A case report on Susac syndrome

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To the Editor: Susac syndrome (SS), a syndrome affecting the brain along with retinal and cochlear microangiopathy, is characterized by acute multiple encephalopathy, branch retinal artery occlusion and sensorineural deafness. This disease was first described in 1979, but it was named formally in 1994.[1] As a rare disease, only approximately 300 cases have ever been reported.[2] In China, SS is seldom reported. This case report describes the clinical and imaging features of a female patient diagnosed with SS.

A 41-year-old woman was admitted to the Department of Neurology on October 27, 2017, mainly complaining of vertigo, vomiting and fatigue for 1 day. She had acute onset and was admitted to the Emergency Department of Fourth Hospital of Hebei Medical University. At the time of admission, her blood pressure was 202/120 mmHg. A quick brain computed tomography (CT) scan showed multiple intracranial lacunar cerebral infarctions [Figure 1A]. After admission, the patient complained of blurred vision with foreign-body sensation. When the ophthalmologist asked for her medical history, she remembered that her visual acuity decline occurred approximately 2 weeks ago.

Although the patient looked indifferently and responded slowly, her general and neurologic examinations were normal. Her vision for both eyes was 0.16/light perception, and intraocular pressures were 17/19 mmHg. Slit-lamp examination of her eyes revealed normal lids, lashes, conjunctiva, and sclera and clear corneas. The anterior chambers, irises, lenses, and vitreous were normal in both eyes. The pupil size of the right eye was normal. However, the left eye was slightly dilated, and its light reflex was blunted.

The ocular fundus of the right eye showed there was some spot-like hard exudation around the inferior temporal branch artery [Figure 1E]. It presented that blocked arterioles could be seen in all of the branches of retinal artery periphery in the left eye [Figure 1F]. The infrared fundus images showed branch artery occlusion in the left eye, and the branch of the superior temporal was blocked more completely. Optical coherence tomography (OCT) of the right eye showed no abnormalities [Figure 1G]. OCT indicated that the inner retinal layers became thinner and the ellipsoid zone of the macular area and the outer segment of the photoreceptor cells showed high reflection in the left eye [Figure 1H]. This patient did not undergo fluorescein angiography (FA) examination because of her poor physical condition.

Magnetic resonance imaging of the head indicated multiple hypointense lesions on T1 and hyperintense lesions on T2 located in the brainstem, bilateral basal ganglia, internal capsule-radiation crown and outer capsule area, thalamus dorsalis, right insular lobe, corpus callosum and central ovale. The boundaries of some lesions were not clear [Figure 1B–1D].

On the diffusion-weighted images, there were multiple hyperintense lesions located in the corona radiata, thalamus dorsalis, corpus callosum and central ovale. The fluid-attenuated inversion recovery images demonstrated many hyperintense foci in the deep white matter of the frontal lobe, occipital lobe and parietal lobe. Additionally, hyperintense foci existed near the bilateral ventricles. All of these foci exhibited no contrast enhancement after the intravenous administration of contrast material. The configurations of the ventricles, anfractuosity, and schizencephaly were normal and without dislocation.

Magnetic resonance angiography of the head indicated that bilaterally cerebral arteries walked naturally, and the vascular wall was not smooth. No obvious signs of stenosis were found.

The otolaryngologist evaluated her hearing. Measurement of pure sound hearing thresholds showed severe deafness in the left ear and deafness in the right ear. Left acoustic immittance showed type A.

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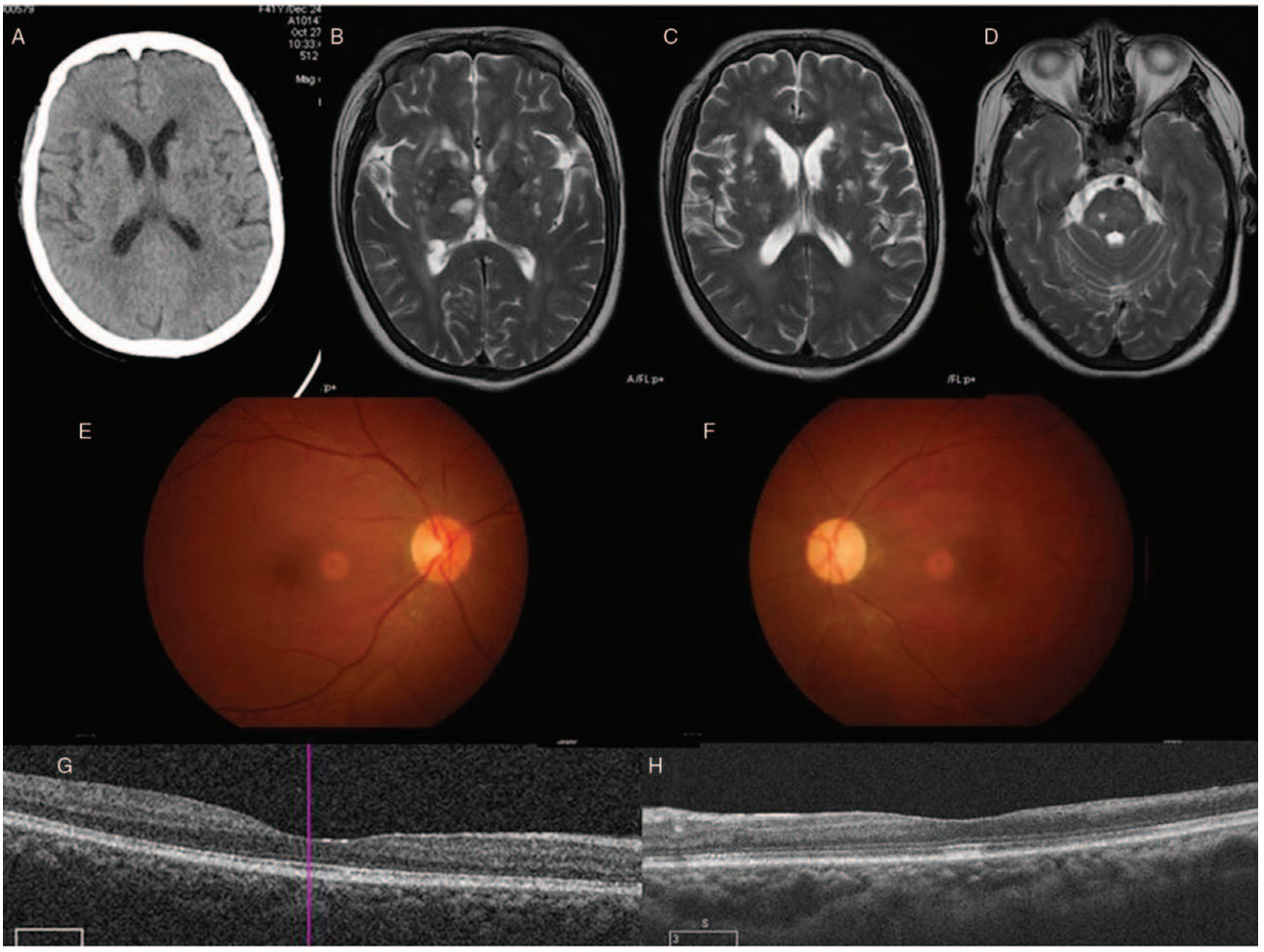

Figure 1: Representative image of the patient. (A) CT scan indicated multiple small hypointense lesions existed in bilateral basal ganglia and pontine. (B, C) Magnetic resonance images showed there were multiple hyperintense lesions on T2 in the periventricular area (right thalamus dorsalis and bilateral basal ganglia). (D) It showed an old hyperintense lesion in pontine. (E) The fundus of the right eye showed there was some spot-like hard exudation around the inferior temporal branch artery. (F) It presented that blocked arterioles could be seen in all of the branches of retinal artery periphery in the left eye. (G) OCT of the right eye showed no abnormalities. (H) OCT indicated that the inner retinal layers became thinner and the ellipsoid zone of the macular area and the outer segment of the photoreceptor cells showed high reflection in the left eye. CT: Computed tomography; OCT: Optical coherence tomography.

Routine blood tests showed that she had elevated levels of platelets, white blood cells, and neutrophil granulocytes and a decreased level of hemoglobin. Urine analysis showed that her epithelial cells were increased and her red and white blood cells were significantly increased. Her serum triglyceride and very-low-density lipoproteins were high. To identify any further risk factors, we undertook more detailed laboratory examinations. Her blood levels of autoantibodies for vasculitis and antineutrophil cytoplasmic antibodies and her erythrocyte sedimentation rate were normal. Her level of C-reactive protein was slightly high.

Taking into account the findings, we considered a final diagnosis of SS. We gave as treatment an intravenous infusion of methylprednisolone (500 mg per day) for three days, followed by oral administration of prednisone (60 mg once a day) with a slow taper. After 1 week, her symptoms of vertigo and headache improved gradually, and she was discharged from the hospital.

SS is a rare disease with an unclear pathogenesis, and an autoimmune etiology is presumed for this disorder. Studies report that up to 30% of patients have found presence of anti-endothelin antibodies, and there is a good response to immunosuppressive therapy.[3] SS affects women three times more often than it affects men.

Typical clinical manifestations include nervous system disease, retinopathy and vestibule cochlear lesions. As reported, only 13% of patients present with the classic triad, but all three symptoms manifest over weeks or months for most cases. For SS, the differential diagnosis includes disseminated sclerosis, primary central nervous system lymphoma, infectious cerebral arteritis, central nervous system sarcoidosis, systemic lupus erythematosus, and so on.

At present, no standard treatment has been developed. Immunosuppression and immunoregulation drugs are mainly recommended. Steroid drugs, anticoagulant therapy, antiplatelet therapy, and calcium antagonists are also used as adjuvant therapy.[4] Some others have suggested that high-dose immunoglobulin intravenous injections and hyperbaric oxygen therapy may be helpful.

SS is characterized by being self-limited, waviness and monophasic. Few are recurrent and progressive. Most patients stabilize or experience relief of their symptoms after 2 years.[5] Thus, a patient who is diagnosed in a timely fashion and receives early treatment will acquire a good prognosis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for the images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Conflicts of interest

None.

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