選擇完整切除，當牽涉神經叢所在之腫瘤則以部份切除，搭配珈瑪刀(光子刀)爲最佳治療方式之選擇。

**關鍵詞**：頭頸部神經鞘瘤，復發率。

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