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CASE REPORT

An intratemporal facial nerve neuroma: a case report

İntratemporal fasyal sinir nöromu: Olgu sunumu

İbrahim KETENCİ, M.D.,[1] Yaşar ÜNLÜ, M.D.,[1] Abdulhakim COŞKUN, M.D.,[2] Erkun TUNCER, M.D.[1]

A forty-one-year-old man presented with right-sided progressive facial nerve paralysis of one year duration. Computed tomography of the temporal bone showed a soft tissue mass in relation with the tympanic segment of the facial nerve with destructive changes in the facial recess region. Magnetic resonance images before and after gadolinium injection revealed a contrast-enhancing mass lesion in the tympanic cavity. The tumor was removed through a transmastoid approach and a cable graft from the greater auricular nerve was used to repair the defect. Histopathologic diagnosis was facial nerve schwannoma. At the end of one-year follow-up, the patient had moderately severe facial nerve dysfunction.

Key Words: Facial nerve/radiography; facial nerve diseases/diagnosis/surgery; facial paralysis/etiology/radiography; magnetic resonance imaging; neurilemmoma/diagnosis; neuroma/diagnosis/surgery/radiography; tomography, X-ray computed.

Kırk bir yaşındaki erkek hasta, sol tarafı tutan, bir yıldır yavaş gelişim gösteren fasyal paralizi şikayeti ile başvurdu. Temporal kemik bilgisayarlı tomografisinde fasyal sinirin ikinci dirseği hizasında, orta kulağa doğru uzanan ve fasyal reses bölgesinde tahrip edici değişiklere yol açan yumuşak doku kitlesi görüldü. Gadolinum ile yapılan menyetik rezonans görüntülemede orta kulak boşluğunda kontrast madde tutan kitle saptandı. Transmastoid yolla girilerek, fasyal sinirdeki kitle cerrahi sınırlarda rezeke edildi ve n. aurikularis magnustan alınan sinir grefti ile tamir edildi. Histopatolojik incelemede fasyal sinir schwannomu tanısı kondu. Ameliyattan bir yıl sonra yapılan incelemede fasyal sinir fonksiyonunda kısmi düzelme görüldü.

Anahtar Sözcükler: Fasyal sinir/radyografi; fasyal sinir hastalıkları/tanı/cerrahi; fasyal paralizi/etyoloji/radyografi; manyetik rezonans görüntüleme; nörilemoma/tanı; nöroma/tanı/cerrahi/radyografi; bilgisayarlı tomografi.

Facial nerve neuromas are uncommon, slow-growing neoplasms that may occur anywhere along the course of the facial nerve from the brain stem to the facial muscles, leading to a wide variety of symptoms, depending on the segment of the nerve involved.[1-3]

The use of computed tomography (CT) and later magnetic resonance imaging (MRI) allowed examination of the temporal bone in great detail, resulting in earlier and more accurate diagnosis.[3-8]

CASE REPORT

A forty-one-year-old man presented with right-sided progressive facial nerve paralysis of one year duration. On physical examination, the tympanic membrane was normal. Schirmer's test was normal. The right acoustic reflex was absent. Audiometric evaluation showed normal hearing on both sides, with 100% discrimination. The nerve was unresponsive to electrical stimulation.

◆ *Departments of [1]Otolaryngology, and [2]Radiology, Medicine Faculty of Erciyes University, Kayseri, Turkey.*

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◆ *Correspondence: Dr. İbrahim Ketenci. Alpaslan Mah., Bahar Sok., Şişli Sitesi, A Blok, No: 20/26, 38040 Kayseri, Turkey. Tel: +90 352 - 437 49 01 / 21432 Fax: +90 352 - 437 91 64 e-mail: ketenci@erciyes.edu.tr*

◆ *Erciyes Üniversitesi Tıp Fakültesi, [1]KBB Hastalıkları Anabilim Dalı, [2]Radyoloji Anabilim Dalı, Kayseri.*

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◆ *İletişim adresi: Dr. İbrahim Ketenci. Alpaslan Mah., Bahar Sok., Şişli Sitesi, A Blok, No: 20/26, 38040 Kayseri. Tel: 0352 - 437 49 01 / 21432 Faks: 0352 - 437 91 64 e-posta: ketenci@erciyes.edu.tr*

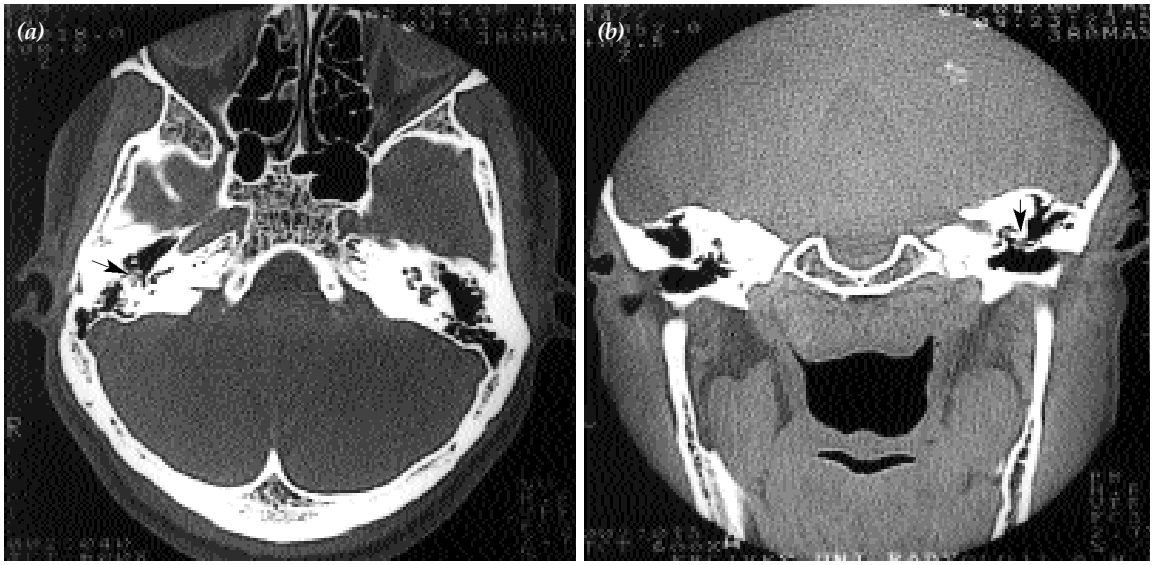

Fig. 1 – *High-resolution* ***(a)*** *axial and* ***(b)*** *coronal computed tomography images showing a mass lesion filling the facial recess and extending into the tympanic cavity.*

A high-resolution CT scan of the temporal bone showed a mass lesion in close relationship with the tympanic portion of the facial nerve, with destructive changes around the facial recess (Fig. 1). Magnetic resonance images revealed a mass lesion in the tympanic cavity, arising from the tympanic portion of the facial nerve. It showed intermediate signal intensity on T_1- and high-signal intensity on T_2-weighted images (Fig. 2). Both CT and MRI findings were suggestive of a facial nerve neuroma.

Exploratory tympanotomy showed a tumoral mass originating from the second genu of the facial nerve and filling the facial recess. The tumor was resected with the facial nerve through a transmastoid approach. The margins were clear and a graft using the greater auricular nerve was placed in the

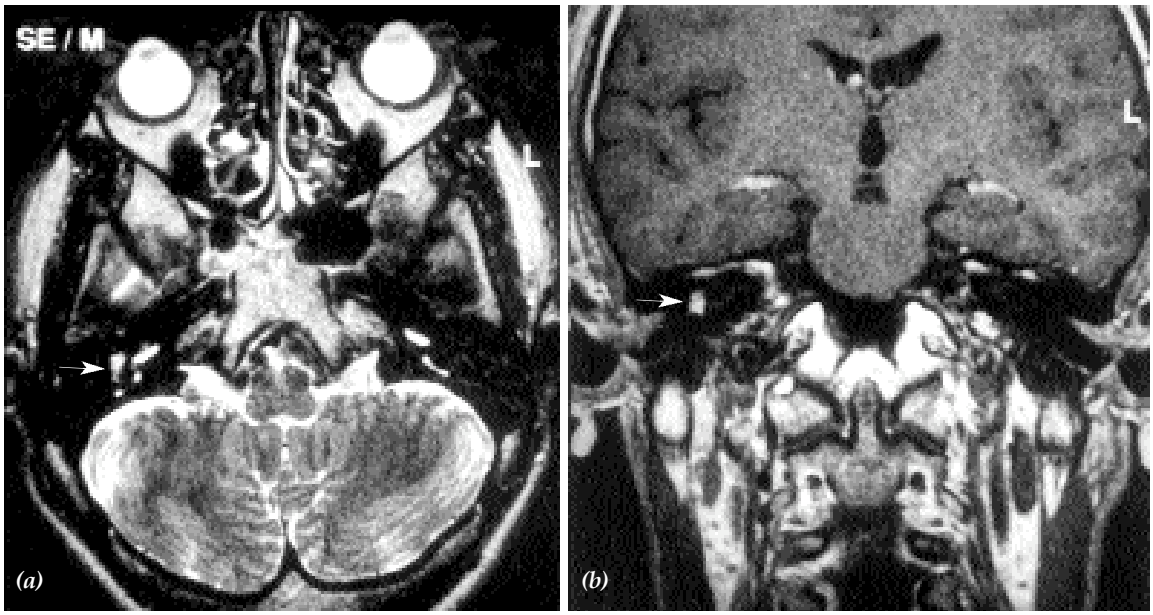

Fig. 2 – ***(a)*** *Axial T_2-weighted magnetic resonance image showing a soft tissue mass with high-signal intensity in the tympanic cavity.* ***(b)*** *Coronal T_1-weighted view after intravenous gadolinium injection showing a contrast-enhancing lesion.*

Fallopian canal. The histological diagnosis was facial nerve neuroma.

At the end of one-year follow-up, House-Brackmann grading scale showed grade IV recovery of the facial nerve function.[9] Audiometric evaluation showed normal hearing on the left, but a conductive hearing loss of 37 dB was detected on the right, with 100% discrimination.

DISCUSSION

Facial nerve neuromas are known to arise from the neuroectodermal sheath of peripheral nerves. Of the cranial nerves, the vestibular portion of the eighth nerve is most frequently involved. Neuromas arising from the facial nerve occur less frequently and may involve any segment of the facial nerve. The increasing number of cases reported in recent years suggests that recognition of facial neuromas is improving. This may also be due to the presence of increased number of fellowship-trained neuro-otologists and neuro-radiologists and to a greater cooperation between surgical subspecialties.[8]

The clinical presentation of facial nerve neuromas varies depending on the portion of the nerve involved. The intratemporal portion of the facial nerve is most commonly involved, causing disturbances in facial nerve function as the most common presenting complaints.[3,6,10,11] The classical history is a slowly-progressing facial paralysis over months to years, although sudden onset of paralysis, fluctuating paresis, and facial tics may also occur.[11] In our patient, a slow but progressive facial paralysis was the only symptom. Intracranial cases typically present with sensorineural hearing loss or tinnitus.[8] O'Donoghue et al.[12] reported that hearing loss and tinnitus were the most common symptoms in their review of 48 patients with facial nerve neuromas.

Recognition of facial nerve neuromas requires proper diagnostic studies. All patients should have a complete neurological evaluation including topographic testing of the seventh nerve and nerve excitability tests. However, the latter tests may be confounded by the fact that these tumors still transmit electrical activity because of their slow growth and the presence of partial regeneration of nerve fibers. Topographic testing of facial nerve function, including tearing, stapedius reflex, taste, and salivation may be helpful, but may not be definitive in the evaluation of the extent of facial nerve neuroma.[3,11] In our case, the Schirmer's test was normal and no acoustic reflex was detected on the affected side.

Although improvements in imaging techniques of the temporal bone have increased the possibility of a correct preoperative diagnosis, tumoral infiltration into normal-looking adjacent nerve segments may make radiological tests unreliable.[3,11] Computed tomography shows segmental involvement of the facial nerve. High-resolution CT allows identification of a soft tissue mass along the course of the facial nerve, often with enlargement of its bony canal.[7] Pressure-induced erosion to the underlying bone may be noted and erosion of the ossicles may be demonstrated in middle ear involvement.[4,7] Facial neuromas may also be associated with internal auditory canal widening, cerebellopontine angle extension, or a middle cranial fossa mass, mimicking vestibular neuromas.[3-5,12]

Magnetic resonance imaging is superior to CT in delineating a soft tissue mass along the facial nerve, especially with contrast material. The arteriovenous plexus along the Fallopian portion of the facial nerve may be visualized in a spotty fashion by contrast enhancement.[11] However, as inflammatory lesions of the facial nerve will also result in contrast enhancement,[11,13] enhanced MRI findings should be considered indicative of tumor only if there is a soft tissue mass.[3,11,13] Hence, clinical findings, CT or MRI should be correlated before diagnosing a facial nerve tumor. In this case, both CT and MRI findings of the temporal bone were consistent with facial nerve neuroma.

An intratemporal facial nerve neuroma generally extends along the course of the facial nerve to a greater distance than clinically or radiologically anticipated.[3,11] Neuromas detected in the middle ear may extend intracranially into the internal auditory canal and cerebellopontine angle or extratemporally into the parotid gland.[14] The major limitation of preoperative imaging is to differentiate from facial acoustic neuromas when the labyrinthine segment or more distal portions of the facial nerve are not involved. The finding of a small tumor with eccentric involvement of the long axis of the internal auditory canal suggests the preoperative diagnosis of a facial neuroma.[15]

The management of facial neuromas should be directed to obtain maximal facial nerve function, preservation of hearing, and prevention of recurrence and complications.[3,8,11] The entire intratemporal course of the nerve should be explored. An

asymptomatic tumor or one associated with moderate facial dysfunction requires particular consideration with respect to timing and type of surgery. Some authors recommend a "wait-and-see" policy for small tumors, with minimal impairment in facial nerve function,[4,16,17] while others advocate surgery as soon as the diagnosis is made.[3] The patient's age and willingness to comply with follow-up imaging must be considered before choosing observation.[8]

On the other hand, the type of surgery must be tailored to the individual case. The nerve can be exposed in its intracanalicular and labyrinthine course either by a middle fossa or a translabyrinthine approach. The latter route is employed in the presence of a non-serviceable hearing loss or a dead ear.[3,11] The tympanic and mastoid segments may be explored through a transmastoid approach. A transmastoid and extralabyrinthine approach when the tumor involves the geniculate ganglion and the lateral portion of the intralabyrinthine segment of the facial nerve.[18] Removal of the tumor generally requires resection of a length of the normal-looking nerve and the use of frozen section is recommended to ensure complete tumor removal.[3,4,11] Cable grafting is the preferred repair technique with the use of a portion of the greater auricular nerve or the sural nerve.[3] If primary anastomosis or cable grafting fails, hypoglossal-facial anastomosis may be necessary.[11] In our case, a transmastoid approach was used for tumor removal and facial nerve repair was made with the use of the greater auricular nerve.

Surgical exploration has been recommended in patients in whom radiological evidence for tumor is lacking despite the persistence of facial palsies progressive in nature or facial palsies that show no return in tone or function after 6 to 12 months.[11]

The recovery of facial nerve function depends largely on the duration of preoperative facial paralysis, the results being invariably poor when it is of long duration.[12,19] In our patient, the duration of facial paralysis was about one year and the recovery of facial function one year after surgery was limited.

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