Case Report

MRI of Susac’s Syndrome

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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]. Clinically, the diagnosis is difficult when the patient presents with only a portion of the triad. In this circumstance, 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. In this case report, we review the MRI findings of Susac’s syndrome.

Case Report

A 27-year-old woman presented with frontal headaches, bilateral hearing loss, and visual changes. She complained of difficulty speaking and walking. Her family reported a recent change in her behavior. They noted mood changes and inappropriate gestures. She described several bouts of urinary incontinence in the prior month. There was no medical or surgical history. On physical examination, mild unsteadiness of gait was observed. The visual acuity of her left eye was 20/70, and funduscopic examination revealed a retinal hemorrhage. The right visual acuity was 20/40, and the fundus was normal. No other neurologic deficits or abnormalities were present.

Laboratory studies were negative for antinuclear antibodies, hepatitis panel, Epstein-Barr polymerase chain reaction, cardiolipin antibodies, and herpes infection. CSF analysis was negative for oligoclonal bands and nonreactive for the Venereal Disease Research Laboratory (VDRL) test. Electroencephalography revealed moderate slowing with no epileptiform activity. A gallium scan was negative for adenopathy and for acute inflammation.

Gadolinium-enhanced MRI was performed next. Multiple foci of increased T2 signal intensity in the periventricular white matter, left middle cerebellar peduncle, corpus callosum, and right internal capsule were shown (Figs. 1A–1C). None of the foci enhanced after the administration of gadolinium. Diffusion-weighted images showed increased signal in the right internal capsule. Because the apparent diffusion coefficient (ADC) map was normal, this finding was attributed to T2 "shine-through." Sagittal and coronal FLAIR images also showed areas of increased signal that corresponded to areas of T2 signal change. In the corpus callosum, multiple lesions of mixed signal intensity on FLAIR sequences were identified (Fig. 1D).

These findings were interpreted as representative of Marburg’s variant of multiple sclerosis or CNS vasculitis. One month later, our patient underwent MRI-guided stereotactic brain biopsy. Six separate specimens were obtained from the white matter of the right frontal lobe. The pathologist’s interpretation was white matter reactive glial changes with evidence of degeneration. The white matter was well myelinated on Luxol fast blue stain and periodic acid–Schiff stain. The axon density was normal with the Bodian method. No definitive evidence of demyelination or neoplasm was found. Electron microscopy showed no viral particles.

The patient’s symptoms improved after plasma exchange but did not improve with steroids. A second MRI examination with gadolinium enhancement was performed 4 months after the initial study. Again shown were multiple periventricular white matter lesions of high signal intensity on T2-weighted and FLAIR sequences (Figs. 1G–1J). The right internal capsule lesion had resolved. The posterior fossa lesions were diminished. Multiple lesions in the body and splenium of the corpus callosum were of increased signal intensity on T2-weighted images and decreased signal on T1-weighted images. None of the lesions enhanced with gadolinium, and they were essentially unchanged. These well-demarcated callosal changes had a “punched-out” appearance (Figs. 1A and 1G). They were centrally located within the corpus callosum fiber tracts and were similar in appear-

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ence compared with their appearance on the initial examination.

Discussion

Susac’s syndrome was first described in 1979 [1, 2]. This syndrome has also been referred to as “SICRET syndrome” (small infarctions of cochlear, retinal, and encephalic tissue) or “RED-M” (microangiopathy with retinopathy, encephalopathy, and deafness) [1]. It involves a clinical triad that includes encephalopathy, retinal arterial branch occlusions, and bilateral hearing loss [1–6]. The exact cause is unknown, but it is believed to be an immune response with microembolization [1, 2].

Susac’s syndrome is seen most commonly in young women 20–40 years old [1–6]. Multiple cases have been reported in men [1, 3]. This entity has also been described in the Japanese-, Chinese-, French-, German-, Spanish-, and English-language literature [1–6]. Therefore, Susac’s syndrome occurs in all races.

The encephalopathic changes can manifest as memory impairment, confusion, behavioral disturbances, ataxia, dysarthria, parainoid psychosis, occasional mutism, and headaches [1–6]. The optic changes include scotomata and vision distortion that are due to branch retinal artery occlusions [1–6]. Retinal irregularities may be seen on ophthalmoscopy examination or fluorescein angiography [2]. Cochleovestibular symptoms involve hearing loss at low and medium frequencies [1, 2, 4]. Of all these symptoms, headaches with encephalopathy are the most common presentation [2].

The classic triad is pathognomonic for Susac’s syndrome, but the three elements are not always present at the same time [1].

Most case reports and articles concerning Susac’s syndrome have been published in the neurology, otorhinolaryngology, and ophthalmology literature [2–6]. In 2003, Susac et al. [1] reported the largest series of cases, and the 27 MRI studies reviewed showed characteristic changes within the corpus callosum. These changes consisted of small well-demarcated, spherical, high-signal-intensity lesions located in the body and splenium of the corpus callosum on FLAIR and T2 sequences [1]. The lesions had a punched-out appearance and were located centrally within the fiber tracts of the corpus callosum. Susac et al. [1] attributed these lesions to microinfarctions. They, along with other researchers,
have reported that the MRI findings are commonly confused with demyelinating processes, typically multiple sclerosis or acute disseminated encephalomyelitis [1, 3, 4]. However, in multiple sclerosis the ependymal undersurface of the corpus callosum is usually involved and callosal atrophy is usually seen [1, 7].

Callosal lesions located centrally within the fiber tracts without callosal atrophy, therefore, are highly suggestive of Susac’s syndrome. In addition, high-signal-intensity changes are usually seen in the periventricular white matter and the deep gray matter [1]. Contrast enhancement of the white matter, gray matter, and leptomeninges may be seen, of which the latter is the least common [1].

During the preparation of this article, two other articles regarding Susac’s syndrome were published. In the first article, Do et al. [8] reported another four cases, but lesions in the corpus callosum were not seen in those cases. In the second article, White et al. [9] retrospectively reviewed the diffusion-weighted images and ADC findings of serial MRI studies. Their conclusion was that Susac’s syndrome may show increased signal intensity on diffusion-weighted imaging, depending on the time of patient presentation. In our patient, no diffusion-weighted images or ADC abnormalities were present with regard to the corpus or other white matter structures (Figs. 1E and 1F).

Although Susac’s syndrome is usually self-limited and may stabilize, it can also lead to complete deafness and blindness [1–3]. If multiple sclerosis is suspected and only corticosteroids are administered, the disease may progress. If Susac’s syndrome is suspected, immunosuppressive or antithrombotic agents (or both) should be considered as part of the treatment regimen [2].

In conclusion, in patients who present without the classic triad of Susac’s syndrome, MRI may aid the clinician in establishing the correct diagnosis. Because treatment plans differ from those for demyelinating diseases, it is important for the radiologist to be aware of this entity. Thus, radiologists should include Susac’s syndrome in the differential diagnosis when punched-out high-signal-intensity lesions are present within the central fibers of the corpus callosum on T2-weighted images.

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References


Fig. 1. (continued)—27-year-old woman with Susac’s syndrome.

G–J, Images of follow-up studies obtained 4 months after A–F.

G–I, Sagittal T1-weighted (G) (500/14), sagittal T2-weighted (H) (4,020/96), and sagittal FLAIR image (I) (9,000/76; inversion time, 2,500 msec; flip angle, 130°) images again show the characteristic "punched out" callosal lesions of Susac’s syndrome.

J, Sagittal T1-weighted image (TR/TE, 500/14) left of midline more clearly shows two cerebellar lesions; these lesions are not significantly changed on this examination compared with initial MRI examination (C).