

Magnetic resonance imaging findings of Susac syndrome

Susac sendromunun manyetik rezonans görüntüleme bulguları

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Abstract

Susac syndrome is a rare disease of unknown pathogenesis. It is caused by a microangiopathy affecting the arterioles of the brain, retina, and cochlea, giving the classic clinical triad of subacute encephalopathy, visual loss secondary to retinal branch occlusions, and sensorineural hearing loss. Magnetic resonance imaging, retinal fluorescein angiography, and audiography findings enable diagnosis. Early therapy may reduce sequelae and improve recovery. Herein we report a case of Susac syndrome with all the findings of the disease.

Keywords: Magnetic resonance imaging, microangiopathy, Susac syndrome

Özet

Susac sendromu patogenezi bilinmeyen nadir bir hastalıktır. Beyin, retina ve kohleanın etkilendiği mikroanjiyopati sonucunda ortaya çıkar. Bu durum hastalığın tipik triadı olan subakut ensefalopati, retinal arteriollerin tıkanmasına bağlı oluşan görme kaybı ve sensorinöral işitme kaybına neden olur. Manyetik rezonans görüntüleme, retinal floresan anjiyografi ve odyografi bulguları tanıya yardımcıdır. Erken tedavi sekel oluşumunu azaltabilir ve iyileşmeyi hızlandırabilir. Bu yazıda hastalığın tüm bulguları ile birlikte bir Susac sendromlu olguyu sunuyoruz.

Anahtar sözcükler: Manyetik rezonans görüntüleme, mikroanjiyopati, Susac sendromu

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Introduction

Susac's syndrome is an uncommon neurologic disorder of unknown cause. It has been described as a clinical triad of encephalopathy, hearing loss, and branch retinal artery occlusions [1]. This syndrome was first described by John O. Susac in 1979 in two young women presenting with the classic clinical triad of subacute encephalopathy, retinal arteriolar branch occlusions, and sensorineural hearing loss [2]. Clinically, the diagnosis is difficult when the patient presents with only a portion of the triad. Radiologist plays an important role when the disease is suspected. In this circumstance, magnetic resonance imaging (MRI) may be crucial in aiding the neurologist to make the diagnosis. Neuroradiologists and general radiologists frequently attribute the MRI findings seen in patients with Susac's syndrome to multiple sclerosis or acute disseminated encephalomyelitis. Herein we aimed to present the MRI findings of a case with Susac syndrome.

Case report

In September 2011, 24-year-old female patient applied to ophthalmology out-patient clinic with increasing vision loss lasting a few months. The patient with accompanying hearing loss was redirected to neurology out-patient clinic for consultation.

She did not have similar disease history in her family and migren. When she was 7 years old, she had been diagnosed as meningitis and treated at intensive care unit. During treatment, she could not recognize her mother and approximately for 20 days, she had been evaluated with loss of movement. After treatment, hearing loss had been determined as sequelae in her right ear.

In the last 2 years, she had complaints such as slowing in her movements, dragging her right lower extremity and increasing vasculary type head ache. During this time, hearing loss has developed in her left ear. Four years ago, she had started to suffer from learning, memorizing difficulties and nervousness.

Two months ago, she had fallen down and hit her head. One day after this trauma, there was vision loss in the upper half of her right eye. Despite the treatment, findings had not regressed, in ophthalmologic evaluation, upper nasal quadrantopia had been found.

In present evaluation, optic disc and neurological findings were normal, visual acuity was complete. Electroencephelography (EEG), electromyography (EMG) findings were normal. In visual evoked potential (VEP) there was advanced slowing (216 ms). There was bilaterally sensorineural hearing loss (left, at low frequency, right, at all frequencies). In fundus examination, macula was normal. There was perivascular cuffing in temporal part of fovea secondary to previous vasculitis (Figure 1a). Retinal fluorescein angiography revealed filling defect at the temporal lower half of the right eye (Figure 1b).

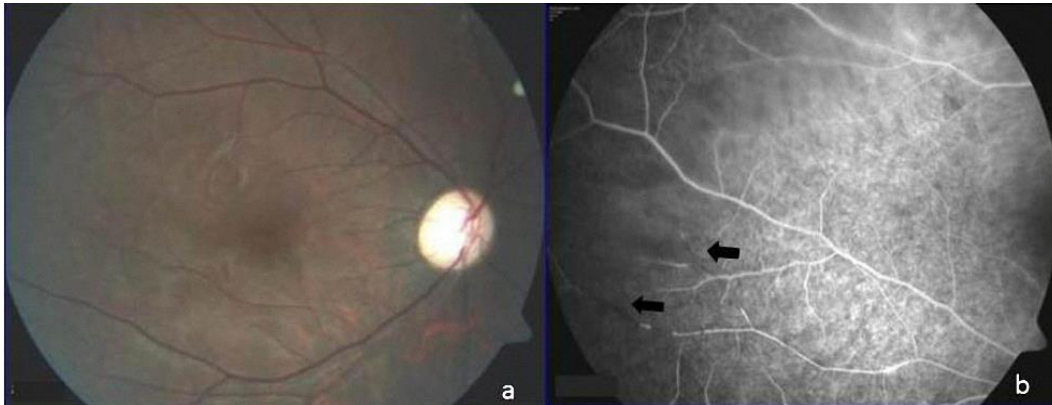


Figure 1 a. In fundus examination, macula was normal. There was perivascular cuffing in temporal part of fovea secondary to previous vasculitis. b. Retinal fluorescein angiography revealed filling defect at the temporal lower half of the right eye (Black arrows).

With these findings, she was referred to our MRI section for cranial MRI examination. In T2-weighted and FLAIR sequences there were hyperintense lesions in both periventricular white matters and corpus callosum. After contrast administration, the lesions did not enhance. Imaging findings of lesions and their discriminations were similar to demyelizing plaques and vasculitis (Figure 2 and 3).

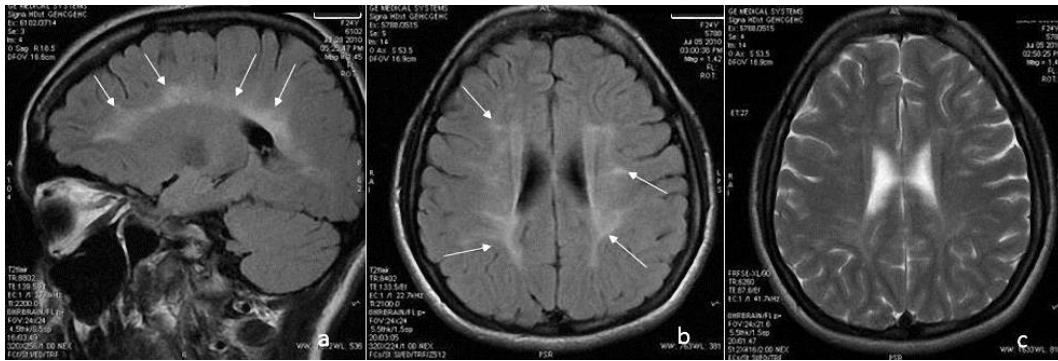


Figure 2. In a. Sagittal FLAIR, b. Axial FLAIR and c. T2-weighted MRI images there are hyperintense foci in periventricular zones and corpus callosum (White arrows).

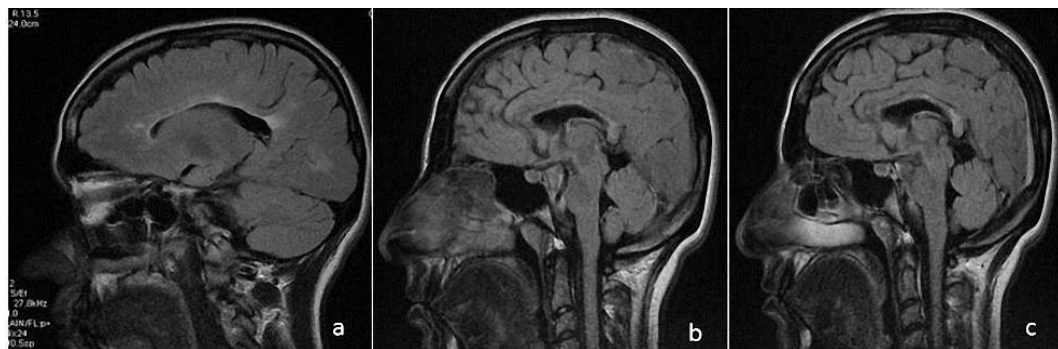


Figure 3. In a. Sagittal FLAIR, b. Sagittal precontrast T1-weighted and c. Sagittal postcontrast T1-weighted MRI images, there are no markedly enhancement.

Our case was diagnosed as Susac syndrome with MRI findings, sensorineural hearing loss and retinal artery occlusions.

Discussion

Susac had first described this pathology, in 1979 with two different female patients who had applied with encephalopathy, retinal artery occlusion and deafness findings. In 1986, Hoyt had named this pathology as ‘Susac syndrome’ [3]. Mass et al. [4], had described this pathology by using the first letters of the findings such as RED-M (retinopathy, encephalopathy, deafness- microangiopathy) [5]. Schwitter et al. [6] had named this syndrome as SICRET (small infarcts of cochlear, retinal, and encephalic tissues), Petty et al. [7], had described as ‘retinocochleocerebral vasculopathy’.

In the literature, most of the cases are between 18-40 years old, healthy female patients with no family history [8]. The disease is very rare and only less than 100 cases have been reported in the literature. Usually, it may be interfered with multiple sclerosis, migren, lupus erythematosus, encephalitis, Meniere disease, thromboembolic infarcts and even schizophrenia [7, 9].

Usually there are active monophasic limiting stages of the disease lasting months and years [9]. As a result of these stages, functional and residual defects can be seen.

Audiometry reveals sensorineural hearing loss that is usually in the low-frequency to midfrequency range. The associated retinopathy is characterized by multiple peripheral retinal arteriolar branch occlusions that can be seen at ophthalmoscopic examination or retinal fluorescein angiography [10-12].

MRI is the referred imaging method for diagnosis. In T2-weighted images, there are many, milimetric, hypertintensities. In postcontrast images, there are diffuse enhancement

in gray and white matters of cerebrum-infratentorial. In acute and subacute stages, the lesions can enhance [7, 8, 11]. Lesions are seen in corpus callosum (genu, corpus and splenium), centrum semiovale, internal capsule, periventricular white matter, brain stem, cerebellum, cerebral and cerebellar periventricular peduncles, basal ganglia and thalamus. These lesions may appear on T1-weighted MRI as hypointense areas during the subacute or late phase of disease, as in this patient. They usually evolve into microinfarction cysts in the late stage of disease. The most striking MRI finding in patients with Susac syndrome is the involvement pattern of the corpus callosum [3, 8, 9, 11]. No abnormal signal intensity and enhancement in cochlea have not been reported in the literature. Cerebral atrophy can be seen in the late stages of the disease. Computed tomography and angiography examinations are applied for differential diagnosis [7, 11].

In our case cranial MRI findings were compatible with the literature but the lesions were generally patchy and the margins of the lesions were insignificant. All of the lesions were mainly in periventricular and corpus callosum regions. There was cranial atrophy additionally. These findings depend on chronic pattern of the disease. Susac syndrome often has a chronic relapsing course punctuated by frequent remissions and exacerbations for 1–2 years [13, 14]. After contrast material administration, there was no enhancement. Retinal fluorescein angiography was performed after recognition of the brain MRI findings. This examination revealed inflamed retinal arterioles with filling defect at the temporal lower half of the right eye indicating vasculitis. In audiometric evaluation there was hearing loss in both ears. These findings were also compatible with the literature. All these MRI, retinal fluorescein angiography and audiometry findings have led us to the Susac syndrome diagnosis.

Susac syndrome can mimic several diseases. The differential diagnosis includes any disorder that can produce multifocal neurologic symptoms, visual impairment, hearing loss, or any combination of these and is therefore very large: Demyelinating disease, connective tissue disease, infection, neoplasm, procoagulant state, and ischemic disease of different etiologies. Several central nervous system infections can cause multifocal neurologic signs, including loss of hearing. The infections may be associated with meningitis, so cerebrospinal fluid examination can help in the detection of microorganisms. Angiography may show abnormalities of arteries at the base of the brain, whereas this examination is usually normal in patients with Susac syndrome [12].

Cranial MRI, retinal fluorescein angiography and audiologic examinations give us great contribution to the diagnosis. As a result of this early treatment regimes can be applied. All the patients with Susac syndrome prediagnosis should be evaluated with clinical, imaging, retinal fluorescein angiography and audiometric examinations.

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