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A Rare Cause of Sudden Hearing Loss: Neurosarcoidosis

Nadir Bir Ani İşitme Kaybı Nedeni: Nörosarkoidozis

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ABSTRACT Sarcoidosis is an inflammatory multi-system disorder of unknown etiology. 5-7% of patients with systemic sarcoidosis are detected symptoms of central nervous system involvement, so it is called neurosarcoidosis (NS). The most prevalent symptom in about 80% of NS is a cranial neuropathy, especially facial and optic nerves. In NS patients, eighth nerve involvement is found in 1-7%. In this case, we present a female patient who has a sense of numbness in left half of the face and sudden hearing loss on the same side that responded to the steroid treatment twice in a week, has been diagnosed possible neurosarcoidosis.

Keywords: Sudden hearing loss; sarcoidosis; neurosarcoidosis

ÖZET Sarkoidozis nedeni bilinmeyen inflamatuar multisistemik bir hastalıktır. Yaklaşık sistemik sarkoidozisli hastaların %5-7'sinde santral sinir sistemini içeren semptomlar ortaya çıkar ki bu durum nörosarkoidozis (NS) olarak adlandırılır. NS vakalarının yaklaşık %80'inde en sık semptom başta fasiyal ve optik sinir olmak üzere kraniyal nöropatidir. Sekizinci sinir tutulumu ise NS hastalarının %1-7'sinde bulunur. Bu vaka sunumunda sol yüz yarısında uyuşma ve sonrasında bir hafta içinde 2 kez steroid tedavisine yanıt veren aynı taraflı ani işitme kaybı ile karşılaştığımız ve olanaklı nörosarkoidozis tanısı alan kadın hastadan bahsedilecektir.

Anahtar Kelimeler: Ani işitme kaybı; sarkoidozis; nörosarkoidozis

Sarcoidosis is an inflammatory disease with multi-systemic involvement. Symptoms related with the central nervous system occur in approximately 5-7% of patients with sarcoidosis which in that case the clinical picture is called neurosarcoidosis (NS).[1] In 80% of NS cases the most common symptom is cranial neuropathy, especially facial and optic nerve neuropathy.[1-3] The involvement of the vestibulocochlear nerve occurs in 1-7% of NS patients.[1,2,4,5]

The histopathological correlation of audiovestibular dysfunction in NS was first described with the autopsy findings in a deaf NS patient in 1984.[6] Researchers observed perivascular lymphocytic infiltration resulting with significant axonal degeneration and demyelization especially in the acoustic, vestibular and facial nerves. The hearing loss and vestibular dysfunction in sarcoidosis were thought to be reversible initially. However in some patients it is thought that, irreversible damage occurrs as a result of the ischemic process secondary to vasculitis.[6]

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In 90% of the cases, the hearing loss is sudden or rapidly progressive on onset. Vestibular symptoms characterized with abnormal vestibular function tests were also reported. The recovery of balance problems may be spontaneous or with medication while at least partial healing in the auditory functions is observed with high dose steroid treatment.[6,7]

Here in this case report, a female patient with paresthezia in the left face half who afterwards experienced sudden hearing loss twice on the same side in only a week and responded to steroid treatment, diagnosed with possible NS will be presented. Written informed consent was obtained from patient who participated in this study.

CASE REPORT

The 38 year old female patient who initially experienced numbness in the left face half for 2 weeks applied to the Otolaryngology policlinic with tinnitus and sudden hearing loss. There was not any vestibular complaint. Both tympanic membranes were intact and normal. Pure tone audiometric evaluation revealed mild sensory neural hearing loss on the left. Right auditory functions were evaluated as normal. 200 mg systemic methylprednisolone was administered. The following day of treatment, pure tone audiometric evaluation revealed normal auditory functions. The patient refused the continuing steroid treatment because of her recovery. A week later, same complaints re-occurred. Another pure tone audiometric evaluation revealed mild sensoryneural hearing loss on the same side. Again, after 4 hours of 200 mg methylprednisolone treatment the hearing functions came to normal. However this time 1 mg/kg methylprednisolone treatment was continued in tapering doses for 10 days.

The patient had no additional systemic diseases. The patient's complete blood count, liver function tests, C-reactive protein level and erythrocyte sedimentation rates were found to be at normal levels. Antinuclear antibody (ANA) test resulted positive whereas anti-neutrophil cytoplasmic antibody (ANCA) test resulted negative. Serum angiotensin converting enzyme(ACE) level was found 76 U/L (Normal ranges: 8-52). The temporal magnetic resonance imaging (MRI) was reported normal. No pathological responses was obtained in the caloric tests. The patient's cranial MRI demonstrated millimetric nonspecific ischemic-gliotic lesions scattered in the periventricular and subcortical white substance in T2-weighted and FLAIR sequences (Figure 1a, b). Because of high serum ACE level performed Pulmonary CT scan revealed hiler lymphadenopathy and a well circumscribed sub-pleural nodule with 11x5 mm dimensions on the posterior basal segment of the right inferior lobe (Figure 2a, b). Tuberculin test was negative. Patient's ophthalmological examination was found normal. The patient was diagnosed with possible NS. Systemic steroid treatment was administered for 6 months (16 mg methylprednisolone oral). The patient then had no complaints once the treatment was ceased.

DISCUSSION

Sarcoidosis is a rare disease with an incidence rate of 11-36 in 100 000 individuals yearly.[8] A study in which 285 patients diagnosed with sarcoidosis concluded that 14 patients (5%) had neurosarcoidosis and aduiovestibular sypmtoms were present in only 2 (0,7%) of these cases.[9]

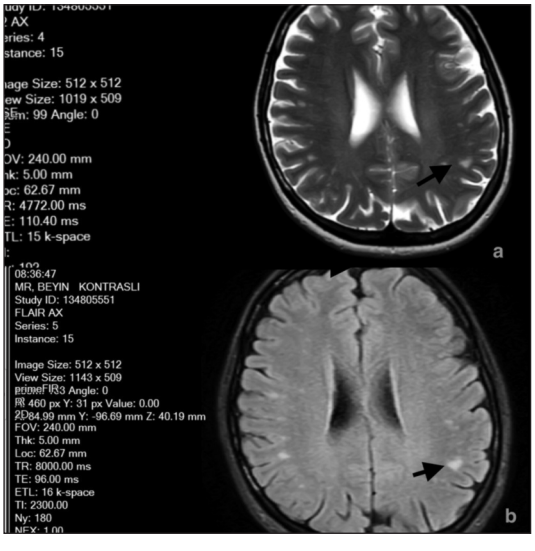

FIGURE 1: Cranial MR images (a, b):Arrows point to the intrcranial lessions.

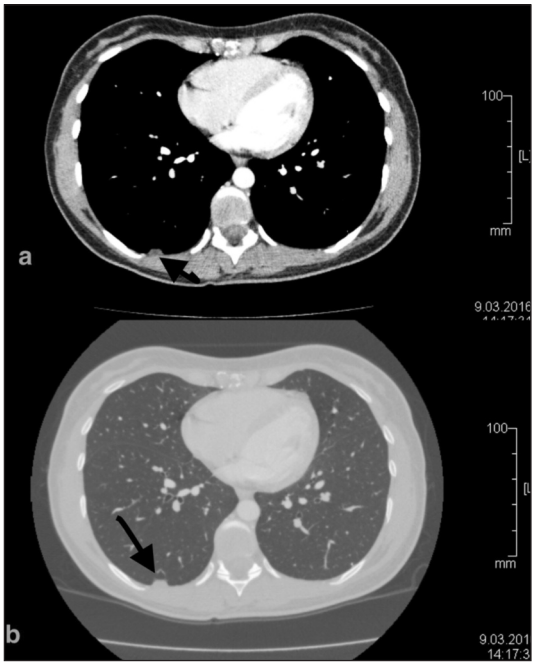

FIGURE 2: Thorax CT images (a, b): Arrows point to the subpleural nodule.

Multiple cranial nerve involvements including the facial and vestibulocochlear nerve, increased uptake of the vestibulocochlear nerve on the MRI and the presence of pleocytosis in cerebrospinal fluid support the diagnosis of NS.[10]

One of the most comprehensive studies in NS patients was reported by Colvin et al. which included 48 published case reports and 2 reviews of cases encountered in audio vestibular clinics.[11] In this study, 50 NS patients were evaluated, 42 (84%) patients presented characteristic histopathological results (biopsy results, postmortem specimen or Kveim test analysis) and most of these patients presented vestibulocochlear symptoms. While hearing loss was observed in 49 patients, 25% of these cases presented with unilateral and 75% of cases presented with bilateral hearing loss. The data revealed 34 patients' records of hearing loss type as 32 (94%) of them had sensorineural hearing loss and 2 (6%) of them had mixed type hearing loss. Conductive type hearing loss was not observed. 22 patients had records of the degree of hearing loss as 1 (5%) patient had normal hearing functions, 6 (27%) patients had mild hearing loss, 7 (32%) patients had moderate hearing loss, 5 (23%) patients had severe hearing loss and 3 (14%) patients had profound hearing loss. Moreover, 30 patients' records of tinnitus complaint were present and in that, 11 had unilateral tinnitus (37%), 6 had bilateral tinnitus (20%). The other 6 patients had tinnitus lacking the laterality information (20%) and in 7 patients tinnitus was not observed at all (23%). 32 patients' vestibular symptoms were evaluated 29 (91%) of them were found to have one or more vestibular system abnormalities.

Zajicek et al. have given three different definitions of NS in 1999.[5] Definite NS requires a positive histological finding in the neurvous system biopsy. For a diagnosis of probable neurosarcoidosis, laboratory support (CSF or MRI) is required as well as evidence of systemic sarcoidosis (histological, Kveim test, and/or two or more indirect indicators: suggestive Gallium scan, chest imaging, or serum ACE). Possible NS is diagnosed when clinical findings are coherent with NS and other causes are excluded.

In 2006, Marangoni et al. have revised this classification.[12] They have suggested that in diagnosing probable NS, the definitive diagnosis of systemic sarcoidosis requires one of the following; positive findings in high resolution thorax computed tomography (CT), a ratio greater than 3.5 of CD4/CD8 ratio in bronchoalveolar lavage or a ratio greater than 5 of CD4/CD8 ratio in cerebrospinal fluid.

According to the Zajicek criteria, this case report matches with possible NS. In this patient, other differential diagnoses have been excluded and NS has been thought after the dramatic response to steroid treatment. This patient had sudden mild hearing loss and recovered after systemic steroid treatment. However, the patient was referred to pulmonology and neurology units a week later when the hearing loss recurred and facial numbness was experienced on the same side. The patient had no neurological symptom except the left-sided facial numbness. Her chest radiography result was normal and thoracic CT showed a subpleural nodule. Even though the patient had symp-

toms of vasculitis in the cranial MRI, it wasn't thought to be the diagnosis after laboratory results. The high levels of ACE supported the NS diagnosis. The patient has been treated with steroid doses that have been decreased for 6 months and she has no current complaints during the continuing follow-ups.

Neurosarcoidosis is rare and when sudden hearing loss occurs, it is very important to think of neurosarcoidosis as a possible diagnosis.

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