Kulak Burun Bogaz Ihtis Derg 2008;18(4):253-256

CASE REPORT

A case of greater occipital nerve schwannoma causing neuralgia

Nöraljiye yol açan büyük oksipital sinir schwannoması: Olgu sunumu

Ahmet URAL, M.D., Alper CEYLAN, M.D.,[1] Erdoğan İNAL, M.D.,[1] Fatih ÇELENK, M.D.[1]

Schwannomas are common tumors of the head and neck region, but they rarely develop in the suboccipital region. A 34-year-old woman presented with a left suboccipital mass causing local pain and neck stiffness. Following physical examination, fine needle aspiration biopsy was performed, which revealed benign cytology. In the light of preoperative radiologic examination and intraoperative observations, the lesion was thought to originate from the greater occipital nerve. Following complete excision of the lesion, histopathological diagnosis was made as schwannoma. Apart from mild neck stiffness and transient local paresthesia in the early postoperative period, no complications were seen during a two-year follow-up period.

Key Words: Neuralgia/etiology; neurilemmoma/complications/surgery; peripheral nervous system neoplasms; spinal nerves.

Schwannomalar baş ve boyun bölgesinin sık görülen tümörleri olmasına karşın, suboksipital bölgede görülmeleri nadirdir. Otuz dört yaşında kadın hasta, sol suboksipital bölgede kitle, lokal ağrı ve boyunda tutukluk yakınmalarıyla kliniğimize başvurdu. Muayene sonrası yapılan ince iğne aspirasyon biyopsi sonucu benign sitoloji olarak bildirildi. Radyolojik değerlendirme ve ameliyat sırası bulgular ışığında majör oksipital sinirden kaynaklandığı düşünülen kitle tam olarak eksize edildi ve histopatolojik tanı schwannoma olarak bildirildi. Ameliyat sonrası erken dönemde boyunda hafif tutukluk ve geçici lokal parestezi dışında, hastanın iki yıllık izlemi sırasında başka sorun görülmedi.

Anahtar Sözcükler: Nöralji/etyoloji; nörilemmom/komplikasyon/cerrahi; periferik sinir sistemi neoplazileri; spinal sinir.

Schwannomas are benign lesions that originate from neural crest cells. They are frequently encountered in the head and neck region and may originate from any peripheral, cranial, or spinal nerves.[1,2] They may cause pain or paresthesia via compression to the neighboring organs, but they may also appear as solitary masses.[2,3] Schwannomas of submandibular, retromaxillary and parapharyngeal regions have been reported.[4,5] A schwannoma presenting with occipital (Arnold) neuralgia has been described recently.[6] We reported a rare case of schwannoma in the form of a suboccipital mass causing neck stiffness and local pain.

CASE REPORT

A 34-year-old woman presented with an insidiously growing left suboccipital mass causing restricted cervical movements and cervicalgia. The lesion was reported to exist for 10 years and it exhibited a rapid growth in recent years. On physical examination, a smooth, semi-mobile and firm mass, about 4x4 cm in size, was determined. Her past medical history was

◆ *Department of Otolaryngology, Atatürk Training and Research Hospital (Atatürk Eğitim ve Araştırma Hastanesi Kulak Burun Boğaz Hastalıkları Kliniği), Ankara; [1]Department of Otolaryngology, Medicine Faculty of Gazi University (Gazi Üniversitesi Tıp Fakültesi Kulak Burun Boğaz Hastalıkları Anabilim Dalı), Ankara, Turkey.*

◆ *Received - December 4, 2006 (Dergiye geliş tarihi - 4 Aralık 2006). Request for revision - January 23, 2008 (Düzeltme isteği - 23 Ocak 2008). Accepted for publication - January 25, 2008 (Yayın için kabul tarihi - 25 Ocak 2008).*

◆ *Correspondence (İletişim adresi): Dr. Ahmet Ural. Mamak Cad. No: 43/1, 06340 Demirlibahçe, Ankara, Turkey. Tel: +90 312 - 291 25 25 / 3618 Fax (Faks): +90 312 - 284 78 07 e-mail (e-posta): ahmetural2001@yahoo.com*

uneventful. The rest of ENT examination and systemic examination were normal. There was no family anamnesis for neurofibromatosis. Ultrasonography revealed a left suboccipital, solid, hypoechoic mass, 39x19 mm in size, located between the muscle layers. Computed tomography (CT) showed a mass measuring 4 cm in diameter between the rectus capitis and semispinalis capitis muscles at the level of the 3rd to 6th cervical vertebrae. The lesion seemed not to cause any destruction to the neighboring tissues. Magnetic resonance imaging (MRI) showed a mass, 3.5x4x4 cm in size, at the level of the 3rd to 5th cervical vertebrae between the paraspinal muscle layers. The tumor had a cystic and necrotic central component and was radiopaque after the injection of radiocontrast material. Despite compression to the surrounding muscle tissue, no true invasion was evident (Fig. 1). These findings were interpreted to be consistent with a soft tissue sarcoma. Fine needle aspiration biopsy revealed benign cytology.

The left suboccipital lesion was completely excised under general anesthesia. The lesion was accessed through a horizontal external incision and the tumor was dissected readily via blunt dissection from the surrounding structures. The lesion appeared to be originating from the greater auricular nerve. Histopathological examination revealed an Antoni A type schwannoma (Fig. 2). Postoperatively, except for transient paresthesia in the left suboccipital region and minimal neck stiffness, no complications were encountered. There were no recurrences in the third postoperative year.

DISCUSSION

Neurogenic tumors constitute only a small portion of head and neck neoplasms.[7] In this heterogeneous tumor group, neurofibromas, schwannomas, and neurinomas make up the benign subgroup, while neurogenic sarcomas, malignant schwannomas, and neuroepitheliomas represent malignant neurogenic tumors.[7] Schwannomas (neurilemmoma, neurinoma) are solitary and encapsulated tumors which originate from the schwann cell sheath.[8] They are more commonly encountered in women and between the 3rd and 6th decades.[1,5] They represent the most common benign tumors of the neural sheath.[6] As well as the cranial and spinal nerve roots, they may originate from the cervical sympathetic chain or cutaneous and muscular branches of the cervical and brachial plexus.[6,7,9-11]

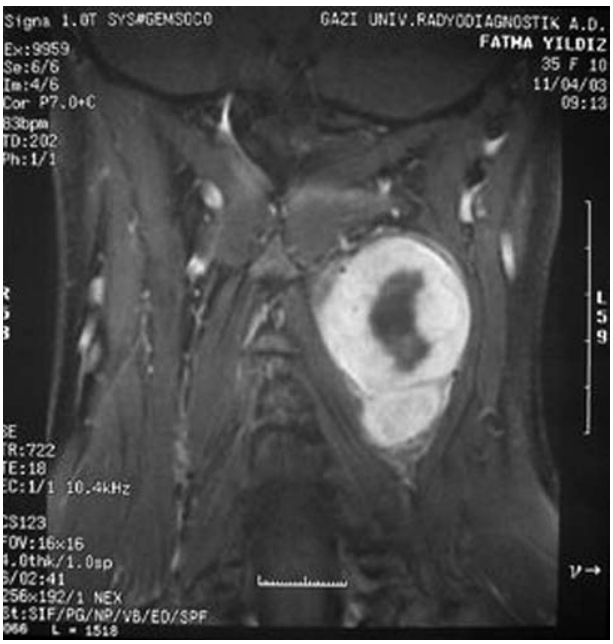

Fig. 1. *The MRI section demonstrating the location of the suboccipital schwannoma and its relationship with surrounding structures.*

Despite being capsulated and well-circumscribed, they may cause compression on the surrounding tissues due to insidious growth pattern. Mostly they are noninvasive, but infiltrative growth patterns may also be seen.[6]

In our patient, the tumor typically originated from the level of the second cervical vertebra, which represents the site of origin for the greater occipital (Arnold) nerve. This nerve is the posterior root of the second cervical vertebra and it receives

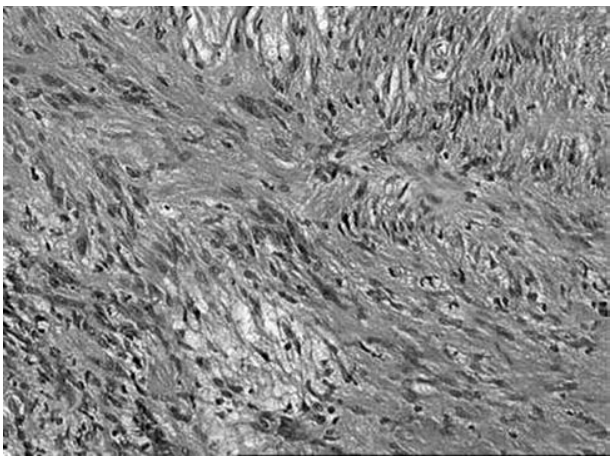

Fig. 2. *Microscopic appearance of Antoni type A schwannoma (H-E x 100).*

concomitant branches from the 1st and 3rd cervical nerves. It innervates paraspinal muscles, especially the semispinalis capitis muscle, and contributes to the cutaneous innervation of the head and neck skin via the posterior cervical plexus. Owing to its close proximity to the muscles, tendons, and neurovascular structures, schwannomas involving this nerve may result in local irritation and pain besides the mass effect. This situation is possibly due to the predominance of Pacinian corpuscles and vasa nervorum in the epi- and endoneural compartments which facilitate the existence of sense of pain due to traction and compression.[6]

These tumors are classified into two groups according to their histopathological appearances: *(i)* Antoni A tumors have a hypercellular and compact texture. The eosinophilic band between two nuclear rows is called as 'Verocay bodies'. The tumor in our patient was reported to be an Antoni A schwannoma. *(ii)* Antoni B lesions have a hypocellular and loose connective tissue. As found in our case, a cystic component was reported in 40% of spinal schwannomas, possibly due to hyalinization and thrombosis of vascular supply of the tumor.[6,12] This may be a valuable finding of MRI to discriminate the lesion from lipoma. The differential diagnosis should involve metastatic and reactive lymphadenopathies, lipomas, and tumors originating from muscle tissue.[12] Rarely, malignant transformation is possible in these tumors and is related with von Recklinghausen disease (type 2 neurofibromatosis). Hence, investigation into family history is as important as careful physical examination.[6]

Ultrasonography and CT scans may provide useful information for head and neck schwannomas, but MRI is the most valuable imaging modality.[13] The relationship between the tumor and neighboring tissue is of vital importance for planning surgical intervention.[6] Surgery is the mainstay of treatment and fine needle aspiration biopsy is usually recommended prior to operation.[13] However, the precise histopathological diagnosis may sometimes be possible only after total excision of the lesion.[14]

The aim of treatment is to perform total excision of the tumor without damaging nerve functions. As in our case, this can be achieved readily in small- and medium-sized and well-capsulated tumors. Consistent with our intraoperative observations, schwannoma tends to show an extrinsic outgrowth pattern from the nerve bundles from which it originates.[15] If infiltration to the surrounding structures is observed in a benign tumor, a partial extirpation may be preferred. However, this can only be feasible in some small lesions. In contrary, for malignant tumors, complete excision is required even at the expense of damage to nerve function.[6] Abramowitz et al.[16] recommended preoperative embolization of head and neck schwannomas, whereas Hood et al.[9] reported safe surgical intervention in their cases.

To our knowledge, only one case has been reported on greater occipital nerve schwannoma with neuralgia.[6] Clinical characteristics of our case were unusual in several aspects, such as anatomic location, the nerve of origin, and presentation. This seems to be the second reported case of greater occipital nerve schwannoma with neuralgia.

In conclusion, schwannomas must be kept in mind in the differential diagnosis of patients with a suboccipital mass and localized pain or neuralgia. Treatment is based on complete excision of the tumoral mass.

REFERENCES

1. Zachariades N. Schwannoma of the oral cavity. Review of the literature and report of a case. J Oral Med 1984;39:41-3.
2. Williams HK, Cannell H, Silvester K, Williams DM. Neurilemmoma of the head and neck. Br J Oral Maxillofac Surg 1993;31:32-5.
3. Yusuf H, Fajemisin OA, McWilliam LJ. Neurilemmoma involving the maxillary sinus: a case report. Br J Oral Maxillofac Surg 1989;27:506-11.
4. Gibbons SD, Wiesenfeld D, Millar H, Busmanis IA. Removal of a retromaxillary schwannoma via a temporal approach. J Oral Maxillofac Surg 1991;49:191-3.
5. Bradley N, Bowerman JE. Parapharyngeal neurilemmomas. Br J Oral Maxillofac Surg 1989;27:139-46.
6. Ballesteros-Del Rio B, Ares-Luque A, Tejada-Garcia J, Muela-Molinero A. Occipital (Arnold) neuralgia secondary to greater occipital nerve schwannoma. Headache 2003;43:804-7.
7. Benzoni E, Cojutti A, Intini S, Uzzau A, Bresadola F. Schwannoma of the sympathetic cervical chain presenting as a lateral cervical mass. Tumori 2003;89:211-2.
8. Zachariades N, Skoura C, Papageorgiou G, Chrissomali E. Giant ancient neurilemmoma of the cervical region: report of case. J Oral Maxillofac Surg 2001;59:668-72.
9. Hood RJ, Reibel JF, Jensen ME, Levine PA. Schwannoma of the cervical sympathetic chain. The Virginia experience. Ann Otol Rhinol Laryngol 2000;109:48-51.
10. Hirabayashi S, Sakurai A, Fukuda O. Neurilemoma of the ansa cervicalis. Plast Reconstr Surg 1987;79:809-11.
11. Bektas D, Caylan R. Bilateral hypoglossal schwannoma: a radiologic diagnosis. Kulak Burun Bogaz Ihtis

Derg 2004;12:45-7.
12. Guerra MF, Campo FR, de Artinano FO, Perez JS. Schwannoma of the brachial plexus: an unusual cause for a cervical mass. J Oral Maxillofac Surg 2001;59:942-4.
13. Keleş E, Yalçın Ş, Sakallıoğlu Ö, Dağlı AF. Üst servikal sempatik zincirden kaynaklanan periferik parafarengeal schwannoma: olgu sunumu. KBB-Forum 2006;5:54-7.
14. Colreavy MP, Lacy PD, Hughes J, Bouchier-Hayes D, Brennan P, O'Dwyer AJ, et al. Head and neck schwannomas - a 10 year review. J Laryngol Otol 2000;114:119-24.
15. Nuss DW, Battalora PA. Pathologic quiz case 2. Schwannoma of the cervical plexus. Arch Otolaryngol Head Neck Surg 1990;116:224-5.
16. Abramowitz J, Dion JE, Jensen ME, Lones M, Duckwiler GR, Vinuela F, et al. Angiographic diagnosis and management of head and neck schwannomas. AJNR Am J Neuroradiol 1991;12:977-84.