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CASE REPORT

Optical coherence tomography angiography findings in Susac's syndrome: a case report

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Abstract
Purpose To report clinical features of Susac's syndrome (SS) using optical coherence tomography angiography (OCTA).
Methods Case report.
Results A 25-year-old Caucasian female with a history of non-pulsatile migraine, tinnitus, and verbal apraxia complaining of peripheral scotoma in left eye due to branch retinal arteriole occlusion (BRAO) was diagnosed as having SS after audiometric test, brain magnetic resonance imaging, and multimodal retinal imaging. Fundus biomicroscopy and fluorescein angiography (FA) revealed the presence of retinal ischemia in the area of BRAO. The OCTA images showed decreased vascular perfusion of both superficial and deep plexuses in the area of the occluded arteriole with increase of vascular density values after treatment with intravenous glucocorticoids, cyclophosphamide, and aspirin.
Conclusions OCTA offers a valid alternative to the standard invasive FA, evaluating vascular perfusion in all capillary plexuses, and monitoring retinal microvascular flow changes during the course of BRAO, without dye injection.

Keywords Susac's syndrome · Optical coherence tomography angiography · Retinal artery occlusion

Introduction

Susac's syndrome (SS), firstly described by Susac in 1979 [1], is a rare disease affecting women more than men, caused by an autoimmune microangiopathy involving the arterioles of the brain, cochlea, and retina [2]. This syndrome is characterized by the classic clinical triad of subacute encephalopathy, sensorineural hearing loss (SNHL), and visual loss secondary to branch retinal artery occlusion (BRAO) [2]. Brain magnetic resonance imaging (MRI), audiometry, and fluorescein angiography (FA) findings enable the diagnosis. Herein we report the utility of the multimodal retinal imaging approach to detect clinical features of SS, and we focus on the optical coherence tomography angiography (OCTA) as a new dyeless method for the imaging of retinal perfusion.

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Case report

A 25-year-old Caucasian female referred to our department complaining of blurred vision and peripheral scotoma in left eye (LE) 15 days after delivery (cesarean section). She also referred a non-pulsatile migraine, tinnitus, and few episodes of verbal apraxia. The medical history revealed a previous type A hepatitis, no ocular trauma or inflammation, and a postpartum hypothyroidism treated with oral tiroxin (25 mcg/daily). Best corrected visual acuity (BCVA) was 20/20 Snellen in both eyes with spherical equivalent refraction of −4.00 diopters. Intraocular pressure was 20 mmHg. Anterior segment examination was unremarkable in both eyes. Fundus biomicroscopy revealed an ischemic retinal whitening in the supero-nasal mid-periphery externally to the vascular arcade of the LE related to a branch retinal arteriole occlusion (BRAO). Posterior segment examination of the right eye was unremarkable. The young patient underwent multimodal retinal imaging including color fundus photography (CFP) using a true color confocal scanner (Eidon, Centervue, Padova, Italy), fluorescein angiography (FA) using Spectralis HRA+OCT (Heidelberg Engineering, Heidelberg, Germany), and optical coherence tomography angiography (OCTA) using XR Avanti (Angiovue, Optovue, Freemont CA, USA). The CFP confirmed a small area of retinal ischemia previously seen in fundus examination and revealed the presence of Gass plaques as yellow–white lipid deposits at the mid-segment of the retinal arterioles (Fig. 1: panel-A, magnification). FA showed mild delayed arterioles filling with retinal hypoperfusion in both supero-nasal and infero-temporal sectors in early phase (Fig. 1: panel-B, top), and typical focal arterial wall hyperfluorescences in late phase (Fig. 1: panel-B, bottom). The OCTA images showed no-flow area, as a decreased vascular perfusion, corresponding to the area of the occluded arteriole on FA in both superficial and deep vascular plexuses (Fig. 1: panel-C, top and bottom), respectively. The flow density map showed impairment in both superficial and deep vascular plexuses (Fig. 1: panel-D, top and bottom), respectively. Laboratory test workup showed: erythrocyte sedimentation rate value of 34 mm/h; C-reactive protein value of 1.49 mg/dl; increased value of both x^1–x^2 globulin (5.5% and 18%, respectively), factor V Leiden mutation (G1691A) absent,

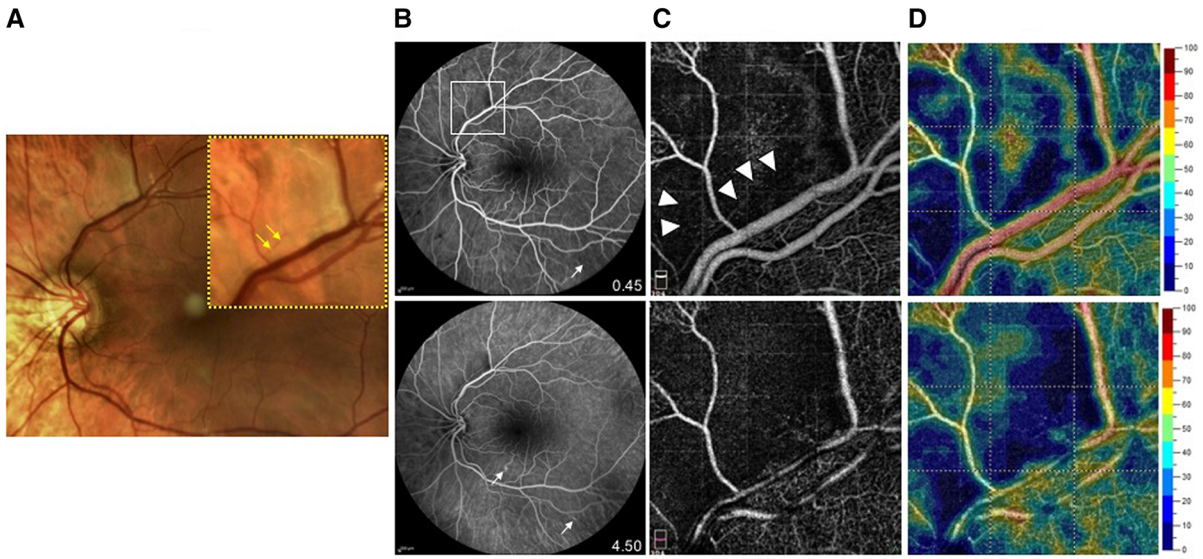

Fig. 1 Multimodal retinal imaging of 25-year-old woman affected by Susac's syndrome and showing multiple branch retinal artery occlusions (BRAOs). Small area of ischemic retinal whitening showing Gass plaques (*yellow arrows* on *dashed-square* magnification) on confocal color picture (panel-A). Fluorescein angiography (FA) showing mild delayed arterioles filling associated with retinal hypoperfusion (*white square* and *white arrow*) in early phase (panel-B, *top*). Late FA showing focal arterial wall hyperfluorescences (*white arrows* on panel-B, *bottom*). Optical coherence tomography angiography (OCTA) showing decreased vascular perfusion in both superficial (panel-C, *top*, arrowheads) and deep (panel-C, *bottom*) retinal vascular plexuses. Color-coded flow density map highlighted vessel density impairment of both superficial (panel-D, *top*) and deep (panel-D, *bottom*) vascular plexuses (the warmer the color, the greater the flow)

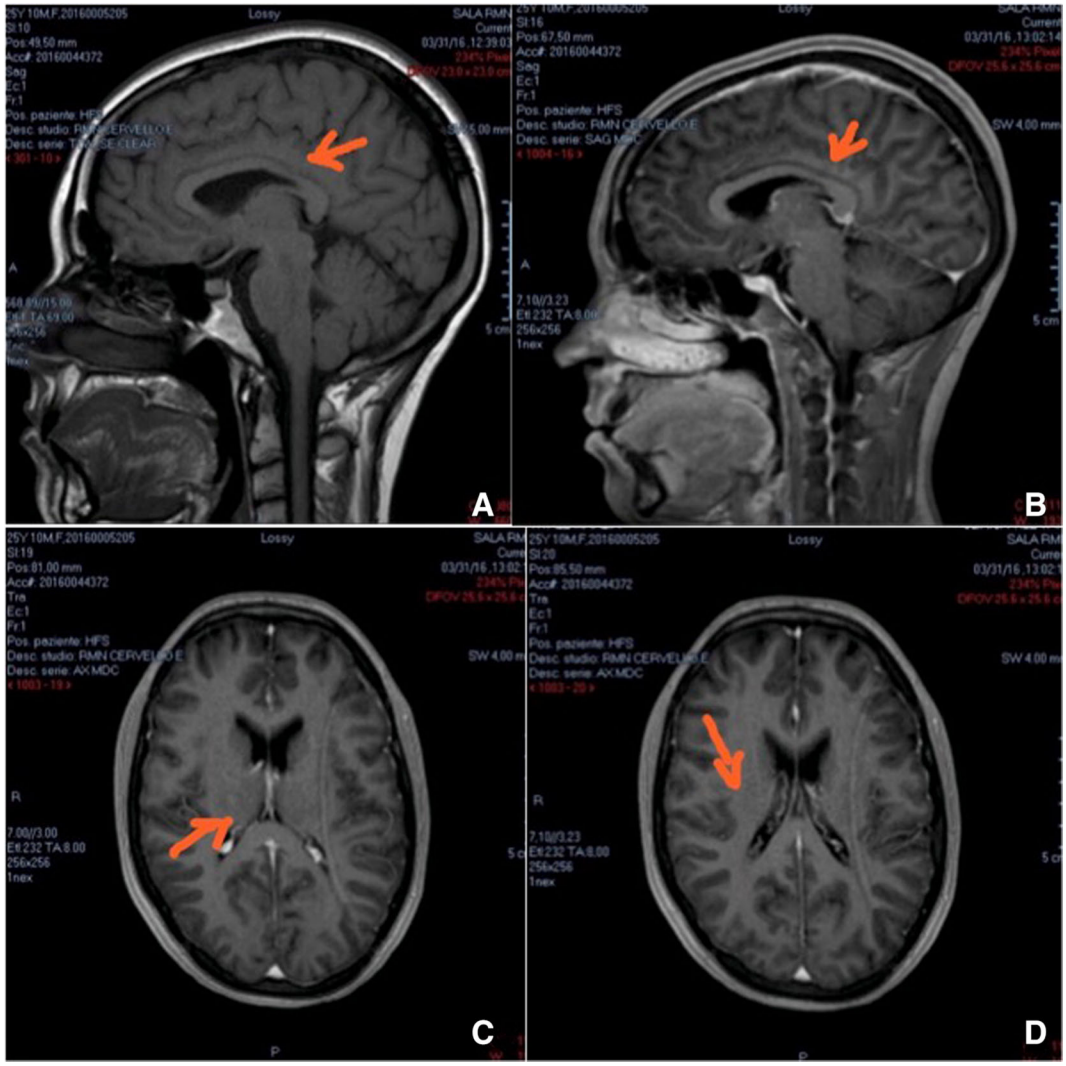

Fig. 2 **a** T1-weighted sagittal sequences: some millimetric hypointense punched-out lesions within the central part of corpus callosum, indicating severe tissue destruction. **b** The lesions do not show enhancement after contrast medium. **c** T1-weighted axial contrast-enhanced sequence showing small slightly enhancing lesion (subacute). **d** T1-weighted axial contrast-enhanced sequence showing small non enhancing lesions (not acute)

homocysteine value of 6.77 mcmol/L, normal values of protein C, S and antithrombin III, negative values of antiphospholipids antibody and negative screening for sickle cell disease. Audiometric test was performed, revealing bilateral mild-low frequency discrimination impairment due to sensorineural hearing loss (SNHL). The MRI showed pathognomonic hypointense punched-out lesions or isointense black hole in typical locations, especially within the central part of corpus callosum, indicating severe tissue destruction at T1-weighted sagittal sequences and many hyperintense little foci located in periventricular white matter, most of all in the posterior part, expression of ischemic injury with the biggest hyperintense lesion located in the trunk of corpus callosum, especially in the right side at T2-weighted axial sequences and FLAIR-coronal sequences (Figs. 2, 3, 4). All these features were suggestive for the diagnosis of Susac's syndrome (SS). High dose of intravenous glucocorticoids, cyclophosphamide, and aspirin was started early. After 3 days, the patient reported an improvement of the symptoms. The CFP images showed partial

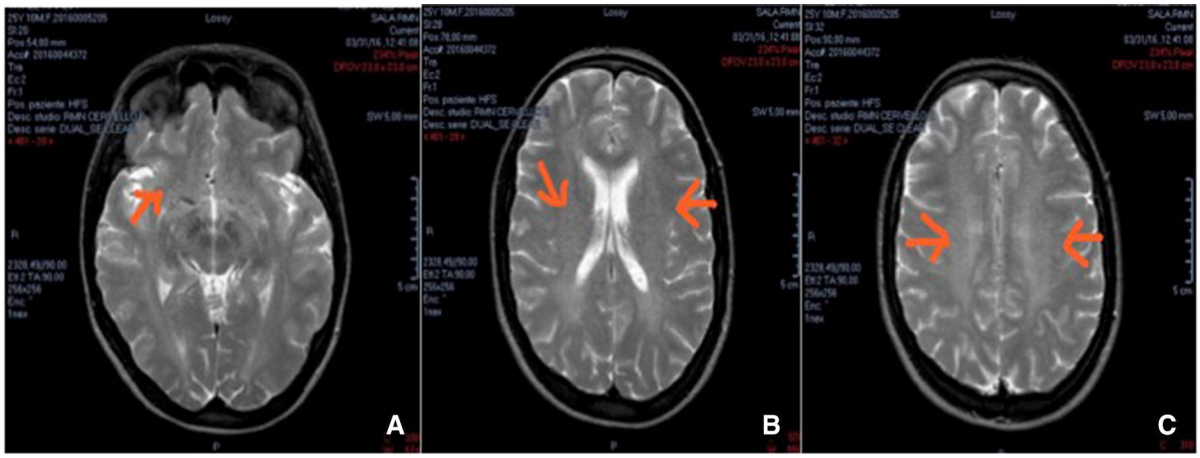

Fig. 3 T2-weighted axial sequences of insula and basal ganglia (**a**), basal ganglia and corpus callosum (**b**), and periventricular white matter (**c**): many hyperintense little foci (mainly millimetric lesions) most of all expression of ischemic injury, the biggest hyperintense lesion is located in the trunk of corpus callosum, especially in the right side, and is a sign of the inflammation process (autoimmune endotheliopathy)

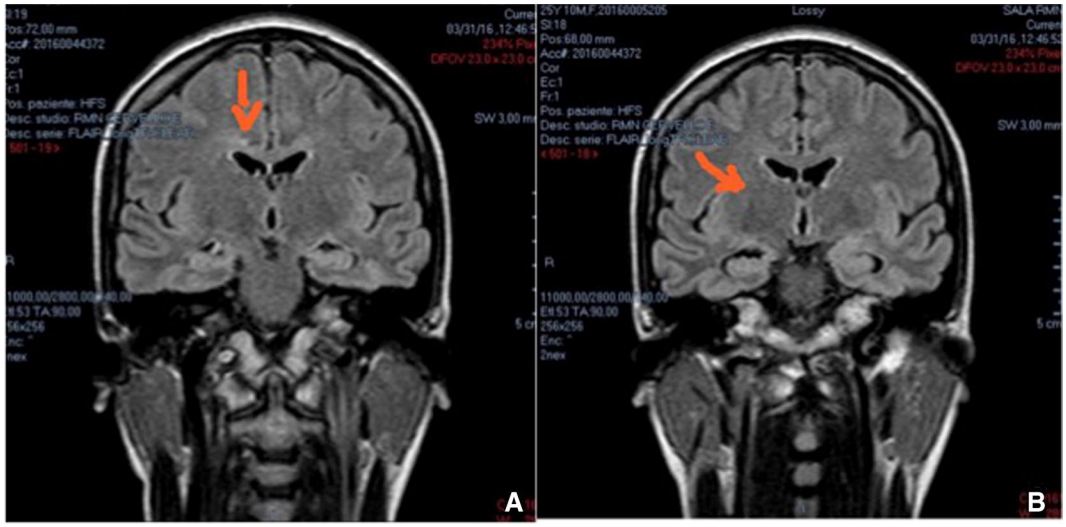

Fig. 4 FLAIR-coronal sequences showing a small hyperintense acute inflammatory lesion in corpus callosum (*right part*) (**a**), and some small spots located in basal ganglia (**b**)

recovery of retinal ischemia and the disappearance of Gass plaques (Fig. 2: panel-A, magnification). The FA images showed normal retinal arterioles perfusion in both early and late frames (Fig. 5: panel-B, top and bottom), respectively. The OCTA images showed increased vascular perfusion of both superficial and deep retinal plexuses (Fig. 2: panel-C, top and bottom), respectively. The flow density map showed improvement of both superficial and deep vascular plexuses (Fig. 2: panel-D, top and bottom), respectively.

Discussion

Susac's syndrome is a rare autoimmune microangiopathy affecting the arterioles of brain, cochlea, and retina [1, 2]. The MRI, audiometry, and FA remain the mainstay of diagnostic evaluation [2]. Agarwal et al. [3] described microstructure alterations of each individual retinal layer by means of spectral-domain optical coherence tomography (SD-OCT) in two patients with SS. Although SD-OCT imaging better analyzed retinal changes than FA, they did not show

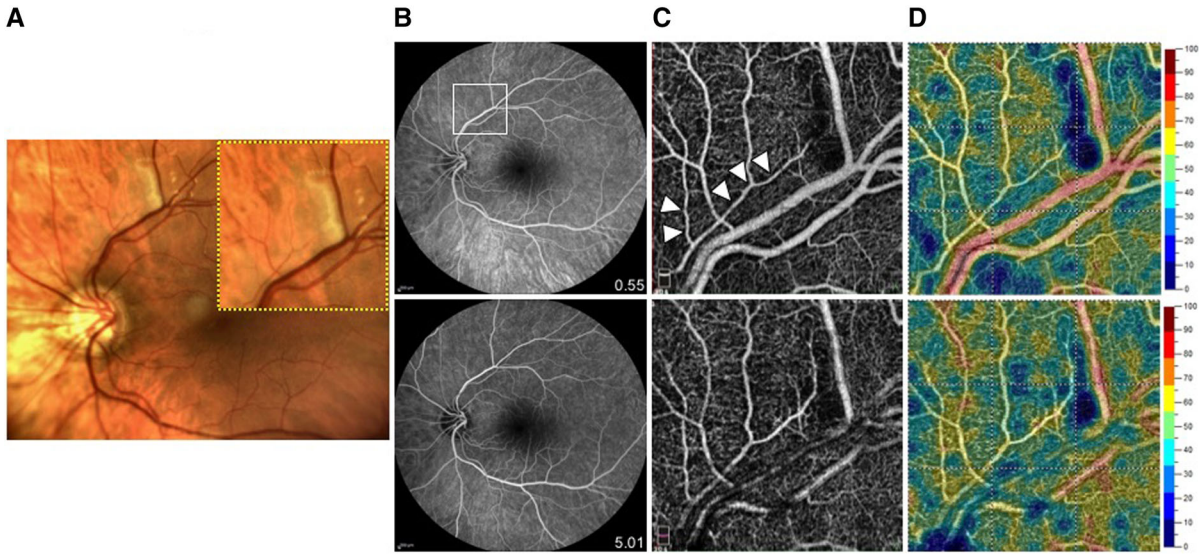

Fig. 5 Follow-up visit features on multimodal imaging of retinal healing after immunosuppressive therapy. Partial recovery of retinal ischemia and the disappearance of Gass plaques on confocal color picture (panel-A, *dashed-square* magnification). Fluorescein angiography (FA) showing normal retinal arterioles perfusion in both early (panel-B, *top*, *white square*) and late (panel-B, *bottom*) frames. Optical coherence tomography angiography (OCTA) showing increased retinal vascular perfusion in both superficial (panel-C, *top*, arrowheads) and deep (panel-C, *bottom*) vascular plexuses. Color-coded flow density map highlighted vessel density improvement of both superficial (panel-D, *top*) and deep (panel-D, *bottom*) vascular plexuses (the warmer the color, the greater the flow)

any information about vascular perfusion. Optical coherence tomography angiography is a new, fast, safe, and dyeless method able to obtain high-resolution, depth-resolved images of retinal microcirculation [4]. Our group previous reported microvascular findings in a patient affected by acute BRAO and correlating the OCTA features with those of the FA [4]. Bonini Filho et al. [5] showed decreased vascular perfusion of the superficial and deep capillary plexuses in patients affected by retinal artery occlusion, but they did not show any significative changes overtime. Herein we firstly report OCTA findings in a young female affected by SS. Particularly we highlighted the alterations of both superficial and deep retinal vascular plexuses, as a low flow area, in course of BRAO. We show a complete restoration of retinal perfusion in both superficial and deep vascular plexuses 3 days after the beginning of the immunosuppressive therapy also confirmed by FA. Susac's syndrome is an important differential diagnosis in several cerebrovascular disorders. Early diagnosis helps to start a prompt therapy, reducing relapses and improving outcomes [6]. Although FA is mandatory for the diagnosis and for the choice of treatment, the assessment and the analysis of deep capillary plexus is limited by light scattering from the inner retinal layers [7]. Optical coherence tomography angiography could offer a valid alternative to the standard invasive FA, because it allows evaluation of vascular perfusion in all capillary plexuses, and monitoring of retinal microvascular flow changes during the course of BRAO, without dye injection [4, 5]. Further prospective longitudinal studies are needed to establish the role of OCTA to monitor disease activity and to establish the efficacy of new therapeutic approaches.

Compliance with ethical standards

Conflict of interest Authors declare that they have no conflict of interest.

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Declaration of Helsinki and its later amendments or comparable ethical standards.

Informed consent Informed consent was obtained from all individual participants included in the study.

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