Bilateral exudative retinal detachment in a patient with cerebral venous sinus thrombosis: a case report

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Case report

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Abstract

Background: Cerebral venous sinus thrombosis (CVST) is a rare cerebrovascular disease, it's ocular symptoms often characterized by a subacute bilateral visual loss, or diplopia and paralysis of eye movements. Fundus examination usually presents as bilateral papilledema and other ocular signs are rare. We report a case of bilateral multiple retinal detachments and finally diagnosed as CVST. Case presentation: A 49-year old woman with progressive headache and bilateral vision deterioration visited our clinic. Ophthaomological examinations including medical history, best-corrected visual acuity, intraocular pressure, slit-lamp biomicroscopy, fundus ophthalmoscopy, fluorescein angiography and Optical coherence tomography and head Magnetic Resonance Venogram (MRV) was also performed. Blood tests for ruling out systemic diseases were also performed. Fundus exam revealed bilateral multiple retinal detachment with sub-retinal fluid and blurred disc margin. Fluorescein angiography (FA) revealed early hypofluorescence in the background stage, multiple pinpoint leakages at the level of retinal pigment epithelium (RPE), and late pooling to outline the boundary of retinal detachment, with some of the leakage shaped as multiple circles in the late stage of FA. OCT revealed retinal and choroid folds, bilateral serous effusion in the sub neuro-retinal area. Lab results showed PPD 1:2000 (+++), T-spot test were positive; Head MRV showed thrombosis formation in the left transverse venous sinus. Conclusions: The patient demonstrated that CVST would cause retinal capillary ischemia and lead to sub-retinal lesions and retinal detachments. Cautions should be taken when patients presenting common ocular characteristics with usual systemic conditions.

Background

Cerebral venous sinus thrombosis (CVST) is a rare cerebrovascular disease which usually affects middle aged population, among which 75% are women. The risk factors of CVST including infection, oral contraceptive usage, puerperium, coagulation disorders and malignancy, etc(Ferro et al. ; Stam). Clinical presentations of CVST can be highly variable, severe headache is the most common symptom, especially in adult patients. The thrombosis can develop in all sinuses, large ones for example the superior sagittal sinus are most likely to be involved(Allroggen & Abbott 2000; Piazza 2012). Ocular symptoms are often characterized by a subacute bilateral visual loss, sometimes with diplopia and paralysis of eye movements. Funduscopic examnination usually reveals bilateral papilledema, which has been broadly reported(Wang et al. 2011). We presented a case of a 49-year-old woman with bilateral exudative retinal detachment and finally diagnosed as CVST.

Case Presentation

A woman in her late-40s visited our clinic with main complaints of progressive headache and bilateral vision deterioration for one week. She denied nausea, vomiting, motor and sensory disturbance, history of trauma, cancer and oral contraceptives. Her blood pressure, neurologic and systemic examinations were negative. Head CT was insignificant. Eye examination showed bilateral best corrected visual acuity (BCVA) were 20/200. Intraocular pressure was normal. Anterior segment examinations were negative.

Fundus exam revealed bilateral multiple disciform neuro-retinal detachment with mobile sub-retinal fluid and blurred disc margin, also noted the multiple polygonal yellowish-white islands in the sub-retinal areas (Fig. 1). Fluorescein angiography (FA) revealed early hypofluorescence in the background stage, multiple pinpoint leakages at the level of retinal pigment epithelium (RPE), and late pooling to outline the boundary of retinal detachment, with some of the leakage shaped as multiple circles in the late stage of FA (Fig. 2). OCT revealed retinal and choroid folds, bilateral serous effusion in the sub neuro-retinal area (Fig. 3). Lab results showed purified protein derivative (PPD) 1:2000 (+++), T-spot test were positive; CT scan of the lungs showed nodules, exudations and cord shadows in double upper lung fields. Blood chemistries, erythrocyte sedimentation rate, pregnancy and autoimmune tests, hyper-coagulation studies were unremarkable. Cerebrospinal fluid tests were normal. The patient initiated with severe headache and head CT was unremarkable, thus the head MRI and MRV were ordered. The head MRV showed thrombosis formation in the left transverse venous sinus. (Fig. 4). The patient was diagnosed as multiple infarctions of choroid arteries and capillaries with cerebral venous sinus thrombosis and was treated with low molecular heparin sodium 4100 IU Bid for anti-coagulation therapy, along with oral anti tuberculosis therapy. One month after, the patient complained that her symptoms improved greatly, fundus examination and OCT showed bilateral exudative retinal detachment and yellowish-white lesions completely resolved. On multispectral imaging, the elschnig spots were presented as subtle depigmentation and pigmentation clumps. As a result, her BCVA improve to 14/20.

Discussion And Conclusions

CVST as a rare cerebral vascular disease, it’s ocular magnification usually are blurred vision and papilledema, resulted by high intracranial pressure. Interestingly, the patient was presented with multiple exudative retinal detachments and multiple pinpoint leakages on FA, which is unusual in CVST and can be easily misdiagnosed as Vogt-Koyanagi-Harada syndrome (VKH) and multiple serous chorioretinopathy. However, Both of them do not cause severe headache and without yellow-white sub-retinal lesions. The sub-retinal yellowish-white lesions are Elschnig Spots, which usually present in aggressive hypertension and pre-eclampsia, indicating infarction of the choriocapillaries(Tso & Jampol 1982; Larcan et al. 1985). In acute stage, patches of RPE overlying infarcted choriod capillaries appear to be yellow and leak fluorescein. Gradually RPE becomes hyperpigmented and circled with hypopigmentation. Also the patient had positive PPD and T-spot test, additionally, her lung CT revealed secondary tuberculosis infection. We believe her age and tuberculosis infective condition put her into a hypercoagulable state and contributed to CVST formation. The underlying mechanism for exudative retinal detachment in CVST patient could be as follows: The tuberculosis infection had initiated the hypercoagulable state, then promoted CVST and choroidal infarction (supported by multiple Elschnig spots, early background hypofluorescence in FA). These thrombosis-induced infarctions destructed the outer retinal barrier (indicated by multiple pinpoint leaks in the RPE layer), which finally led to accumulation of subretinal fluid and detachment of the neuroretina and RPE(Kaur et al. 2008). Treatment for CVST including mainly anti-coagulation therapy as well as systemic supporting therapy. In this case,

we performed a combination of anti-coagulation and anti-tuberculosis therapy, the retinal lesions were relieved to a great extend and the patient complained great visual improvement.

Abbreviations

CVST: Cerebral venous sinus thrombosis

BCVA: Best corrected visual acuity

FA: Fluorescein angiography

RPE: Retinal pigment epithelium

PPD: purified protein derivative

MRV: Magnetic resonance venogram

VKH: Vogt-Koyanagi-Harada

Declaration

Ethics approval and consent to participate The study was conducted in accordance with the Declaration of Helsinki and was approved by Institutional Review Board of the Central South University. The written informed consent was obtained from the patient.

Consent to publish The written informed consent was obtained from the patient.

Availability of data and materials The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.

Competing interests The authors declared no competing interest in this study.

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Authors' Contributions The first author(LL) is responsible for data collection and the draft of the manuscript. The correspondence author (LG) is responsible for review and revision of the manuscript. All authors have read and approved the manuscript.

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Figures

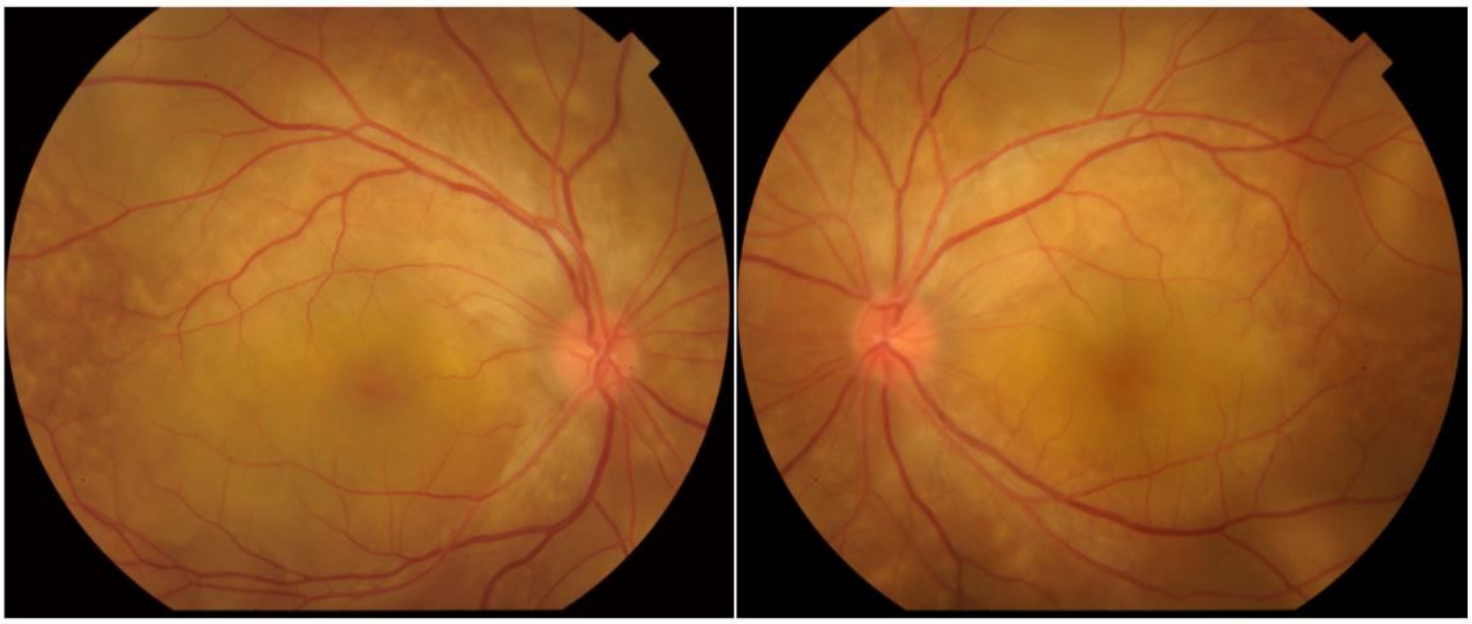

Figure 1

Digital fundus photography showed bilateral multiple retinal detachments and blurred optic disc margin, also noted the multiple polygonal yellowish-white islands in the sub-retinal areas.

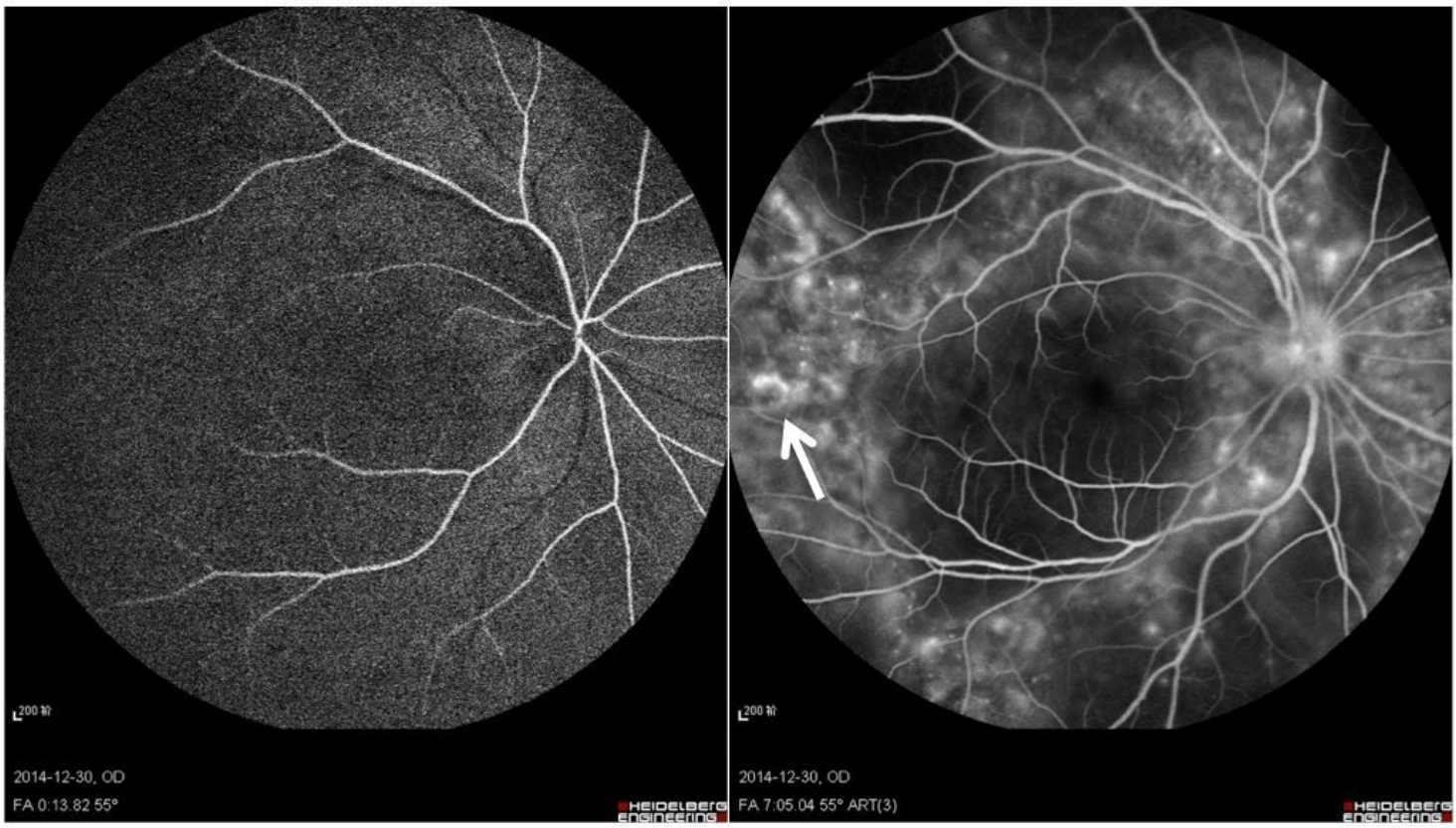

Figure 2

Fluorescein angiography (FA) revealed early hypofluorescence in the background stage, multiple pinpoint leakages at the level of retinal pigment epithelium (RPE), and late pooling to outline the boundary of retinal detachment, with some of the leakage shaped as multiple circles in the late stage of FFA (Arrow).

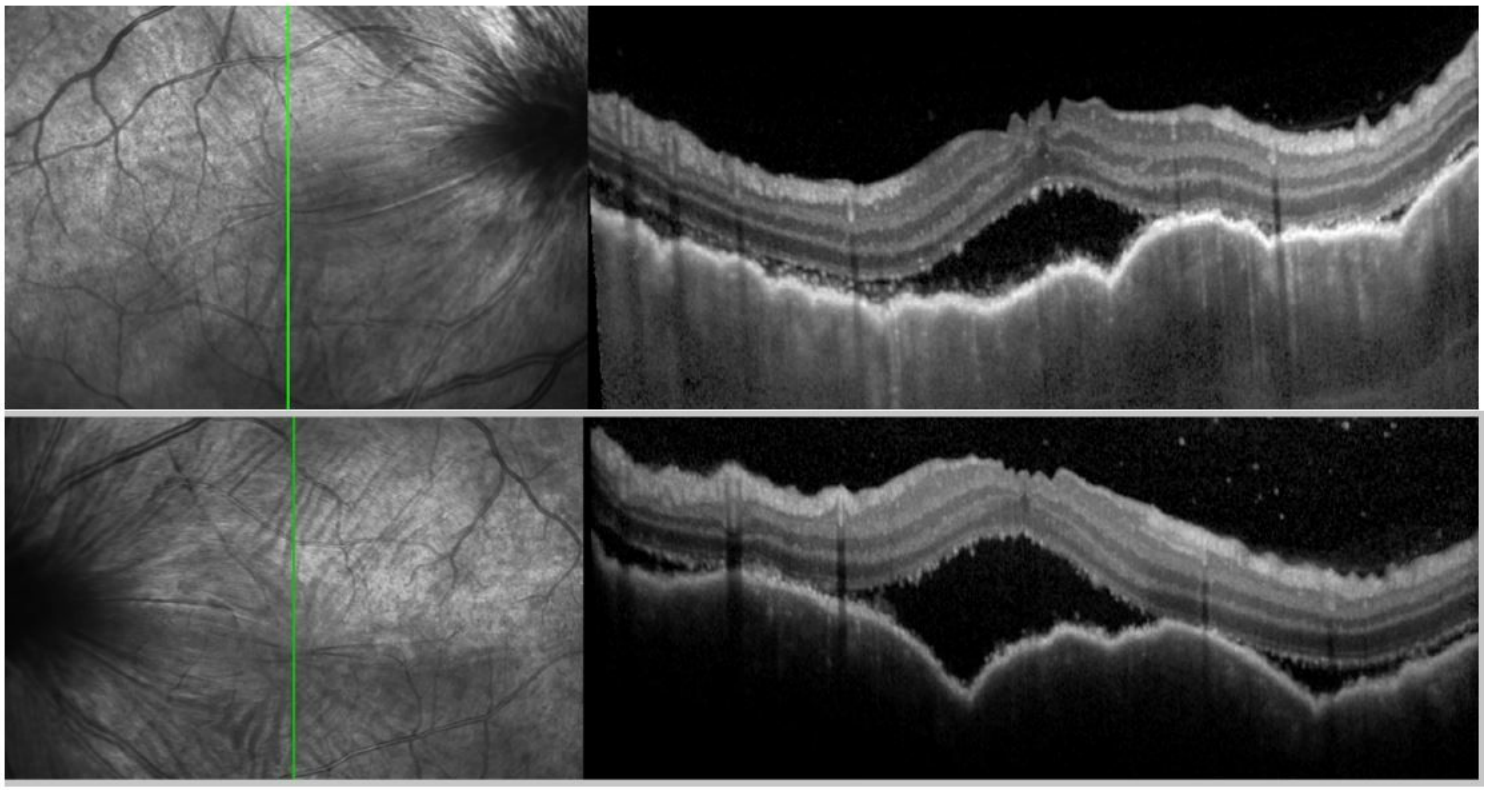

Figure 3

Optical coherence tomography revealed retinal and choroid folds, bilateral serous effusion in the sub neuro-retinal area.

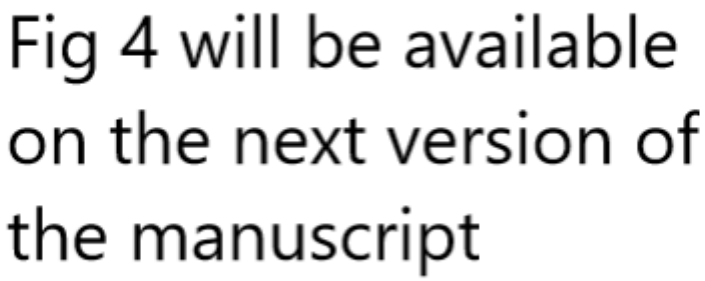

Figure 4

Head Magnetic Resonance Venogram showed thrombosis formation in the left transverse venous sinus.

Supplementary Files

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- CAREchecklistEnglish2013.pdf