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CASE REPORT
OLGU SUNUSU

Orta Kulakta Fasiyal Sinir Schwannomu

Facial nerve schwannoma located in the middle

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ÖZET

Schwannomlar periferik sinir schwann hücrelerinden kaynaklanan benign tümörlerdir. Baş ve boyun bölgesinde yaygın görülürler. Orta kulakta nadir görülmekle birlikte paragangliomalardan sonra ikinci en sık benign orta kulak tümörüdür. Orta kulakta en sık fasiyal sinirden ve korda timpani, stapedial sinir gibi dallarından kaynaklanır. Fasiyal sinir kaynaklı schwannomlar fasiyal sinir semptomlarından ziyade iletim tipi işitme kaybına neden olur. 3 ay önce gelişen periferik fasiyal paralizisi olan 59 yaşında erkek hastanın, yaklaşık 10 yıldır sol kulakta işitme kaybı yakınması mevcuttu. Yapılan radyolojik incelemelerde sol orta kulak boşluğunu tama yakın dolduran yumuşak doku dansitesinde, glomus tümörü, enflamatuar değişiklikler düşündüren patolojik görünüm mevcuttu. Hastaya timpanomastoidektomi yapıldı. Orta kulağı dolduran mavi morumsu renkte kitle izlendi. Kitlenin patolojik tanısı schwannom olarak rapor edildi. Bu sunuda nadir görülen bir orta kulak benign tümörü olan schwannom olgusu sunuldu.

Anahtar Kelimeler: Schwannom; fasiyal siniri; orta kulak

ABSTRACT

Schwannomas are benign tumors originating from peripheral nerve, schwann cells, and located in head and neck region commonly. They are the second most common middle ear tumors after paragangliomas, and they may often arise from the facial nerve and also from its branches such as the chorda tympani and the stapedial nerve. Facial nerve schwannomas cause conductive hearing loss rather than usual symptoms of facial nerve. Our patient was 59-year -old man, who had hearing loss on his left ear for 10 years and who had facial paralysis for 3 months before coming to our clinic. Temporal bone computerized tomography scan showed soft tissue mass involving the timpanic cavity completely, which suggest glomus tumor or inflamatory changes. Tympanomastoidectomy operation was performed. The histopathological diagnosis of the mass was reported as schwannoma. In this case, we presented the schwannoma as a rare benign tumor of the middle ear.

Keywords: Schwannoma, facial nerve, middle ear

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INTRODUCTION

Paragangliomas are the most common type of middle ear and mastoid tumors. Facial nerve schwannomas (FNS), cholesterol granulomas, primary cholesteatoma, choristoma, hemangioma, menengioma are all the other of middle ear tumors (1,3). Shwannomas, in head and neck region, can be originated from IX, X, XI. cranial nerves (4). It originates from facial nerve and its branches such as chorda tympani, stapedial nerve, major petrosal nerve or Jacobson nerve (tympanic branches of N. glossofarengeus) or Arnold nerve (auricular branches of N. Vagus). The most common neurogenic tumor of middle ear is facial nerve schwannoma (5,6,7,8). In this report, a case with schwannoma originating from facial nerve's second angle area was presented.

CASE

A 59-year-old man was admitted to our hospital for House-Brackmann(H-B) grade 3 left-sided peripheral fascial paralysis, and in his history, he had a-day-pain in the left ear and otorrhagia before facial nerve paralysing. In his autoscopic examination, left tympanic membrane was intact, non-transparent and opaque. Patient has been suffering from hearing loss approximately for 10 years, but he has never experienced discharching from both ears. In the magnetic resonance (MR) imaging of temporal region shows that tumor with an contrasted area sized 17x8x19 mm, occupied from internal jugular vein's superolateral to petrous apex. There was changes in contrasted area to be inflamation or glomus tumor behind the membrane (see figure 1).

Loss of ventilation in the left middle ear and mastoid, a soft-tissue density in the mastoid antrum and an erosion in incus were seen in temporal computerised tomography (CT). Systemic steroid treatment was began to the patient for facial paralysis. In post-treatment evaluation, facial paralysis was established as H-B grade 2. Facial EMG (Electromyography) shows the middle-severe grade partial axonal degeneration findings in the facial nerve. In the pre-operative assesment of the patient, benign middle ear tumor was suspected. Our surgical procedure was middle ear exploration and intact canal wall mastoidectomy. Tumor was grossly totally removed in the surgery. During operation it was observed that the middle ear and antrum were full of polypoid soft tissues. Dehiscence was at the second genu of facial nerve canal and it was established that the tumoral tissue was originating from the second genu. Pedunculated mass was filling the middle ear and laying to the eustachian tube as 5-7 mm.

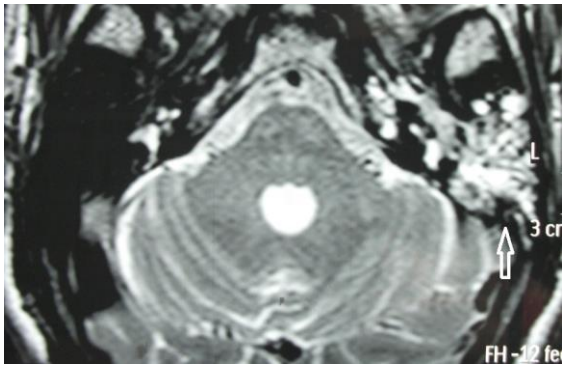

Figure 1: Axial T2-weighted MR image shows hyperintense mass. There is either glomus tumor or inflammatory conditions in left middle ear.

Tumoral tissue was removed from middle ear and antrum. Incus long arm and manibrium mallei was eroded. Head of stapes and its cruras was not seen. Residues of incus and malleus were removed. Graft, obtained from temporal muscles' fascia, was used in underlay procedure and hearing reconstruction was postponed to the second operation. Post-operative pathological diagnosis was schwannoma (see figure 2a-2b).

Facial EMG showed findings that were acute period severe axonal degeneration at the 15th day of post-operative. There was no change in the facial functions of patient and no recurrence during the 7 months of following period.

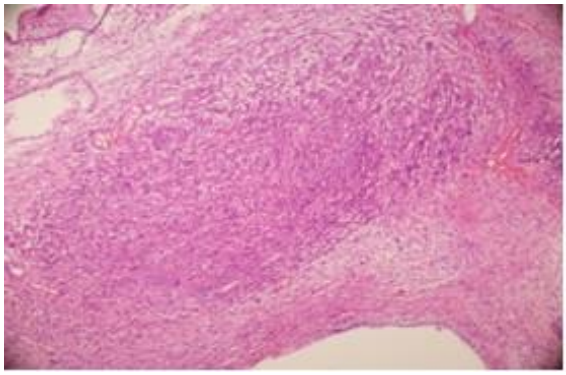


Figure 2a: A lesion, composed of compact spindle cells which was created a short bundles (HEX200). **Figure 2b:** Spindle cells, stained positive with S-100 (S-100X200).

DISCUSSION

Facial nerve schwannomas (FNS) are rare benign tumors of temporal bone (9). Low developing facial paralysis or paresis and hearing loss can be seen in FNS as well as tinnitus, otalgia, vestibular symptoms or mass in the external ear canal. Facial tics and paresis are findings that support the primary facial nerve tumor (10). In our case, there were otalgia, left facial paralysis followed to the otoragia and moderate mixt type hearing loss. FNS may be confused with glomus jugulare. However, schwanomas can be identified with discrete boundaries as they are less vascular than glomus tumors (11). As clinically facial twitching and progressive paresis strongly suggests a primary facial nerve tumor (12).

Surgical excision and nerve grafting seem to be a choice of conventional primary treatment of the FNS, which is an enlarging tumor with facial function H-B ≥ 4 (12). The surgical intervention can produce facial paralysis as H-B grade 3. However, nerve functions be reversible for long period of time. Some authors have applied the partial resection with “peeling” or “stripping” techniques to the tumor (13) and reported that they have gained successful results as H-B grade 1 to 2; however, this procedure has not been suggested for treatment of FNS because it has much more potential for leading to worse results than pre-operative period (11). Another applied method to treat the FNS is stereotactic irradiation. A study, managed by Kida et al in 14 patients, has been reported that 13 of them were either improved or unchanged in terms of facial functions and all tumor’s sizes were either remained constant or decreased (10).

There is a few studies on the FNS and their long-term results are still limited. In this case, middle ear and **transmastoid approach** was applied to the tumor resection. We did not decompress to the facial canal, and we procrastinated the reconstruction of hearing to the second session. There was no significant change in the post-operative facial nerve function.

In conclusion, there should be carried out differential diagnosis in order to distinguish middle ear schwannomas from the other middle ear benign tumors. The extension and the characteristics of tumor can be detected with CT and MRI. Erosion of the anatomic structures shown by CT may suggest the correct origin of the tumor, which was detected an erosion in incus in our case. Tumor may involve any division of the facial nerve, but have a preference for the perigeniculate area, the tympanic segment, and the mastoid segment (9).

The important goal of the treatment of FNS is to decide observation or surgical resection of the tumor.

The treatment option depend on tumor size, facial nerve function, and area of tumor involvement (12). There are many options for the treatment of FNS; furthermore the basic principle of treatment is both to maintain the functions of facial nerve at the best level and to realize optimize function of facial nerve as

soon as possible.Ultimately, surgical resection remains its status as gold standard.

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