

# CLINICAL CHALLENGES

PETER SAVINO AND HELEN DANESH-MEYER, EDITORS

## Retinal Infarcts in a Patient with an Acute Confusional Syndrome

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*(In keeping with the format of a clinical pathologic conference,  
the abstract and key words appear at the end of the article.)*

### Case Report

A 46-year-old man came to the emergency room with confusional syndrome and behavioural disturbances. He smoked compulsively, but had great difficulty lighting a cigarette and had an alteration of his sleeping rhythm. He also had impaired cognition and memory, ataxia, and dysarthria. Two days prior to presentation, he developed a headache (without neck stiffness) and paresthesias in the right half of his face and body.

His past medical history was significant for headaches thought related to alcohol consumption. Social history included moderate alcoholic intake and occasional cannabis and methadone consumption. Routine laboratory studies, including complete blood count (CBC), electrolytes, and urine and blood drug screens, were normal, and no focal neurologic abnormality was found. Brain CT with contrast was normal. A lumbar puncture showed a moderate increase of the protein in cerebrospinal fluid (CSF) to 153 mg/dl.

In the emergency room the patient improved, so he was diagnosed by the neurologist as having a complicated migraine, and he was discharged with analgesic treatment. However, 72 hours later, the neurologic symptoms became worse, and the patient was again confused, somnolent, disoriented, and had incoherent speech.

He was admitted to the hospital and repeat laboratory tests (CBC, electrolytes, ESR, blood and urine toxic screen), and a new brain contrast enhanced CT were normal. Repeat lumbar puncture showed more CSF protein (194 mg/dl) than in the first lumbar puncture.

A magnetic resonance image (MRI) disclosed multiple small foci of cerebral microinfarctions in white and gray matter, but especially in the corpus callosum (Fig. 1). Cerebral catheter arteriography was normal. An electroencephalogram showed general slowing of electrical activity, suggestive of diffuse cerebral encephalopathy. The patient did not have visual acuity loss. The ophthalmologic

abnormalities were limited to the fundus examination as shown in Fig. 2.

*What diagnoses are you considering?*

*What further investigations would you recommend?*

### Comments

#### COMMENTS BY JOHN O. SUSAC, MD

This 46-year-old man presented with headache, encephalopathy, and focal sensory complaints. He was afebrile and had a normal CT scan. His MRI scan showed microinfarcts in gray and white matter—including the corpus callosum. Fig. 1 is an axial Flair image that shows only a small part of the corpus callosum (and no lesions in genu or splenium). I believe that this man has Susac syndrome (SS)—microangiopathy of the brain, retina, and cochlea—and in this instance, the sagittal Flair is best to demonstrate the callosal lesions. Fig. 2 shows branch retinal artery occlusions that constitute the second part of the triad of SS. The elevated protein in his spinal fluid and diffusely abnormal EEG are consistent with his encephalopathy. Arteriography is normal in SS because the size of the affected precapillary arterioles is only 100 microns.

I would recommend fluorescein angiography to see the characteristic staining of the retinal arterioles and an audiogram. In addition, I would send serum for antiendothelial cell antibodies that have been described in SS.<sup>8</sup>

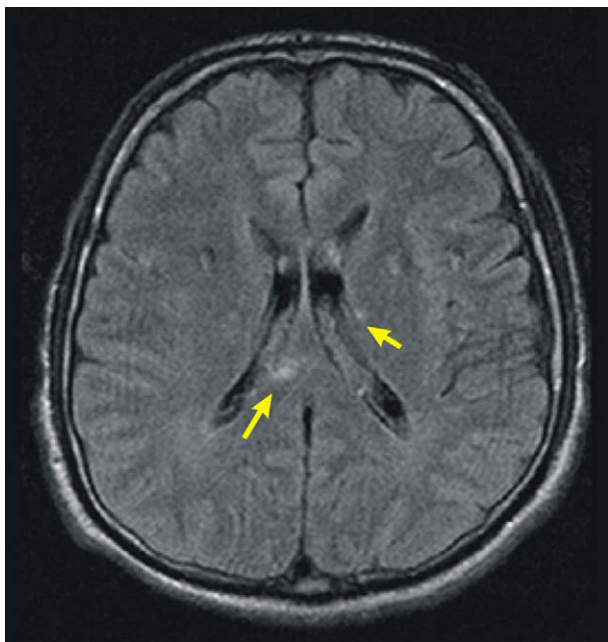


Fig 1. Flair MRI of the brain shows multiple microinfarctions in white matter and the corpus callosum (arrows).

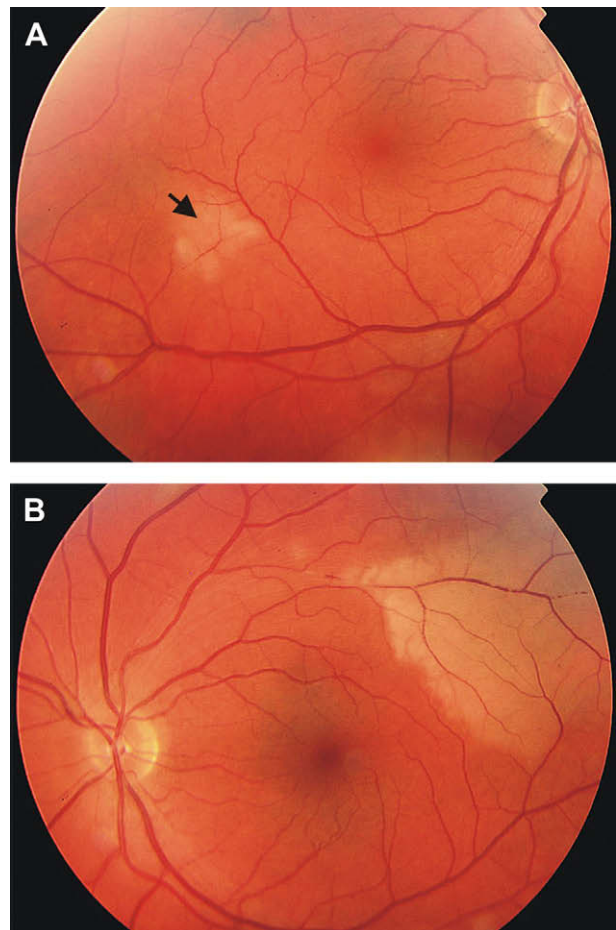


Fig 2. A. Fundus of the right eye with a peripheral retinal infarct (arrow) in the inferior temporal vascular arcade. B. Fundus of the left eye with a peripheral retinal infarct in the superior temporal vascular arcade.

### Case Report (Continued)

Fluorescein angiography showed hyperfluorescence of the arteriolar walls (Figs. 3, 4 and 5).

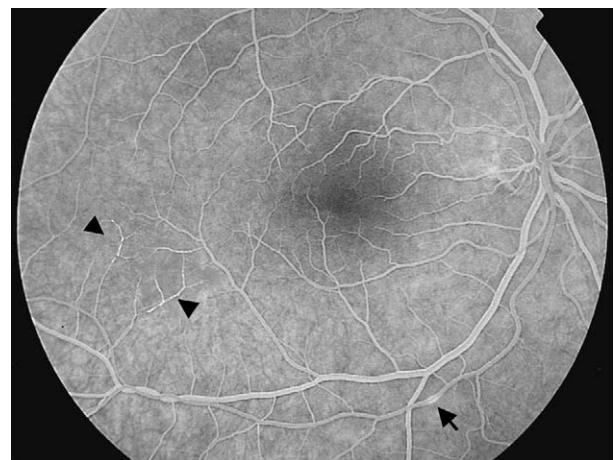


Fig 3. Fluorescein angiogram of the right eye revealed multiple vessel occlusions in the temporal inferior arcade (arrows).

Because both cerebral and retinal microinfarctions were found, Susac syndrome was suspected. An audiometric evaluation, however, was negative.

*What treatment would you institute?*

### Comments (Continued)

#### COMMENTS BY DR. SUSAC

Treatment requires immunosuppression. A treatment protocol has recently been proposed.<sup>6</sup> We would treat this man aggressively with high-dose steroids, including frequent pulses of intravenous methyl prednisolone and intravenous immunoglobulin (IVIG). After the first week, we then would decide whether to add cyclophosphamide or mycophenylate mofetil. We have also used rituximab in several patients. Treatment must be early, aggressive, and sustained.<sup>4,5</sup>

### Case Report (Continued)

Treatment was begun with aspirin 300 mg/24 h and intravenous methylprednisolone 1 g/day for 3 days, followed by an oral taper. The neurologic symptoms quickly resolved with this regimen. However, as the corticosteroid was tapered to 40 mg per day, the symptoms recurred. Again he developed apathy, confusion, and sleeping rhythm alterations. He again was admitted and a new MRI revealed new ischemic lesions in the right basal ganglia region. Contrast enhancement showed leptomeningeal enhancement compatible with meningoencephalitis. The ophthalmologic examination did not show new retinal infarcts. The diagnosis of an exacerbation of Susac syndrome was made and treatment with methylprednisolone 1 g/day for 3



Fig. 4. Fluorescein angiogram of the left eye revealed multiple vessel occlusions in temporal superior and inferior arcades (arrows).



Fig. 5. Fluorescein angiogram of the left eye showing hyperfluorescence of superior vessels (arrow head) and vascular occlusions (arrows).

days was reinstated and he was administered 5 days of IVIG. The patient had a good response with complete resolution of the symptoms.

### Discussion

Susac syndrome is a rare disease of unknown pathogenesis. It is characterized by an arteriolar microangiopathy that affects brain, retina, and cochlea, producing encephalitis, retinal infarcts, and neurosensory hearing loss.<sup>3,7</sup> This disease is commonly underdiagnosed<sup>9,11</sup> and requires a high index of suspicion to make the correct diagnosis. For this reason the ophthalmoscopic and the Fluorescein angiographic findings are extremely important.<sup>1,2</sup> An appropriate therapy, given as soon as possible, is important in order to reduce permanent sequelae, such as sensorineural hearing loss, visual loss, and cognitive deficits.<sup>10</sup>

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The authors reported no proprietary or commercial interest in any product mentioned or concept discussed in this article.

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**Abstract.** A 46-year-old man presented with acute confusional syndrome, ataxia, dysarthria, and right hemiparesthesia. Brain MRI showed small bilateral infarcts and fluorescein angiography revealed multiple peripheral retinal infarcts bilaterally. No visual loss was present, and no other organs were involved. The diagnosis of Susac syndrome (microangiopathy of the brain, retina and cochlea) was made and immunosuppressive therapy begun. (*Surv Ophthalmol* 54:503–506, 2009. © 2009 Elsevier Inc. All rights reserved.)

**Key words.** cerebral infarct • retinal infarct • Susac syndrome