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Intracanalicular Contrast Enhancement on Magnetic Resonance Imaging in Ramsey Hunt Syndrome Mimicking Acoustic Neuroma: Case Report

Manyetik Rezonans Görüntülemede İntrakanaliküler Kontrast Tutulumu ile Akustik Nörinomu Taklit Eden Ramsey Hunt Sendromu: Olgu Sunumu

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ABSTRACT

Magnetic resonance imaging (MRI) is a gold standard method for the diagnosis of lesions that are located in the internal acoustic canal. A 60- year-old man with vesicular lesions on his left auricle, erupted after the facial paresis was presented. He also had vertigo and hearing loss and his complaints did not recover after one month. MRI was obtained and an abnormal contrast enhancement was found at the intracanalicular area, similar to the view of an acoustic neuroma. During follow-up, this image of contrast enhancement was still evident in the MRI with minimal reduction. Abnormal contrast enhancement of the labyrinthine segment and other cranial nerves localized on the intratemporal area could be seen frequently in the Ramsey Hunt Syndrome. However, a contrast enhancement confined to the intracanalicular area and persistence of findings more than eight weeks is not frequent. Ramsey Hunt Syndrome may mimic acoustic neuroma with its late clinical and MRI findings. At the differential diagnosis, repeated clinical and radiological evaluation of the patient is crucial.

Keywords
Herpes zoster oticus; neuroma, acoustic; magnetic resonance imaging

ÖZET

Magnetik rezonans görüntüleme (MRG) internal akustik kanal yerleşimli lezyonlarda altın standarddır. Altmış yaşında erkek hasta fasial parezi ile başvurmuş, izleminde aurikulada veziküler lezyonlar çıkması üzerine tedavi altına alınmıştır. Bir ay sonra halen fasial parezinin devam etmesi ve beraberinde vertigo ve aynı tarafta işitme kaybı olması üzerine çekilen MRG görüntülerinde intrakanaliküler anormal kontrast tutulumu tespit edilmiş; bu görünüm akustik nörinomdan ayırt edilememiştir. İkinci ayda yapılan kontrol MRG' da aynı kontrast tutulum devam etmiş ancak minimal gerileme tespit edilmiştir. Hasta uzun süreli takiplerinde fasial fonksiyonlarını yeniden kazanmıştır. Ramsey Hunt Sendromunda intratemporal bölgede labirint ve diğer kranyal sinirlerin anormal kontrast tutulumu sık görülen bir bulgudur. Ancak sadece kanal içinde görülen ve sekiz haftadan daha uzun süren anormal kontrast tutulum sık görülen bir durum değildir. Ramsey Hunt Sendromu, geç dönem klinik ve MRG bulguları ile akustik nörinomu taklit edebilir. Hastanın tanısının serolojik olarak teyit edilmesi önemli olmakla beraber tanı koyarken en önemli nokta tekrarlanan çekimlerde ve takiplerde yapılan değerlendirmedir.

Anahtar Sözcükler
Herpes zoster otikus; nöroma, akustik; manyetik rezonans görüntüleme

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INTRODUCTION

Ramsey Hunt Syndrome is a neurological disease composed of two subgroups.[1] Ramsey Hunt Syndrome type 1 is named as "Herpes zoster oticus" and found in 1907 as a result of recurrent infection with reactivation of Varicella Zoster virus which had already present in the geniculate ganglion.[1,2] In this syndrome, acute ipsilateral facial paralysis, otalgia, hearing loss, vertigo, tinnitus, and multiple cutaneous herpetic lesions may be seen.[1]

The other subgroup (type II syndrome) is known as "dyssynergia cerebellaris myoclonica". It is an autosomal recessive, progresive, myoclonic epilepsy characterized with ataxia secondary to mitochondrial encephalopathy.[1]

Magnetic resonance imaging (MRI) is an important tool in the evaluation of the cerebellopontine angle, internal acoustic canal and diseases of the inner ear.[3,4] Higher quality imaging of the internal acoustic canal can recently be obtained using high resolution three dimensional Fourier transformer sequence technique (3DFT-MRI).[3,5] MRI images obtained in the Ramsey Hunt Syndrome can be usually demonstrated by incremental increase in contrast enhancement of cochlea, vestibule and semicircular canals together with the seventh and eighth nerves.[3] This contrast enhancement is especially localized at the labyrinthine segment and geniculate ganglion of the facial nerve.[4] In the literature, this kind of an enhancement usually has been reported at the acute phase of disease and proposed as a result of infection.[6]

Here, we present a case of a Ramsey Hunt Syndrome that is different from most of the cases reported in the literature by showing a contrast enhancement by the eighth week of his complaints. In addition, the contrast enhancement was confined to the intracanalicular area as assessed by MRI, so that it could not be differentiated from acoustic neuroma easily. We also discuss the limited enhancement of intracanalicular area in Ramsey Hunt Syndrome in the light of the present literature.

CASE REPORT

A 60-year-old man presented with facial asymmetry and left ear pain that lasted for one day. On the examination, facial paresis on the left side (Hause Brackmann stage 3) and edema, hyperemia and sensitivity on the left auricle and external auditory canal were recorded. The patient was hospitalized for steroid treatment and blood glucose regulation due to the history of diabetes mellitus. On the second day of his complaints, vesicle formation was observed on the auricle and his facial paresis was progressed to palsy (Hause Brackmann stage 6) with beginning of a severe rotational vertigo. His audiological examination showed a mild-moderate sensorineural hearing loss of 40 dB on the left side. Systemic acyclovir treatment with dose of 400 mg, five times daily and anti vertiginous treatment was added to the therapy with 1 mg/kg/day of prednisolone. After administration of acyclovir and a decreasing dose of steroid medical treatment for 10 days the patient was discharged from the hospital and prescribed an oral vertigo therapy.

After pne month, the examination of the ear was normal, however, vertigo was still present but not as serious as the initial symptoms. Complete left facial nerve palsy and moderate sensorineural hearing loss of 40 dB in the left ear were the major complaints of the patient. An electromyelogram showed electrophysiological signs of complete facial nerve damage, therefore, magnetic resonance imaging (MRI) of the internal auditory canal with the technique of high resolution three dimentional Fourier transform sequences (3DTF-MRI) was performed. Axial and coronal T_1-weighted images were obtained before and after performing gadolinium contrast enhancement. These images demonstrated marked intracanalicular enhancement on the affected side as a mass with 5 mm diameter on the complex of 7^{th} and 8^{th} nerves (Figure 1). At the second follow up that was performed 8 weeks after the initial presentation, his facial paralysis had worsened to Hause Brackmann stage 4. However, his hearing loss did not change. On the follow up MRI, marked contrast enhancement was still evident with minimal regression (Figure 2). We decided to follow the patient periodically by performing repeated examinations and MRIs.

DISCUSSION

Ramsey Hunt Syndrome (Herpes zoster oticus) is the second most seen herpes infection of head and neck region after herpes zoster ophtalmicus.[3] Varicella zoster virus could be a latent form observed at the ganglions through hematogenous way on the viremic phase or through sensorial nerves from vesicules after the viremic phase of primary respiratory infection.[2] Varicella zoster virus may reactivate and cause Ramsey Hunt Syndrome.[1,2]

The pathological findings of the facial nerve in Ramsey Hunt Syndrome are examined in various studies in opposition to Hunt's original theory explained in 1907 that the inflammatory processes do not start only

at geniculate ganglion or not always comprise only this area.[7] In this disease, when facial paralysis accompany the audiological and/or vestibular findings it is known that inflammatory process is localized at nerves inside the internal acoustic canal.[8]

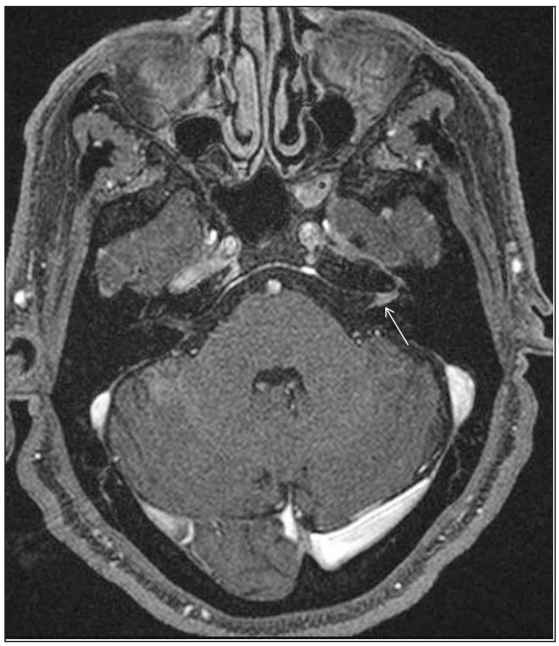

Figure 1. The contrast enhancement inside the left internal acoustic canal is shown on T1 axial scan.

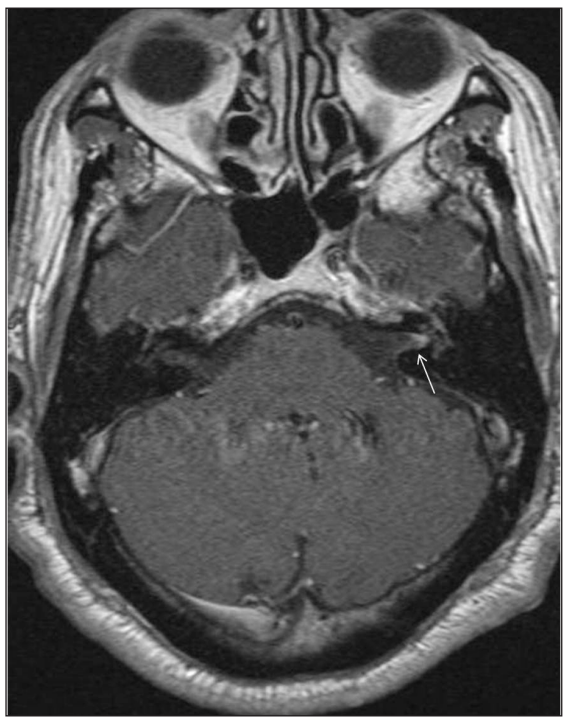

Figure 2. This contrast enhancement is smaller minimally shown on T1 axial scans.

With the improvement in the techniques on MRI especially the use of 3DFT-MRI,[3] evaluation of the internal acoustic canal and structures of inner ear became easier and prevalence of the disease increased. By this way, the contrast enhancement on MRI has been shown not only in acoustic neuroma but also in non-neoplastic diseases, inflammatory neuropathies and post-traumatic facial paralysis. MRI became an important technique for the diagnosis of sensorineural hearing loss with non-neoplastic causes and/or facial paralysis.[4]

Gadolinium diethylenetriamine penthaasetic acid (Gd-DTPA) is a paramagnetic contrast substance that was traditionally used in the imaging of cerebral area and showed a marked enhancement on T1 weighed MRI in cases of destruction of the blood-brain barrier and/ or increased vascular permeability because of impermeable feature of Gd-DTPA normally.[7,8] Enhancement of contrast substance on peripheral nerves is explained as a result of increase in vascular permeability and/or destruction of blood-nerve barrier due to inflammation.[8]

There are many case reports and clinical researches in the literature about the use of MRI with enhancement of Gd-DTPA in the diagnosis and prognosis of Ramsey Hunt Syndrome.[6-9] Yanagida et al.[8] examined MR images of 14 patients with Ramsey Hunt Syndrome and found an enhancement of contrast substance at the distal part of the internal acoustic canal and the labyrinthine segment in most of the cases. This enhancement was especially evident in patients presented with more findings of the seventh cranial nerve. When the findings of the cochleovestibular nerve were added to symptoms of the patients, the contrast enhancement could be seen in the canal or in the inner ear at the images of the patients. Tada et al.[9] could not establish a relationship between findings of MRI and clinical symptoms in patients of Ramsey Hunt Syndrome and proposed that MRI was more specific in the differential diagnosis between Ramsey Hunt Syndrome and tumors.

In a recent study by Kim et al.,[6] the results of decompression surgery of 13 patients with Ramsey Hunt Syndrome was reported. The findings of MRI and surgical results of patients were comparable and the labyrinthine segment was the most frequently established area for the enhancement and pathology.

Unique intracanalicular enhancement on MRI is not prevalent in the patients with Ramsey Hunt Syndrome.[10,11] Particularly, isolated intracanalicular enhancement on MRI is rarely seen during non-infectious period after the acute phase of infection.[11] In this case,

although the correct diagnosis and treatment were applied in the event that herpes vesicules were exist at the present, MRI scans could be evaluated easily as acoustic neuroma. The contrast enhancement on MRI in that patient persisted for a long time which may cause confusion about the diagnosis because of the presence of the contrast substance only in the intracanalicular compartment. Downie et al.[10] published a case of Ramsey Hunt Syndrome with contrast enhancement on MRI with a six months follow-up time. However, this contrast enhancement was found to be located in cochlear and vestibular area by MRI. Goldsmith et al.[11] had reported a case of unique intracanalicular enhancement on MRI in Ramsey Hunt Syndrome which was not easily differentiated from acoustic neuroma resembling our case.

MRI was obtained to identify the continued symptoms of hearing loss, vertigo and facial paralysis although the vesicules on the ear of the patient were improved. We thought that MRI may show the progression of neuritis of the cochleovestibular nerve. Initially, we were a little bit confused about the diagnosis because of persistence of contrast enhancement only in the intracanalicular compartment mimicking acoustic neuroma. After the second MRI, the diagnosis of inflammatory neuritis was certain due to minimal decreasing in the contrast enhancement. However, this was again a rare condition that the contrast enhancement on MRI in RHS or in any inflammatory neuritis was localized only in the intracanalicular area and showing progression more than two months.

In our case, use of different techniques of MRI together with repeated MRI scans was effective in the diagnosis and improved our clinical findings. The evaluation of internal acoustic canal and the cerebellopontine angle was achieved accurately in a short duration. In addition, the reconstruction of scans could be done easily so that the inner ear and the cochleovestibular and the facial nerve have been shown simply with the technique of 3DFT-MRI instead of other conventional spin-echo MRI techniques.[3,5]

When the intracanalicular enhancement was present at the scans of the patients with facial paralysis with sensorineural hearing loss, one of the first possible diagnoses would be acoustic neuroma. However, patient history should be reviewed for varicella zoster infection too. In such cases, mucocutaneous vesicules may not be present and /or cochleovestibular findings may be more evident than facial paralysis.[2] The combination of antiviral and steroid management is suggested to be started in these patients.[1,2]

Although isolated intracanalicular enhancement on MRI is thought to be an acoustic neuroma, even if MRI findings were not evident at the acute phase, patient should also be investigated for Ramsey Hunt Syndrome and other inflammatory neuropathies. It should be kept in mind that a small intracanalicular acoustic neuroma may be confused with a self inflicted disease of Ramsey Hunt Syndrome. Therefore, the most important point in the diagnosis of this disease is the repeated MRI scans and the evaluation of clinical symptoms of the patient during follow-up.

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