

Giant facial schwannoma with intracranial extension: a case report

Gokhan Canaz¹, Izzet Durmusalioglu¹, Mustafa Ali Akcetin², Nur Topyalin³, Cemile Ozdemir⁴, Ali Osman Akdemir²

¹Bakirkoy Research and Training Hospital for Neurology, Neurosurgery and Psychiatry, Department of Neurosurgery, Istanbul, TURKEY

²Haseki Training and Research Hospital, Department of Neurosurgery, Istanbul, TURKEY

³Van Yuzuncu Yil University, Department of Neurosurgery, Van, TURKEY

⁴Haseki Training and Research Hospital, Department of Pathology, Istanbul, TURKEY

Abstract: *Introduction:* Facial nerve schwannoma is a rare benign tumor which may originate from any segment of the facial nerve. We present a case of a giant cystic facial nerve schwannoma that showed extension to the middle cerebral fossa, together with its clinical and radiological characteristics, and a short review of the related literature.

Case Report: A 52-year old female patient attended with complaints of numbness in the right half of her face, and mild hearing loss. Examinations revealed House-Brackmann Grade 3 peripheral facial paralysis, sensorineural hearing loss of 30% in her right ear. She described three episodes of facial paralysis in the last six years. Cranial MRI and CT scan revealed a round cystic lesion of about 40x44x38 mm, located centrally and medially in the right temporal area. The surgical operation was performed and the lesion was totally extracted. The histopathological investigation reported the lesion as a schwannoma.

Conclusion: Facial nerve schwannoma is a rare, benign tumor that occurs in different localizations. It may commonly be mistaken in prediagnosis and may be confused with other clinical states due to its clinical characteristics. It has to be kept in mind in the prediagnosis of patients with facial paresis and hearing loss.

Key words: Schwannoma, Facial Palsy, Tinnitus, Hearing Impairment

Introduction

Facial nerve schwannoma (FNS) is a benign tumor that is rarely seen, and is generally difficult to diagnose before surgery. It may originate from any segment of the facial

nerve through its extension from its origin in the cerebellopontine angle, to the parotid gland. Schwannomas account for 8% of all intracranial tumors. Of these, FNSs account for about 1.9%. FNSs that extend to the middle cerebral fossa are rare, as they commonly

originate from the geniculate fossa (9, 10). Here, we present a case of a giant cystic facial nerve schwannoma that showed extension to the middle cerebral fossa, together with its clinical and radiological characteristics, and a review of the related literature.

Case

A 52-year old female patient attended with complaints of numbness in the right half of her face, and mild hearing loss. Her physical examination revealed House-Brackmann Grade 3 peripheral facial paralysis, and hearing loss. The audiometric evaluation revealed a sensorineural hearing loss of 30% in her right ear. In her anamnesis, the patient described three episodes of facial paralysis: the first one occurred six years previously, the second four years, and the most recent one 15 days previously; a radiological investigation had not been performed. Cranial contrast enhanced magnetic resonance imaging (MRI), revealed a round cystic lesion of about 40x44x38 mm with regular margin, and with a peripheral capsule of about 8 mm maximal thickness, located centrally and medially in the right temporal area (Figure 1). In the computerized tomography (CT), we observed erosion of the petrous bone and widening in the facial hiatus on the right side (Figure 2). The patient was operated with a pre-diagnosis of meningioma, metastasis or schwannoma. The tumor was accessed using a right pterional craniotomy and sylvian dissection. The tumor was grey-yellow colored and it has hemorrhages in some areas. The tumor was easily dissected from the surrounding tissue. A tissue specimen was transferred for frozen

section investigation. The pathological examination excluded malignancy. The mass was totally extracted following internal decompression. The optic nerve, carotid cisterns, and anterior communicating artery and its segments were intact. It was established that the petrous bone and dura were eroded in the area where the mass was adjacent to the temporal bone; therefore the dura was repaired in this region using a galeal graft, and the operation was completed. The histopathological investigation reported a schwannoma from the palisading pattern with S-100 protein positive, glial fibrillary acidic protein (GFAP) and epithelial membrane antigen (EMA) negative (Figures 3A,B).

A post-operative physical examination of the patient revealed progression of right facial paralysis to House Brackmann Grade 4 level; her hearing loss was also determined to progress. An investigation of the facial nerve electromyography (EMG) revealed severe axonal damage in the facial nerve. A control EMG was recommended one month later. Almost complete hearing loss was determined on the right side in her audiometric evaluation. In the post-operative contrast enhanced cranial MRI, it was determined that the mass has been totally excised (Figure 4). The patient was discharged from hospital on the post-operative 14th day. The second control EMG was administered about two months later, and the result was compatible with a lesion causing complete axonal damage to the right facial nerve segments that branches off from the orbicularis oculi and frontal muscles, and severe axonal damage in the segment that branches off from the orbicularis oris muscle.

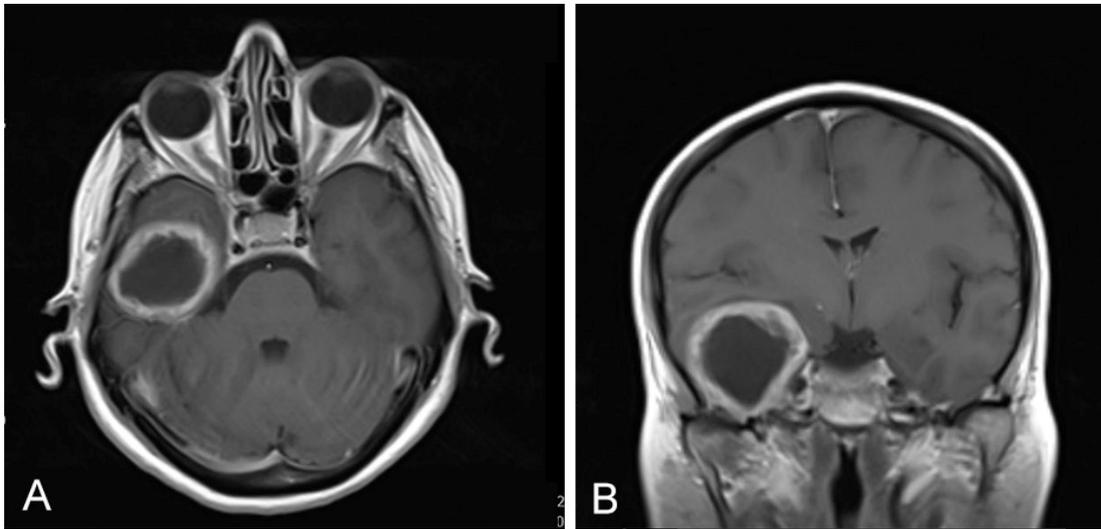


Figure 1 - Contrast enhanced cranial MRI: A round cystic lesion of 40x44x38 mm in size with a regular margin, and with a peripheral capsule of 8 mm in maximal thickness, located centrally and medially in the right temporal lobe. A: Axial section. B: Coronal section

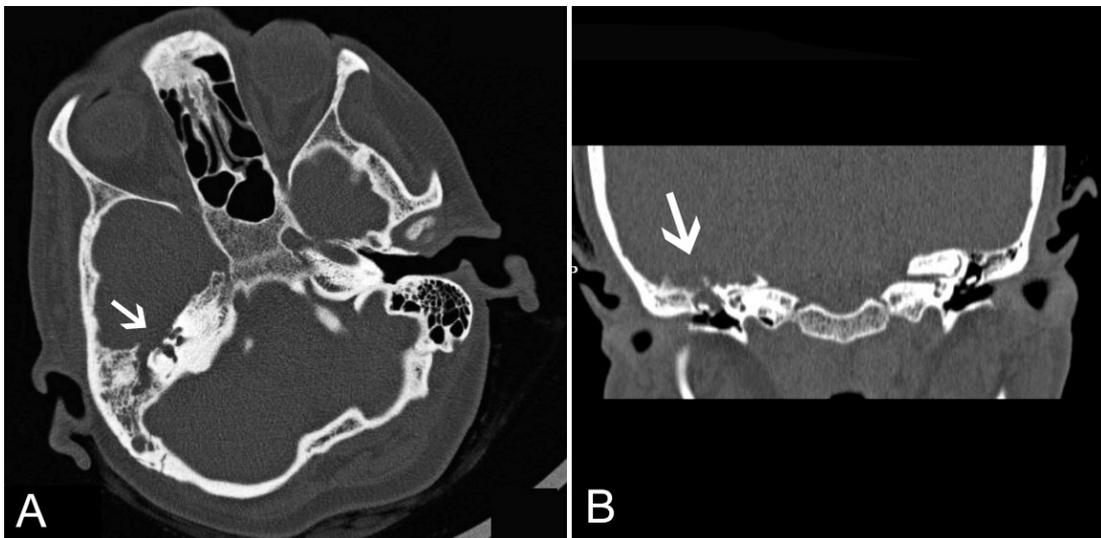


Figure 2 - A: Widening and erosion in the facial hiatus in axial section, in the temporal CT. B: Erosion in the right temporal bone petrous segment, in the coronal section

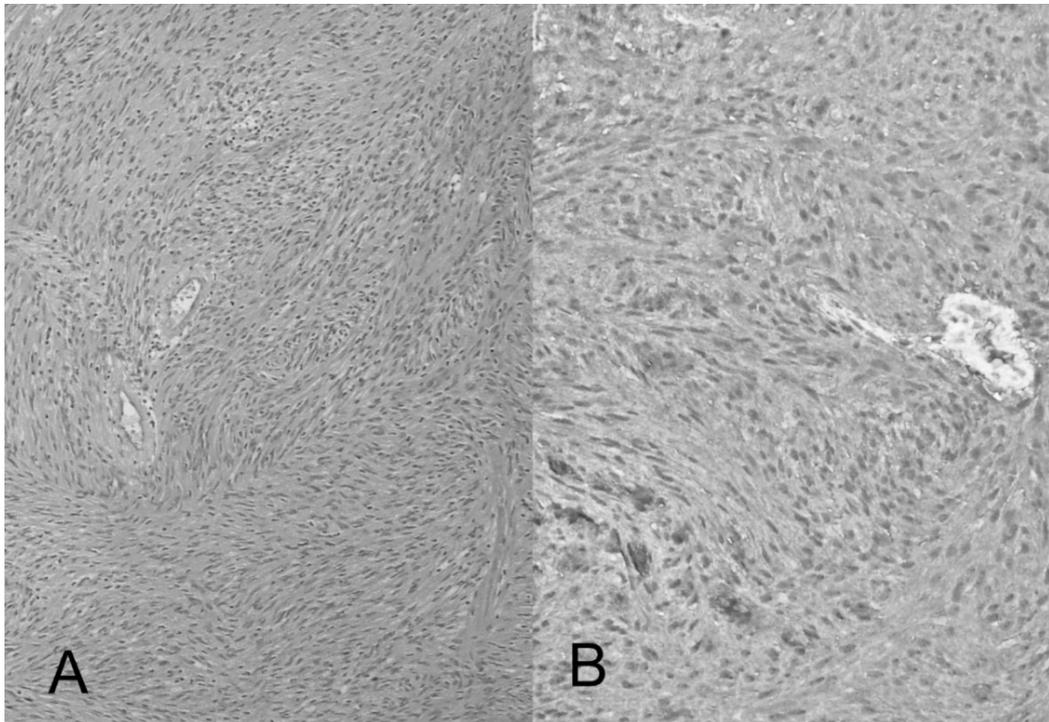


Figure 3 - A: Bundles of fusiform cells by hematoxylin staining, magnified 100 times. B: Diffuse positive reaction by S-100 immunohistochemical staining

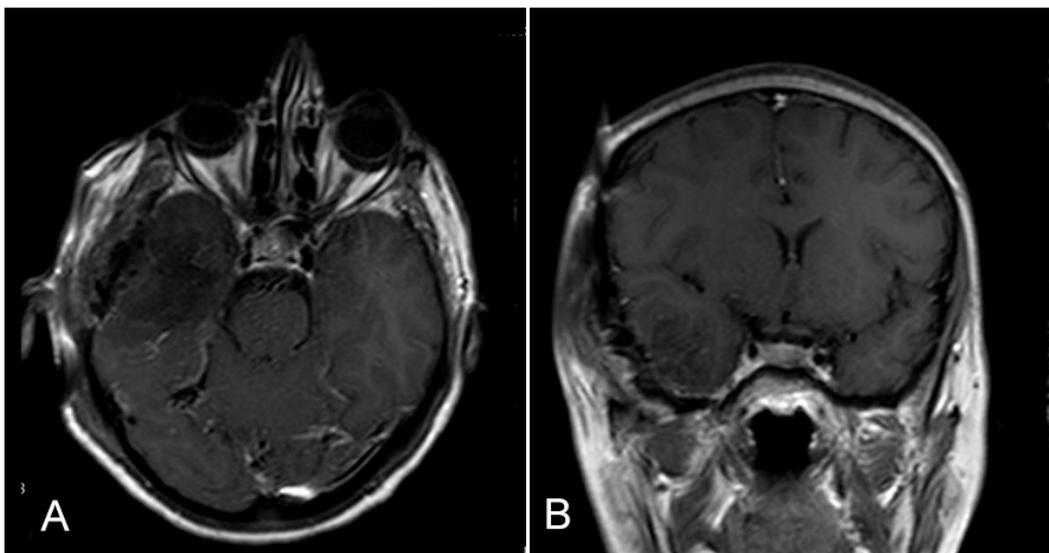


Figure 4 - Postoperative changes in the postoperative contrast enhanced cranial MRI

Discussion

Schwannomas are simply benign tumors that originate from the Schwann sheaths of the peripheral neural cells (cranial and spinal). Giant size described as > 4.0 cm in maximal extrameatal diameter by Samii et al. (7). They arise from sensory nerves, and are also known as neuromas. Schwannomas that originate from cranial nerves account for 8% of intracranial tumors, and they occur at a relatively high frequency. FNS accounts for 1.9% of all intracranial tumors. A FNS generally extends intracranially to the cerebellopontine angle, and to a lesser extent, to the middle cerebral fossa (11). Its rare occurrence in middle cerebral fossa, and the presence of more frequently seen lesions in this area that have similar appearances radiologically, make the differential diagnosis and accurate pre-diagnosis difficult.

FNS occurs clinically as a facial nerve paresis or paralysis with a long-lasting, fluctuating and progressive course. It has to be kept in mind that hearing loss (sensorineural or conductive type) occurs as frequently as facial paralysis. It has found to occur even more frequently in some studies, including large case series (4). This clinical feature often leads to false pre-diagnosis of vestibular schwannoma. Symptoms are often found to be associated within the region of the facial nerve in which the lesion is located. The symptoms are generally characterized with various distributions of the facial, otologic, and mass effect signs; however this is not a clinically pathognomonic sign for the diagnosis of FNS. A high correlation has been detected between

the degree of facial nerve dysfunction and intratemporal location; otologic complaints like sensorineural hearing loss, tinnitus, and vertigo have been reported most frequently in patients with intracranial tumors (2). Because the differential diagnosis of acoustic neuroma and FNS is difficult, diagnosis of FNS is almost always ascertained perioperatively or postoperatively.

There are different studies and classifications related to the localization and frequency of FNSs on the facial nerve. In many studies, it has been reported that FNS most frequently occurs in the geniculate ganglion along the course of the facial nerve (4). In a study comprising 24 patients with a histopathological diagnosis of FNS, the facial nerve has been investigated in eight separated segments, as follows: cerebellopontine angle, internal acoustic canal, labyrinth, geniculate fossa, nervus petrosus major, tympanic, mastoid, and intracranial. In this study, the geniculate fossa was established to be the most frequent location (83%); of these tumors located in the geniculate fossa, 60% occurred with extension to the labyrinth, and 30% were with petrosal nerve involvement. Only two cases were determined with intracranial extension (8.3%), and both of these originated from the geniculate fossa. Facial neuropathy was identified in these two cases with intracranial extension, and one case with sensorineural hearing loss (11). In a study, the location of FNS was classified as intratemporal, extratemporal and intracranial, and the most frequent location was in the intratemporal region (61%); tumors with extratemporal (parotid area) and intracranial

(cerebellopontine angle) locations, were reported in equal frequencies. In this study, extension to the middle cerebral fossa was not included in the intracranial location, and one tumor was reported with intracranial extension occurred in the intratemporal location (2).

High-resolution enhanced CT and MRI, are the most important visualization methods in the radiological diagnosis of FNS. CT may not be considered absolutely essential; however it is important in the visualization of the changes in the temporal bone, to determine the origin of the tumor, and when planning surgery. Erosion of the petrous bone, internal acoustic canal and a widened facial hiatus can easily be visualized using CT, which also helps in the preoperative differential diagnosis (8). A contrast enhanced MRI is the preferential diagnosis for FNS, independent of the size and location of the tumor. These tumors classically occur in MR imaging as hypo-hyperintense in the T1 weighted sequences, hyperintense in the T2 weighted sequences, and with contrast enhancement (3). Large tumors commonly show cystic differentiation, and peripheral contrast enhancement.

FNSs with extension to the middle cerebral fossa, are benign tumors, and they frequently cause mass effect; the first choice of treatment is therefore surgery. Different studies make different recommendations regarding the surgical indication and approach; however the most common suggestion for tumors beyond House-Brackmann stage 3, is surgery (4). FNSs are tumors that grow very slowly (0.02 cm³ /year); however surgery must not be

delayed when there is involvement of the middle fossa (6). Taking into consideration the sense of hearing, and location of the tumor in the facial nerve segment, suboccipital, infratemporal, frontotemporal, transpetrosal, and retrosigmoid approaches may be undertaken by neurosurgeons, with the participation of nose-ear-throat specialists, if needed. FNSs are tumors of the nerve sheath, and it is theoretically possible to extract them without damaging the nerve; however following middle fossa surgery, this segment of the facial nerve is almost always extracted together with the tumor. This results in permanent facial paralysis. There are studies in the literature using different methods of reconstruction; however nerve function better than that of House-Brackmann stage 3 cannot be maintained (2, 5). The general methods applied include the interposition of the nervus auricularis major, and hypoglossal nerve anastomosis by grafts from the sural nerve or vestibular nerve. Additional techniques including sutures and fibrin tissue adhesives without sutures are also used. Facial nerve conduction can be maintained in some of these studies; however clinical improvement does not occur, due to atrophy developing in the muscles that it innervates. In cases with unimpaired hearing occurring with tumors extending to the middle cerebral fossa, transmastoid- middle fossa combined surgery is recommended. Hearing may be maintained to some extent following surgery, in small or moderate tumors (3-15 mm); in large tumors (>18 mm), but it has also been reported that it's not possible to protect the sense of hearing (1).

Conclusion

Facial nerve schwannoma is a rare, benign tumor that occurs in different localizations. It may commonly be mistaken in prediagnosis, and may be confused with other clinical states due to its clinical characteristics. It is possible to protect the sense of hearing, and the facial nerve functions in cases diagnosed at an early stage; therefore it has to be kept in mind in the differential diagnosis of patients with facial paresis and hearing loss.

Correspondence

Gokhan Canaz, Bakirkoy Research and Training Hospital for Neurology, Neurosurgery and Psychiatry, Department of Neurosurgery, Bakirkoy / Istanbul, Turkey.

Phone: +90 212 409 15 15

Mail:gokhancanaz@gmail.com

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