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Original Article* / *Çalışma - Araştırma

Extracranial head and neck schwannomas: A series of 14 patients

Ekstrakraniyal baş boyun schwannomları: 14 hastalık seri

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ABSTRACT

Objectives: This study aims to evaluate patients operated due to extracranial head and neck schwannomas.

Patients and Methods: A total of 14 patients (6 males, 8 females; mean age 42.6 years; range 7 to 73 years) who were treated for extracranial head and neck schwannomas between January 1995 and December 2015 were retrospectively reviewed. Demographic data, tumor location, surgery, and clinical characteristics of the patients were discussed in light of the literature.

Results: All patients were evaluated radiologically and with fine needle aspiration biopsy preoperatively. Total removal of the tumor was achieved in all patients. All tumors had benign character. Mean follow-up period was 14 months. Recurrence or malignancy was not detected in any patient.

Conclusion: Schwannomas may present in a variety of sites in the head and neck region, and cause usually non-specific symptoms. Therefore, the preoperative diagnosis requires a clinical suspicion. Preoperative magnetic resonance imaging is a useful and simple method for diagnosis.

Keywords: Head and neck neoplasms; nerve sheath neoplasms; neurilemmoma; Schwann cells.

ÖZ

Amaç: Bu çalışmada, ekstrakraniyal baş boyun schwannomları nedeniyle ameliyat edilen hastalar değerlendirildi.

Hastalar ve Yöntemler: Ekstrakraniyal baş boyun schwannomları nedeniyle Ocak 1995 - Aralık 2015 tarihleri arasında tedavi gören 14 hasta (6 erkek, 8 kadın; ort. yaş 42.6 yıl; dağılım 7-73 yıl) geriye dönük olarak incelendi. Hastaların demografik verileri, tümörün yerleşim yeri, cerrahisi ve klinik özellikleri literatür eşliğinde tartışıldı.

Bulgular: Tüm hastalar ameliyat öncesinde radyolojik olarak ve ince iğne aspirasyon biyopsisi ile değerlendirildi. Tüm hastalarda total tümör eksizyonu gerçekleştirildi. Tüm tümörler benign karakterde idi. Ortalama izlem süresi 14 ay idi. Hiçbir hastada nüks veya malignite saptanmadı.

Sonuç: Schwannomlar baş boyun bölgesinde farklı alanlarda ortaya çıkabilir ve genellikle non-spesifik semptomlara yol açar. Bu nedenle, tanı için klinik şüpheye ihtiyaç vardır. Ameliyat öncesi manyetik rezonans görüntüleme tanı için yararlı ve kolay bir yöntemdir.

Anahtar Sözcükler: Baş boyun neoplazmları; sinir kılıfı neoplazmları; nörilemmom; Schwann hücreleri.

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Neurogenic tumors of the head and neck are rare entities regarding presentation, diagnosis, and treatment.[1] These tumors consist of a group of miscellaneous neoplasms, including neurofibromas, schwannomas, granular cell myoblastomas, neurogenic sarcomas, malignant melanomas, and neuroepitheliomas.[2]

Schwannomas are benign, encapsulated and solitary tumors that arise from perineural Schwann cells. They constitute 5% of benign soft tissue tumors.[3,4] About 25-48% of them occur in the head and neck region. They are classified according to their nerve of origin and their location within the head and neck.[4,5] The most frequently involved cranial nerves are the trigeminal, abducens, facial, vestibular, vagus, glossopharyngeal, spinal accessory, and the hypoglossal nerves. Other frequent neural origins are the sympathetic chain and the brachial plexus.[2] These tumors compress rather than infiltrate their neural origin.[6] According to site, they may be intracranial, extracranial or transcranial.[1] Intraparotid, nasal/paranasal, mastoid, parapharyngeal, hypopharyngeal, tongue base or cervical locations are considered extracranial schwannomas.[1,2,4,7] Unusual extracranial locations include the esophagus, larynx, and thyroid gland.[2] In terms of neck site, extracranial head and neck schwannomas (ECHNS) can be medial or lateral. Medial tumors originate from the glossopharyngeal, vagus, accessory and hypoglossal nerves or the sympathetic chain. Laterally, they arise from cervical or brachial plexus.[8]

The current study focuses on extracranial schwannoma cases treated in our clinic in the last 20 years. Our series of extracranial head and neck schwannomas is the third and largest among the national Turkish literature.[9,10]

PATIENTS AND METHODS

This is a retrospective study conducted at department of Otorhinolaryngology of Dokuz Eylül University hospital. The medical records of 14 patients (6 males, 8 females; mean age 42.6 years; range 7 to 73 years) diagnosed with ECHNS and managed in our department from January 1995 to December 2015 were reviewed retrospectively. Patients who were followed up regularly after surgical or non-surgical treatments and volunteered for this study were included, whereas non-willing and not regularly controlled cases were excluded. Fourteen participants who met the inclusion criteria were discussed in this report. Gender, age, symptoms, diagnostic tools, tumor characteristics, treatment methods and outcomes were evaluated. This study obtained the approval of Local Institutional Review Board and Ethical Committee in accordance with Helsinki Declaration.

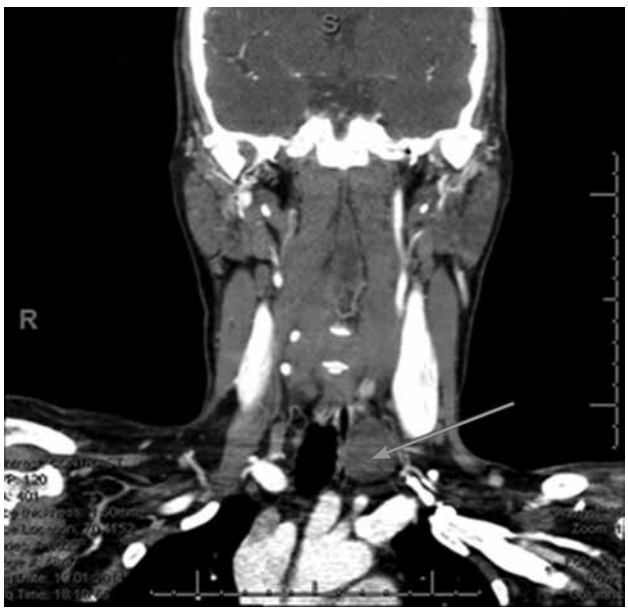

Figure 1. *Contrast-enhanced cervical computed tomography, coronal section shows the supraclavicular extracranial head and neck schwannomas of patient number four; note the well-circumscribed and non-infiltrative mass with lower signal density than surrounding muscles (gray arrow).*

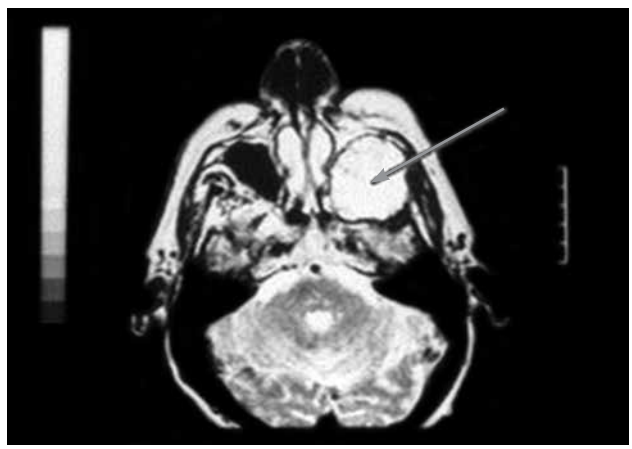

Figure 2. *Contrast-enhanced maxillofacial computed tomography, axial section shows the left maxillary sinus extracranial head and neck schwannomas of patient number eight; note the contrast-enhanced tumor (gray arrow).*

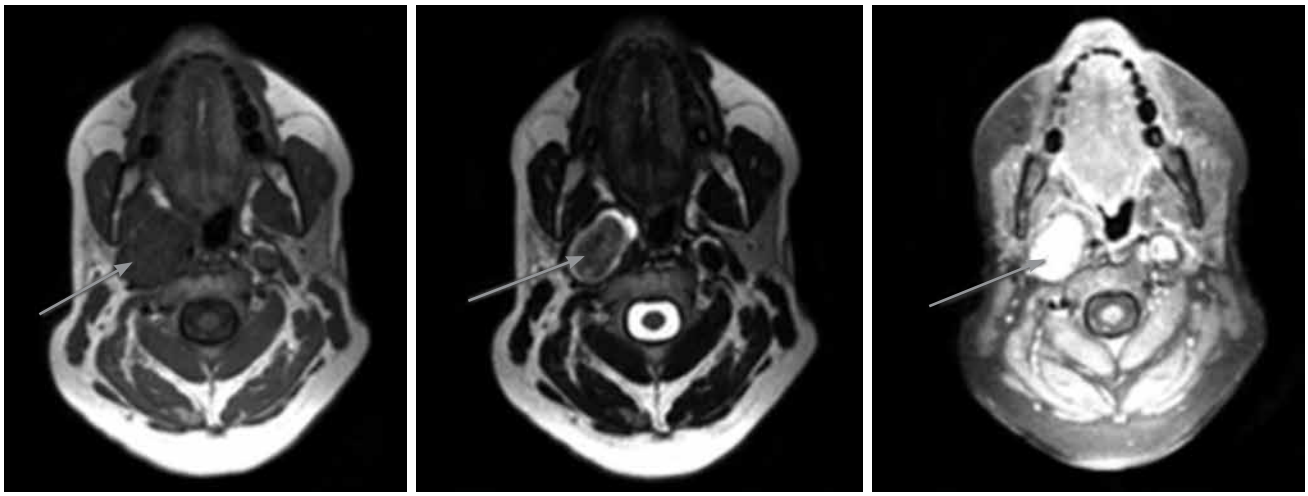

Figure 3. *Axial T_1-weighted, T_2-weighted, and contrast-enhanced T_1-weighted cervical magnetic resonance imaging sections demonstrate the parapharyngeal extracranial head and neck schwannomas of patient number three; note the homogenous intensity, iso-intense, well-shaped, non-vascular soft tissue mass in the pharapharyngeal space on T_1-weighted; peripheral hyper-intensity of the same mass on T_2-weighted; and contrast-enhancement on contrast-enhanced T_1-weighted images (gray arrows).*

RESULTS

Eight patients presented with a painless neck mass. Among them, tumor origins were the vagal nerve in three, the cervical plexus in two, the hypoglossal nerve in one, and the cervical sympathetic chain in one patient. Other tumor sites where the origin of nerve could not be determined clearly were the parotid gland in two patients, and pyriform sinus, tongue, nose, and maxillary sinus in one patient each.

The routine preoperative diagnostic test battery included ultrasonography (US), computed tomography (CT), magnetic resonance imaging (MRI), and fine needle aspiration biopsy (FNAB). Ultrasonography was employed for neck masses as an initial radiological diagnostic tool. All patients' US results indicated non-specific masses. Further evaluation was provided by contrast-enhanced CT and MRI. Contrast-enhanced CT showed hypodense and well-limited lesions with peripheral contrast

Table 1. The number, age, gender, major symptoms and tumor location of the patients are presented

Case	Age/Gender	Location of the tumor and nerve origin	Symptom	CT	FNAB
1	13/M	Tongue	Swelling in tongue	Schwannoma	Schwannoma
2	7/F	Neck (cervical sympathetic chain)	Neck mass	Tumor	Nondiagnostic
3	34/F	Parapharyngeal space	Neck mass	Tumor	Schwannoma
4	47/F	Neck (cervical plexus)	Neck mass	Tumor	Schwannoma
5	51/M	Right pyriform sinus	Dysphagia	Tumor	Nondiagnostic
6	73/M	Neck (N. vagus)	Neck mass	Schwannoma	Schwannoma
7	58/F	Nose	Stuffiness	Tumor	Schwannoma
8	66/M	Left maxillary sinus	Malar swelling	Tumor	Schwannoma
9	52/F	Right parotid gland (N. facialis)	Intraparotid mass	Schwannoma	Schwannoma
10	41/F	Right parotid gland (N. facialis)	Intraparotid mass	Tumor	Nondiagnostic
11	17/F	Neck (cervical plexus)	Neck mass	Tumor	Nondiagnostic
12	48/M	Neck (N. vagus)	Neck mass	Tumor	Nondiagnostic
13	30/M	Neck (N. hypoglossus)	Neck mass	Tumor	Nondiagnostic
14	60/F	Neck (N. vagus)	Neck mass	Tumor	Schwannoma

CT: Computed tomography; FNAB: Fine needle aspiration biopsy.

Table 2. The number, tumor location, nerve origin, perioperative and postoperative neural status of the patients are presented

Case	Location of the tumor and nerve origin	Preoperative neural status	Postoperative neural status	Neural outcome six months after operation
1	Tongue	Normal	Normal	Normal
2	Neck (cervical sympathetic chain)	Normal	Paralysis	Paralysis
3	Parapharyngeal space	Normal	Normal	Normal
4	Neck (cervical plexus)	Normal	Horner's syndrome (ptosis)	Ptosis
5	Right pyriform sinus	Normal	Normal	Normal
6	Neck (N. vagus)	Normal	Vocal cord paralysis (severe aspiration)	Improved and well-tolerated after medialization thyroplasty
7	Nose	Normal	Normal	Normal
8	Left maxillary sinus	Normal	Normal	Normal
9	Right parotid gland (N. facialis)	Normal	House-Brackman grade 2 facial paralysis	Normal (improved)
10	Right parotid gland (N. facialis)	Normal	House-Brackman grade 4 facial paralysis	House-Brackman grade 3 facial paralysis (improved)
11	Neck (cervical plexus)	Normal	Normal	Normal
12	Neck (N. vagus)	Normal	Vocal cord paralysis (hoarseness)	Vocal cord paralysis
13	Neck (N. hypoglossus)	Normal	Paralysis	Normal (improved)
14	Neck (N. vagus)	Normal	Vocal cord paralysis (hoarseness)	Vocal cord paralysis

enhancement (Figure 1 and 2). Contrast-enhanced MRI was also applied to all patients (Figure 3). None of the MRI images were demonstrative for identification of nerve origin.

All patients underwent FNAB after the radiologic examination. Our results showed that only eight cases (57%) displayed a specific diagnosis of schwannoma (Table 1). None of the patients experienced either a neural or a non-neural complication due to the FNAB. After the preoperative investigations, all patients underwent surgery. The diagnosis of schwannoma was confirmed by histopathologic examination. Total removal of the tumor was achieved in all subjects, and none of the cases had a recurrence during the follow-up course (mean 14 months).

A few expected postoperative complications like hoarseness, aspiration, incomplete facial paralysis and Horner's syndrome arose. Patient number six presented with severe aspiration after surgery and was treated with medialization thyroplasty. The neural deficits encountered in the other patients are noted in Table 2. In a seven-year-old patient with neck schwannoma, bilateral vestibular schwannomas were coincidentally detected in the internal acoustic meatus. She was followed up with the diagnosis of neurofibromatosis type 2 by Pediatric Oncology.

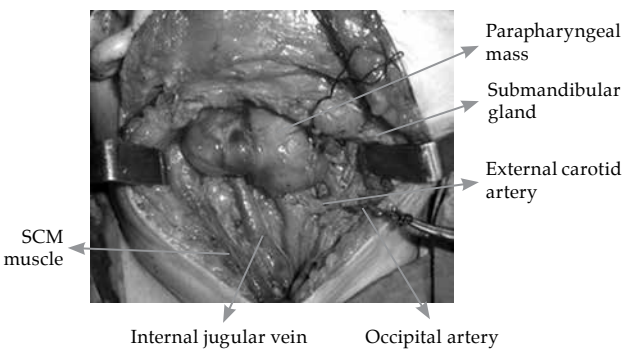

Figure 4. *Intraoperative photo of patient number three showing parapharyngeal extracranial head and neck schwannomas and specific anatomical landmarks (gray arrows).* *SCM: Sternocleidomastoid.*

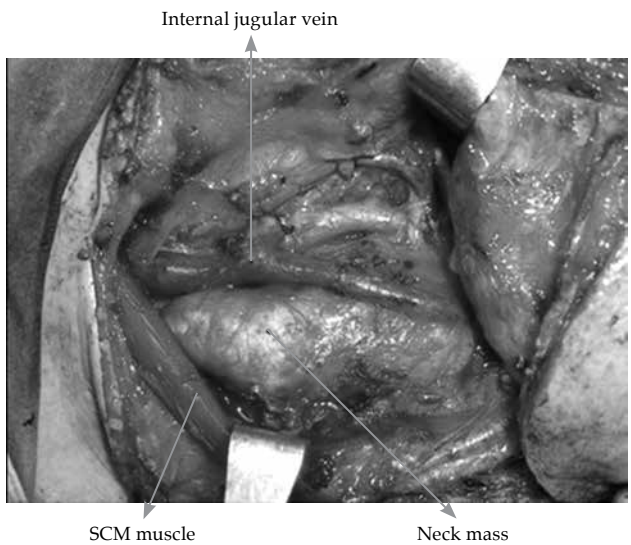

Figure 5. *Intraoperative photo of patient number six showing cervical extracranial head and neck schwannomas and specific anatomic landmarks (gray arrows).* *SCM: Sternocleidomastoid.*

All of the tumors were benign. Demographic data, major symptoms, tumor locations of the cases are presented in Table 1. Preoperative and postoperative neural outcome is summarized in Table 2. Perioperative views of patient number three and six are shown in Figure 4 and 5, respectively.

DISCUSSION

Schwannomas may occur at any age but are most common between the ages of 20 and 50 with no apparent predilection for race or sex.[4,7] Although originating most frequently from sensory nerves, head and neck schwannomas can occur in all cranial nerves except the olfactory or optic nerves, which are extensions of white matter. Schwannomas involving cranial nerve eight are located intracranially and comprise 90% of intracranial schwannomas. Trigeminal, glossopharyngeal, accessory and vagal nerve schwannomas can be located both intra- and extracranially.[1]

Schwannomas usually present as slow growing solitary masses and are usually palpated incidentally.[5] Symptoms like pain and neurological deficits are related to malignancy. Malignant schwannomas are rare and constitute only 5% of all soft tissue tumors.[11] A complete clinical history and physical examination of the whole body is the first step of diagnosis. There are no specific signs or symptoms since they depend fundamentally on the location of the tumor or the involved neural structures.[2] Trigeminal schwannomas have been reported to occur in the Gasserian ganglion presenting with facial numbness, trigeminal neuralgia, weakness of masticatory muscles, proptosis, limitation of ocular movements, visual field defects and in the nasal and paranasal region with symptoms of nasal obstruction, epistaxis, and hyposmia. Facial schwannomas involving the extratemporal facial nerve may present with facial palsy. Symptoms of vagal or glossopharyngeal schwannomas include hoarseness, dyspnea, dysphagia, cough, and syncope.[2] The overall incidence of motor weakness has been reported to be 41%.[12] None of our cases had a neural deficit initially (Table 2).

Ultrasonography, contrast-enhanced CT, MRI, and FNAB are useful in the preoperative and differential diagnosis.[12] The US primarily suggests a hypoechoic, homogeneous mass including cystic and lobulated components but has poor diagnostic sensitivity.[3,13] In our research, all neck masses were preoperatively examined with a neck US. Computed tomography and MRI were performed in all cases. Characteristic findings on CT scans are well-circumscribed and non-infiltrative mass with lower signal density than surrounding muscles. Homogeneous contrast enhancement may be seen. However, the diagnostic sensitivity of CT has been stated as 35.7%.[5] Our CT results indicated a poorer diagnostic sensitivity (three patients, 21%) despite the nonspecific appearance on sonography and CT scans; MRI would suggest the diagnosis, revealing low signal intensity on T_1-weighted images and high signal intensity on T_2-weighted images.[11] The diagnostic sensitivity of MRI has been stated as 80%.[13] In our series MRI findings suggested the diagnosis of schwannoma in all patients (100%). Fine needle aspiration biopsy also plays a role in differential diagnosis but is not sensitive enough.[13] Additionally, facial schwannomas, to avoid the risk of facial paralysis, are not usually subjected to FNAB.[2] In our data, FNAB provided a specific diagnosis in eight cases (57%). Open biopsy may provide higher diagnostic sensitivity about 86%, but is inadequate regarding patient compliance.[14] Accordingly, the definitive diagnosis is established by histopathologic examination of the surgical specimens.[5,13] Histologically, schwannomas have characteristic growth patterns, named Antoni A and Antoni B types, which reveal the clear-cut diagnosis.[5] The use of immunohistochemical S-100 protein also assists in identifying Schwann cells.[1]

The consensus on the management of ECHNS consists of whole tumor removal with a favorable functional outcome. Therefore, treatment of schwannomas has been described as a cautious extracapsular dissection or even enucleation of the tumor from the nerve to preserve the function and integrity of the nerve for benign schwannomas.[15] The relative avascular nature of the tumor allows for dissection within the capsule and separation from the parent nerve, however, because the neural fascicles splay over the tumors, it is frequently impossible to preserve the associated nerve.[2] The total sacrifice rate of the associated nerve is 56% in the literature, whereas 64% of the rest had permanent and 29% had transient neural deficits.[16] Thus, the patient operated on for an

extracranial head and neck schwannoma may ultimately experience postoperative permanent or transient neurological morbidities such as dysphagia, Horner's syndrome, and hoarseness. Of our cases, 57% experienced neural deficits immediately after surgery. Four improved after six months and one required a medialization thyroplasty.

Because of the surgical morbidity, surgeons have advocated a "watch and wait approach" before removing the tumor until it causes significant symptoms.[8,11,17,18] Yafit et al.[17] defined a management algorithm for ECHNS. According to this algorithm, "being symptomatic" is the principal determinant in the transition from observation to treatment. Asymptomatic benign lesions, as long as they remain stable are good candidates for observation. Symptomatic cases were committed to surgery. During surgery, despite the low malignant degeneration and recurrence rate, it should be important to remove the whole mass.[11,13] Extracranial head and neck schwannomas ineligible for surgical treatment should be considered for radiotherapy.[17] Recent data supports radiotherapy in schwannomas originating from cranial nerves III-VI for non-surgical candidates resulting in local control rate of 90-100%.[19,20] Close follow-up is mandatory.[11] Our clinical experience resulted in no recurrence during the mean monitoring time.

In conclusion ECHNS may present in a variety of sites in the head and neck region. According to our results, the most common symptom was a painless neck mass. In our series, we performed a diagnostic radiological evaluation battery including US, CT, and MRI in a consecutive manner but could be able to verify the diagnosis only using MRI (100% diagnostic accuracy). Further, MRI demonstrated a better diagnostic confirmation than FNAB in our series.

Another important result of our case series was demonstrating the safety of preoperative FNAB. We used FNAB for all participants and did not experience any neural or non-neural complications. The overall success rate of our surgical procedures was quite satisfactory and the final neural outcome was compatible with the literature. Based on our clinical experience, we recommend that surgeons, who are in charge of carrying out such surgical procedures, be well experienced in identifying common presentation characteristics and surgical procedures of these tumors, and dealing with potential postoperative complications.

We believe that this report, which discusses a wide variety of schwannoma cases, will be a useful reminder for head and neck surgeons.

Declaration of conflicting interests

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