Case report

EJO European Journal of Ophthalmology

European Journal of Ophthalmology
1–3

DOI: 10.1177/1120672120946540
journals.sagepub.com/home/ejo
SAGE

Optical coherence tomography angiography in Tuberous sclerosis complicated with macular choroidal neovascularization

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Abstract

This study describe the optical coherence tomography angiography (OCTA) features of a young patient with Tuberous sclerosis complicated with CNV unilateral macular choroidal neovascularization during the ranibizumab therapy. OCTA scans of macular region of right eye, revealed a dense microvascular network confirming the diagnosis of CNV. After four monthly intravitreal injections, OCTA revealed a decrease of size and activity of CNV. OCTA is a valid, non-invasive, dyeless, and reliable method that could improve the diagnosis and management of CNV in child with Tuberous sclerosis.

Keywords

Tuberous sclerosis, OCT angiography, CNV

Date received: 28 May 2020; accepted: 1 July 2020

Introduction

Tuberous sclerosis or tuberous sclerosis complex (TSC) is an autosomal-dominant, neurocutaneous, multisystem disorder characterized by hamartomas that affect multiple organs, including skin, brain, heart, lungs, kidney, and eyes.[1]

Retinal hamartoma can be found in the retina and optic nerves in 30% to 50% of patients and tends to be bilateral and multiple, becoming during infancy. Therefore, the majority of hamartomas not cause visual impairment and are non-progressive.[1,2]

Here, we report, for the first time, a case of a TS patient with unilateral choroidal neovascularization evaluated with Optical Coherence Tomography Angiography (OCTA) during anti-vascular endothelial growth factor treatment.

Case presentation

A 9 year-old male suffering of incomplete form of tuberous sclerosis complex (formes frustes) presented with headache and decrease in vision in his right eye since 3 months. Visual acuity was 20/60 in the right eye and 20/20 in the left eye.

The patient had a story of visual disturbance in the right eye, did not take any medication and he had not any recent medical occurrence.

On examination, he had any findings on the biomicroscopy of the both eyes.

Slit Lamp examination showed translucent, smooth, and avascular cornea and anterior chamber was clear. The lens was clear and there was no neovascularization of the iris. Intraocular pressure was 14 mmHg by applanation tonometry.

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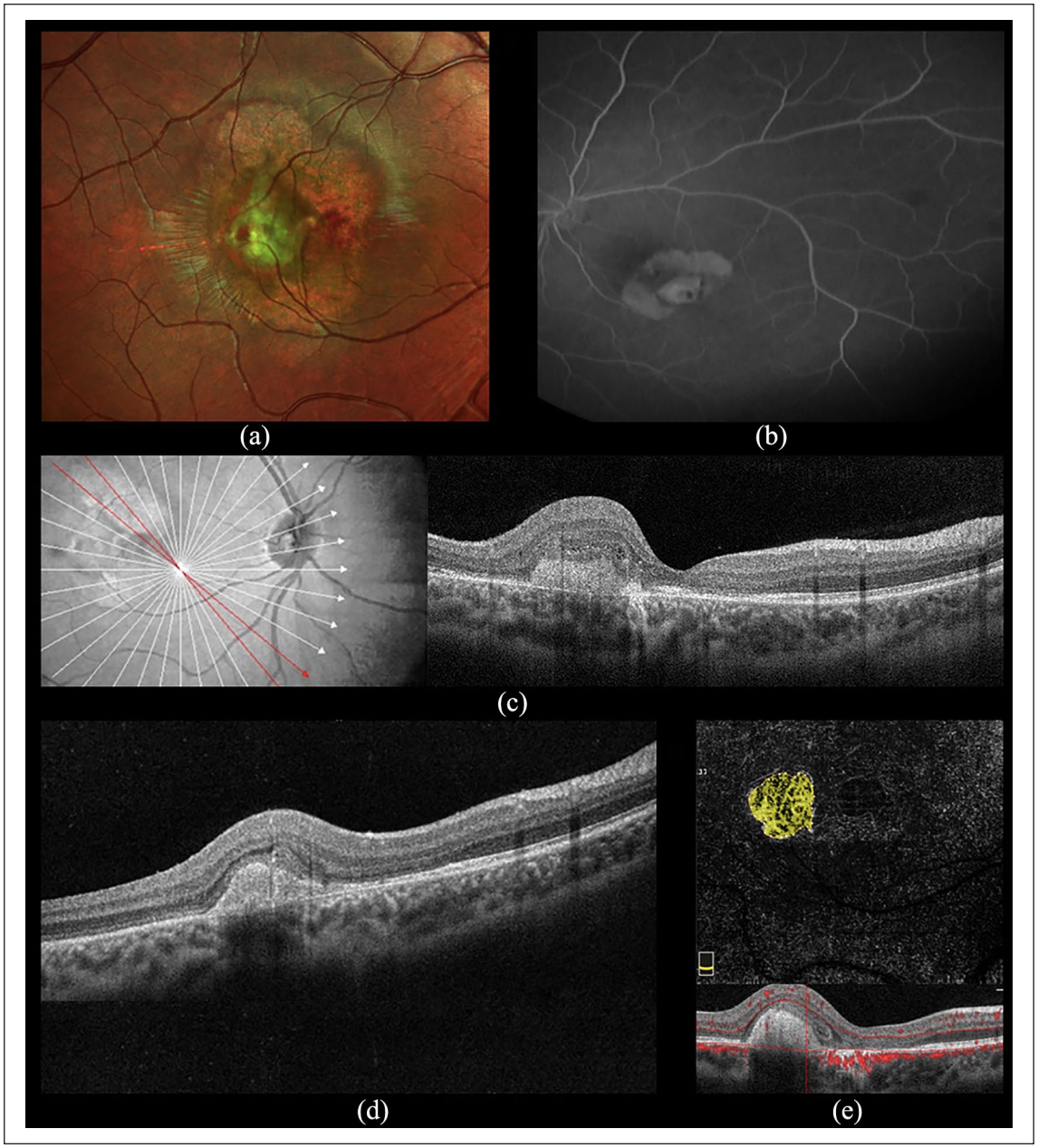

Figure 1. Right eye of a 9 year-old patient with Tuberous sclerosis complicated with macular CNV (a) color fundus examination shows large yellowish lesion with subretinal hemorrhage located in the macula region (b). Fluorescein angiography showing leakage of CNV in late phases. Structural OCT showing the typical aspect of type 2 CNV (c), with shaded margins and intraretinal fluid (d). OCTA (6 × 6) showing an interlacing vascular network at the level of the choriocapillaris segmentation (e).

Fundus examination revealed a large yellowish lesion with subretinal hemorrhage located in the macula region in the right eye (Figure 1(a)).

There were no visible calcification or retinal detachment.

Fluorescein angiography (FA) of the same eye revealed slight hyperfluorescence in the region due to choroidal neovascularization (Figure1(b)).

Structural OCT and OCTA (RTVue XR Avanti, Optovue, Inc., Freemont, California, USA) confirmed a diagnosis of CNV in his right eye (Figure 1 (c–e)), in particularly OCTA showed a interlacing network that is characterized by dense vascular hyper-intensity.

This appearance is often associated with neovascular activity on FA and structural OCT.[3]

Four monthly intravitreal injection of ranibizumab (0.5 mg) were made in his right eye.

Visual acuity improved from 20/60 to 20/30 in the right eye after Ranibizumab therapy.

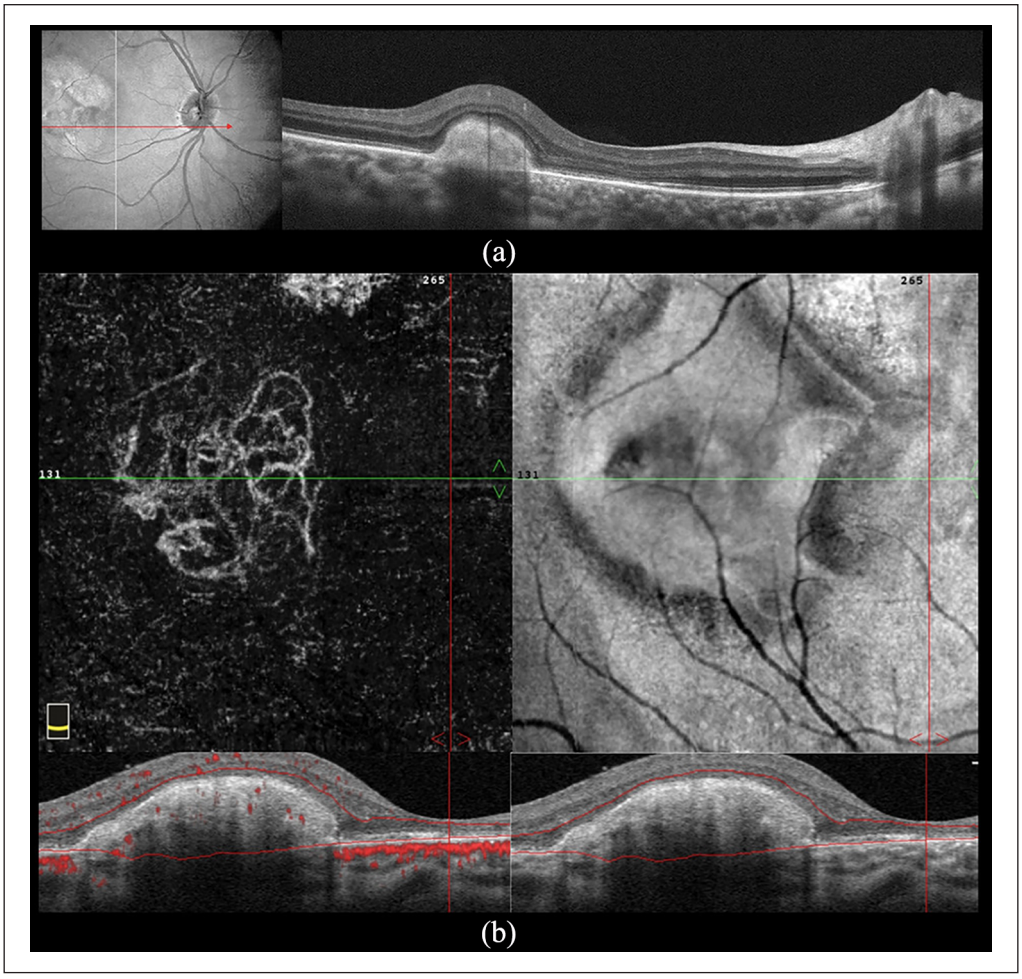

Figure 2. The same patient after anti-VEGF therapy. Structural OCT showing resolution of CNV activity, with integrity of external limiting membrane (a). OCTA (6 × 6) / en face image revealing a tangled vascular network at the choriocapillaris segmentation, with a reduction of the neovascular area (b).

Structural OCT showed the resolution of intraretinal fluid and integrity of external limiting membrane. OCTA revealed a decrease of size of CNV and the presence of tangled network with loosely laced (Figure 2 (a) and (b)). This pattern can be associated with the absence of neovascular activity.[3]

The statement of consent to publish this case was gathered from the patient.

Conclusion

OCTA is a reproducible and non-invasive examination, which provides a reliable follow-up over time as for the neovascular size area and activity in child with Tuberous sclerosis.

Declaration of conflicting interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

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