

Multiple Schwannoma of the Parapharyngeal Space

Parafaringeal Bölgenin Multipl Schwannomu

Hanifi KURTARAN, MD, Türker YILMAZ MD, Betül KAYMAKCI, MD, Nebil ARK, MD,
Davut AKTAŞ, MD, Cemil MUTLU, MD

Fatih University Faculty of Medicine, Department of Otorhinolaryngology Head and Neck Surgery, Ankara

ABSTRACT

A very rarely seen case of multiple schwannoma in parapharyngeal space is presented. A fifty-four year old male patient admitted to the otorhinolaryngology clinic with complaints of painless swelling in the neck and dysphagia. Physical examination revealed a solid tumor in the posteroinferior part of the right parotid gland with irregular borders. The same tumor could be seen intraorally, displacing the ipsilateral tonsil medially and the uvula contralaterally. In computerized tomographic examination multiple masses were detected in the right parapharyngeal space, which were interpreted as conglomerated lymphadenopathies. The masses were totally excised by using modified "Blair incision" and open cervical approach. Pathological diagnosis of the mass was reported as "multiple schwannoma". After two years of follow up no recurrence has been observed.

Keywords

Neurilemmoma, head and neck neoplasms

ÖZET

Parafaringeal bölgede çok nadir görülen bir multipl schwannom vakası sunuldu. Ellidört yaşında erkek hasta boyunda ağrısız şişlik ve yutma güçlüğü şikayetleri ile kulak burun boğaz kliniğimize başvurdu. Hastanın fizik muayenesinde; sağ parotis glandı posterior inferior komşuluğunda düzensiz sınırları olan solid kitle mevcuttu. Aynı kitleye ağız içinden bakıldığında; kitlenin tonsili mediale, uvulayı karşı tarafa doğru ittiği görüldü. Bilgisayarlı tomografik incelemede; sağ parafaringeal bölgede multipl kiteller mevcuttu ve bu kiteller konglomere lenfadenopati olarak yorumlandı. Modifiye "Blair insizyonu" ve açık servikal yaklaşım yoluyla kiteller total olarak çıkarıldı. Kitlenin histopatolojik tanısı "multipl schwannom" olarak rapor edildi. Hastanın post operatif iki yıllık takibinde rekürrens gözlenmedi.

Anahtar Sözcükler

Schwannom, baş ve boyun tümörleri

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Correspondence

Dr. Hanifi KURTARAN

Alparslan Türkeş Caddesi,

No:57 06510, Emek, Ankara Turkey

Tel: +90 312 203 51 09

Fax: +90 312 221 36 70

E-mail: hanifikurtaran@yahoo.com

INTRODUCTION

Schwannoma –neurinoma, neurolimmoma- was firstly described by Verocay in 1908.¹ It is a benign tumor originating from the Schwann cells that make up the myeline sheath of the peripheral nerves.^{2,3}

It is usually seen on the cranial nerves IX, X, XI, XII and the cervical sympathetic chain in the parapharyngeal space as a single solid mass. Multiple simultaneous tumors of the head and neck region have been seen very rarely,⁴⁻⁸ and, up to date, only one case of multiple schwannoma in the parapharyngeal space was reported in the literature.⁷ Herein, we present another case of multiple schwannoma with 13 synchronous masses.

CASE REPORT

A fifty-four year old male patient admitted to the otorhinolaryngology clinic with complaints of dysphagia, snoring, disturbance of phonation and gradual painless swelling in the neck and throat for one and a half years.

In physical examination, a gross submucosal mass was observed in the oropharynx. In the right side of the neck, close to the posterior edge of the parotid gland, a firm, painless and hardly mobile mass with slightly irregular borders was palpable. His vital signs and laboratory findings were all normal.

Cervical CT findings were as follows (Figure 1): There was a mass lesion with distinct borders localized at the inferomedial border of the right parotid gland descending inferiorly to the level of the thyroid cartilage through the right parapharyngeal space. The diameters of the mass were 3.5 x 8 cm.



Figure 1. Axial CT view of the mass. There is a mass lesion (arrow) with distinct borders localized at the inferomedial border of the right parotid gland descending inferiorly to the level of the thyroid cartilage through the right parapharyngeal space.



Figure 2. The specimen of mass.

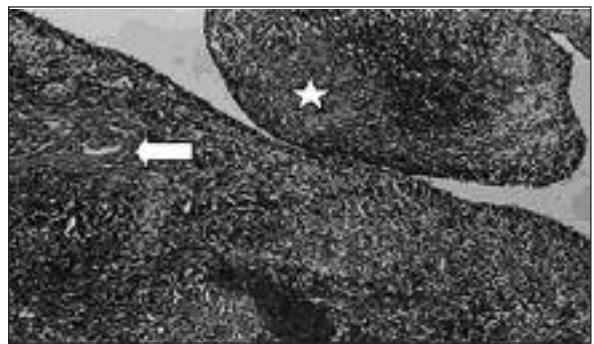


Figure 3. The microscopic view. The tumor reveals schwannoma consisting of Antoni A containing crowded spindle cells with palisading nuclei (arrow) and Antoni B containing loosely arranged spindle cells in abundant myxomatous matrix (star) areas. Tumor tissue was counterstained with hematoxylin and eosin ($\times 10$).

For therapeutic purposes, the mass was excised totally. The masses were located at the parapharyngeal space deep to the neurovascular sheath and parotid gland. Multiple masses were seen and they were separate from each other. The biggest one was 4x3x3 cm (Figure 2). All of the masses were originating from the cervical sympathetic plexus and had no relation with any of the cranial nerves. The pathological examination of the masses revealed the diagnosis of “multiple schwannoma” (Figure 3).

A slight Horner’s syndrome was present postoperatively, however no intervention required. No recurrences were seen in two years follow up period.

DISCUSSION

Schwannomas are benign, slowly growing, encapsulated peripheral nerve tumors composed of Schwann

cells and collagen fibers. They can occur in any region of the body where there is a nerve with a Schwann sheath.⁴ Most of the cases (25% to 45%) are seen in the head and neck region⁹ such as face, scalp, intracranial cavity, orbit, nasal and oral cavities, parapharyngeal space middle ear, mastoid, larynx, and medial and lateral regions of the neck.¹⁰

Schwannomas are often asymptomatic in the early stages. As the lesion grows, it compresses the nearby anatomical structures and causes pain, dysphagia, cough, voice changes and cranial nerve paralysis.⁹

Branchial cysts, lymph nodes, paraganglioma, tumors of the parotid gland, minor salivary gland neoplasms, lipomas, neurofibroma, teratoma, rhabdomyosarcoma and deep neck infection should be considered in the differential diagnosis of the schwannoma at the parapharyngeal space.

Magnetic resonance imaging (MRI) provides the best preoperative information regarding extension and

relationships to adjacent structures.¹¹ Differential diagnosis of parapharyngeal space masses can be made by showing the position changes of parapharyngeal fat column or internal carotid artery by MRI. Tumors compressing parapharyngeal fat column are usually extraparotid in origin. Tumors from retrostyloid location can cause an anteromedial displacement and tumors from prestyloid location can cause a posterolateral displacement of internal carotid artery.

Schwannoma is a generally single and well encapsulated benign tumor. The treatment is surgical excision. The most common method is cervical approach¹² as applied in this case. The prognosis is quite well for the totally excised tumors and recurrence rate is very rare.

We aim to alert physicians for the importance of early recognition and treatment of multiple schwannomas. In conclusion, multiple schwannoma must be included in the differential diagnosis in patients with neck swelling and multiple masses.

REFERENCES

- Verocay J. Multiple geschwulste als systemerkrankung am nervosen. Verocay J. Festschrift F. Chiari. 1st ed. Wien and Leipzig: W.Braunmiller; 1908. p.378-415.
- Weiss SW. Benign tumors of peripheral nerves. Enzinger FM, Weiss SW. Soft Tissue Tumors. 3rd ed. St Louis: Mosby-Year Book Inc; 1995. p.821-88.
- Huang JH, Simon SL, Nagpal S, Nelson PT, Zager EL. Management of patients with schwannomatosis: report of six cases and review of the literature. Surg Neurol 2004;62(4): 353-61.
- Shoss SM, Donovan DT, Alford BR. Tumors of the parapharyngeal space. Arch Otolaryngol 1985;111(11):753-7.
- Attie JN, Friedman E, Rothberg MS. Submandibular and axillary schwannomas not associated with von Recklinghausen's disease. J Oral Maxillofac Surg 1984;42(6):391-4.
- Michida A, Ryoike K, Ishikura S, Hamada T. Multiple schwannomas of the neck, mediastinum, and parapharyngeal space: report a case. J Oral Maxillofac Surg 1995;53(5):617-20.
- Santoro R, De Meester W, Coscarelli S, Polli G. Multiple neurinomas of the parapharyngeal space. Eur Arch Otorhinolaryngol 1997;254(6):301-3.
- Karpati R, Loevner L, Cuning D, Yousem D, Li S, Weber R. Synchronous schwannomas of the hypoglossal nerve and cervical sympathetic chain. AJR 1998;171(6):1504-7.
- Ganesan S, Harrar RP, Owen RA, Dawkins RS, Prior AJ. Horner's syndrome: A rare presentation of cervical sympathetic chain schwannoma. J Laryngol Otol 1997;111(5):493-5.
- Mikaelian DO, Holmes WF, Simonian SK. Parapharyngeal schwannomas. Otolaryngol Head Neck Surg 1981;89(1):77-81.
- Saito DM, Glastonbury CM, El-Sayed IH, Eisele W. Parapharyngeal space schwannomas: preoperative imaging determination of the nerve of origin. Arch Otolaryngol Head Neck Surg 2007;133(7):662-7.
- Stell PM, Mansfield AO, Stoney PJ. Surgical approaches to tumors of the parapharyngeal space. Am J Otolaryngol 1985;6(2):92-7.