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CASE REPORT

Systemic Lupus Erythematosus and Antiphospholipid Syndrome Related Retinal Vasculitis Mimicking Ocular Cysticercosis: a Case Report

Chan Wu[1], Fang-tian Dong[1]*, You-xin Chen[1], Qian Wang[2], Rong-ping Dai[1], and Hua Zhang[1]

[1]Department of Ophthalmology, [2]Department of Rheumatology, Peking Union Medical College Hospital, Chinese Academy of Medical Sciences & Peking Union Medical College, Beijing 100730, China

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MAKING accurate and timely diagnosis is often challenging when patients with a systemic disease first present with ocular manifestations. The possibility that vasculitis associated with systemic lupus erythematosus (SLE) and antiphospholipid syndrome (APS) can be misdiagnosed as cysticercosis has not been discussed in the literatures.

CASE DESCRIPTION

A 45-year-old woman was referred to the emergency department of our hospital with progressive vision loss bilaterally for 5 days. She also reported dyspnea and palpitation in the past 4 months which were worsening for a week before her presentation. She was suspected with ocular cysticercosis (OCC) in a local hospital because she was diagnosed with neurocysticercosis after a seizure 7 years ago. Cysticercal serum enzyme-linked immuno-electrotransfer blot assay was positive at that time, and intermittent treatment for cysticercosis was administered. She had consequent seizure recurrence several times since then and developed uncontrollable hypertension for 7 years, interstitial lung disease and anemia for 2 years. She suffered from spontaneous abortion 5 times. Her family history was unremarkable.

On examination, the best-corrected visual acuity (BCVA) in the right eye was 20/400, Jr7 and 20/200, Jr5 in the left eye. Intraocular pressure was normal bilaterally. Slit lamp examination of the right eye showed mild pupillary irregularity with mild relative afferent pupillary defect. Both eyes showed clear lens with no anterior chamber or vitreous reaction and no subconjunctival cyst. Dilated fundus examination revealed diffuse vasculitis with widespread intraretinal hemorrhages in both eyes. Additionally, yellow-white lesions of approximately 1.5 prism diopters were noticed across the macular area of both eyes. The optic nerve head showed 0.6 cupping in the right eye and 0.5 in the left eye with no papillary edema (Fig. 1A). Ultrasonography revealed vitreous opacities and round dense echoes corresponding to the macular lesions bilaterally (Fig. 1B). Optical coherence tomography (OCT)

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*Corresponding author E-mail: d_fangtian@sina.com

showed thickening neuroepithelium with a hyper-reflective portion within it in the macular area (Fig. 1C). Fluorescein angiography (FFA) of both eyes revealed slower filling of vessels and peripheral areas of nonperfusion. It also showed blocked fluorescence in the macular areas. The late phase FFA showed significant dye leakage from the retinal veins and abnormal vessels in nonperfusion areas (Fig. 1D).

The patient's general condition deteriorated 3 days later, and she was found to have severe mitral regurgitation with suspected vegetation. She also began to complain of pain in the right eye. Ophthalmological examination showed the BCVA of the right eye decreased to counting fingers at 1 meter. Intraocular pressure of the right eye was 43 mm Hg, while that of the left eye was normal. Slit lamp examination revealed significant iris neovascularization and corneal edema in the right eye. Dilated fundus examination revealed worsening intraretinal hemorrhages. Retinal vasculitis and neovascular glaucoma were considered. Panretinal photocoagulation and topical anti-glaucoma eye drops were administered immediately.

Her complete blood cell count revealed pancytopenia with normal eosinophil cell count. The urinalysis revealed proteinuria and red blood cells. The erythrocyte sedimentation rate was mildly elevated and the complement (CH50, C3, C4) levels decreased. Antibody measurements showed positive antinuclear antibody (ANA) and anticardiolipin antibody (ACLA), while the anticysticercus antibody was negative.

Magnetic resonance imaging of the brain showed multiple discrete white matter lesions in the right periventricular area and bilateral temporal lobes. The patient was diagnosed as systemic lupus erythematosus and antiphospholipid syndrome with lupus nephritis and neuropsychiatric lupus. Prednisolone in combination with cyclophosphamide was initiated. Intravitreal injection of triamcinolone acetonide and bevacizumab was applied to the right eye, and panretinal photocoagulation was continued in both eyes.

After above treatment for 2 weeks, the patient's clinical condition was significantly improved. The ophthalmological examination showed improved vision in both eyes. The BCVA of the right eye was 20/1000, and the left eye was 20/50. Intraocular pressure of the right eye reduced to 20 mm Hg. Slit lamp examination revealed partial regression of iris neovascularization in the right eye and the cornea turned clear. The right pupil was larger and relative afferent pupillary defect was remarkable. Fundus examination showed the optic disc was pale in the right eyes with a cup-disk ratio of 0.9. The hemorrhages in both eyes were absorbed, and the macular yellow-white lesions disappeared (Fig. 2A). OCT showed the neuroepithelium of the fovea was thinning and the hyper-reflective area disappeared (Fig. 2B).

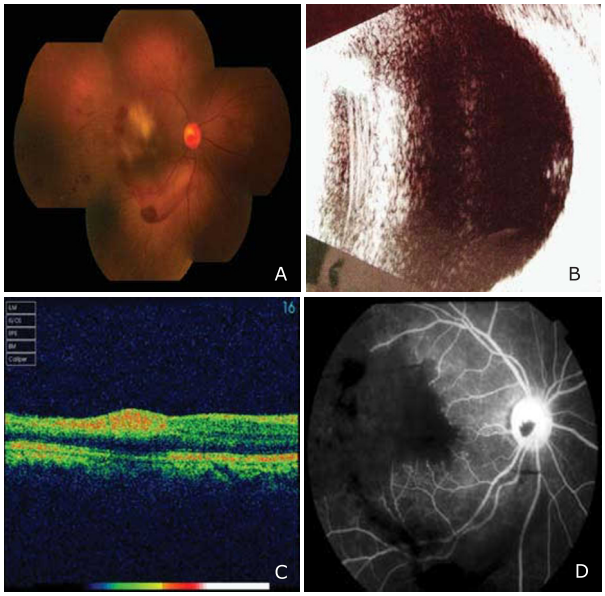

Figure 1. Patient at initial visit.

A. Fundus photograph showing diffuse vasculitis with widespread intraretinal hemorrhages and yellow-white lesion across the macular area in the right eye.

B. Ultrasonography showing round dense echo corresponding to the macular lesion.

C. Optical coherence tomography showing thickening neuroepithelium with a hyper-reflective portion within it in the macula.

D. Fluorescein angiography showing blocked fluorescence in the macular area with vaso-occlusion.

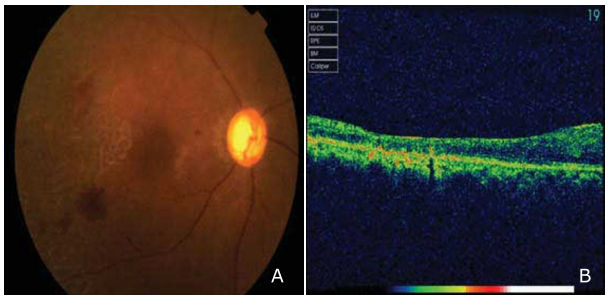

Figure 2. Patient after treatment.

A. Fundus photograph showing the hemorrhages and yellow-white lesion in the macula were absorbed.

B. Optical coherence tomography showing the neuroepithelium in the macula was thinning without hyper-reflective signals.