OCTA-Monitoring of Acute Posterior Multifocal Placoid Pigment Epitheliopathy (APMPPE): Transition from active to inactive disease

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Background

Acute posterior multifocal placoid pigment epitheliopathy (APMPPE), one of the white-dot-syndromes, is characterised by transient central or paracentral vision loss due to multiple yellow-white inflammatory deep retinal placoid lesions in otherwise young healthy adults. In one third of the cases, a preceding flu-like illness is present. So far, the pathogenesis of APMPPE is not completely understood, but an underlying autoimmune pathomechanism is presumed. Fluorescein angiography (FA) findings in the acute phase of the disease are an early blockage of the choroidal background fluorescence under the active lesions, and late staining. In indocyanin green angiography (ICGA) the lesions remain hypocyanescent till the late phase.
Particularly due to optical coherence tomography angiography (OCTA) analyses, recent findings indicate that the primary site of the disease is most likely the choriocapillaris, as ischemic changes herein have been observed. [1,2,3] The underlying vascular layer of Sattler's is preserved, which leads to the frequent recovery of the damaged outer retina and retinal pigment epithelium (RPE). [1,2,3]

Case report

An otherwise healthy 25-year old male was referred to our clinic with a three-day history of blurry vision in his right eye and bilateral scotomas in his visual fields (Fig. 1 and 2). Flu-like symptoms two weeks prior to presentation were reported. His best-corrected visual acuity was 20/32 in the right eye and 20/16 in the left eye. Funduscopy showed bilateral multiple creamy yellowish-white placoid lesions on the posterior pole corresponding to the perception of scotomas (Fig. 1a). His past ocular and family history of ocular and autoimmune diseases, laboratory testing and systemic investigation were unremarkable, and an infectious cause or cerebral vasculitis were ruled out. FA, ICG, fundus autofluorescence and in particular OCTA corresponded with the diagnosis of APMPPE. No treatment, but only observation was conducted. After seven weeks, the patient showed functional and anatomical improvement proven by a visual outcome of 20/20 in the right and 20/12.5 in the left eye with partial reperfusion in the choriocapillaris and recovery of the outer retinal layers on OCT and OCTA (Fig. 6).

Results: Transition from Active to Inactive Phase of the Disease

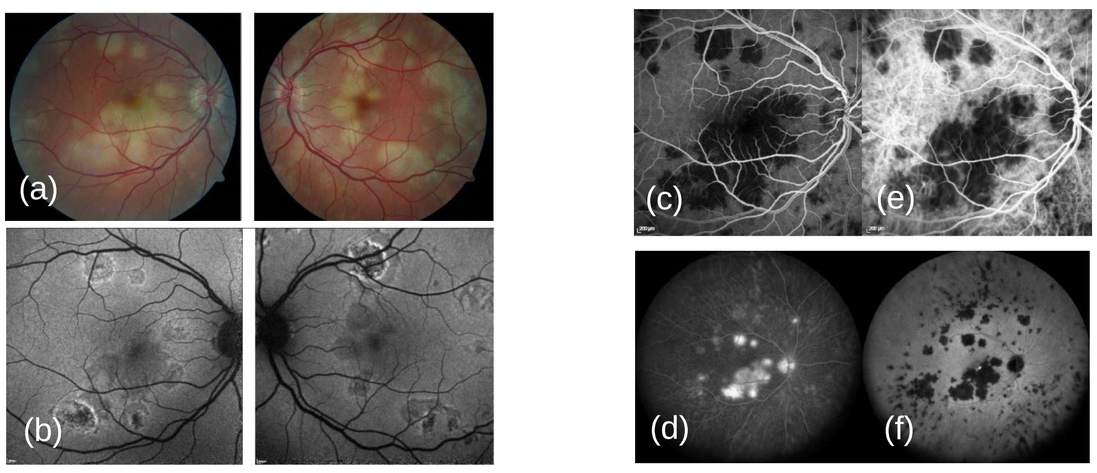


Figure 1. Day 1 (a) Bilateral multiple creamy yellowish placoid lesions on color fundus photography. (b) Bilateral fundus autofluorescence shows corresponding hypoautofluorescence with hyperautofluorescent edges. (c, d) Fluorescein angiography of the right eye with typical (c) early placoid hypofluorescence which turns hyperfluorescent in the (d) mid-late phase. Indocyanin green angiography (right eye) with placoid hypocyanecent lesions during the entire angiogram from the early (e) to the late phase (f).

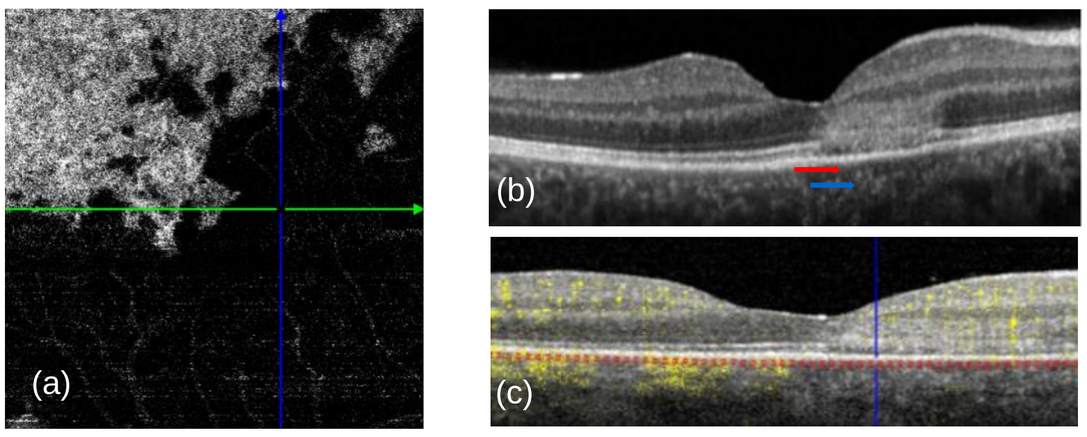


Figure 2. Day 1. (a, b) En face optical coherence tomography angiography (OCTA, a) and OCT (b) demonstrate choriocapillaris (CC) ischemia (red arrow) in the corresponding lesions of the right eye of Figure 1. (b) OCT shows a hyperreflectivity beginning in the outer plexiform layer to the retinal pigment epithelium (RPE) without CC flow (yellow) below these lesions on OCTA B-Scan (c) but with preservation of the flow in Sattler's layer (b, blue arrow).

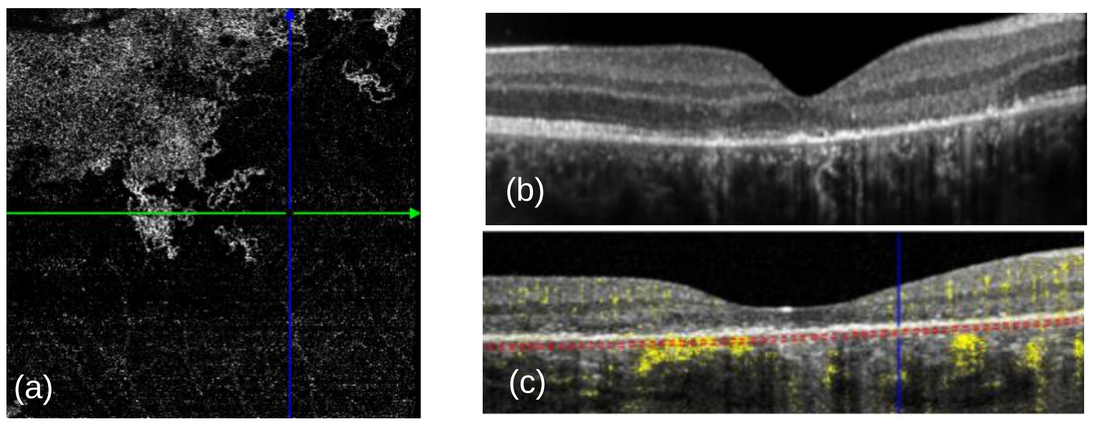


Figure 3. Day 9. (a, c) Optical coherence tomography angiography (OCTA) shows improvement of the hypoperfusion area at the level of choriocapillaris, more flow (yellow) in the OCTA compared to day one (c) and decreased hyperreflectivity and swelling of the outer retina on OCT (b).

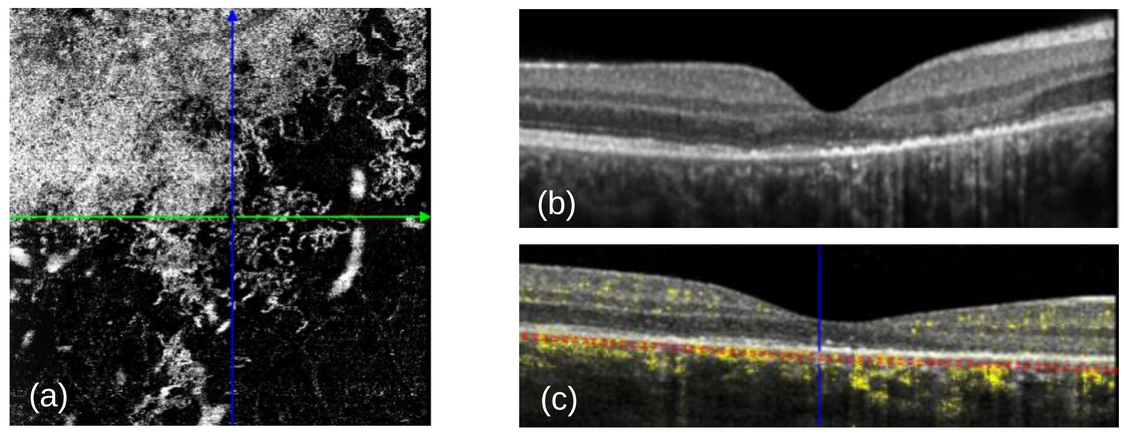


Figure 4. Day 15. (a, c) Optical coherence tomography angiography (OCTA) shows less hypoperfusion at the level of choriocapillaris. (b) OCT shows focal atrophy from the outer nuclear layer with partial recovery of the RPE-Bruch's Complex.

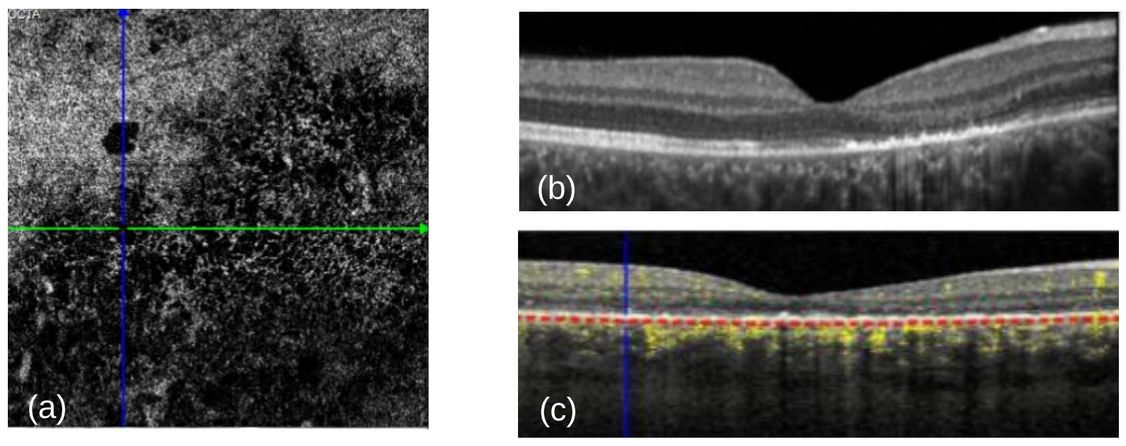


Figure 5. Day 37. (a) En face optical coherence tomography angiography (OCTA) with further reperfusion in the choriocapillaris and flow in the OCTA B-Scan (c). (b) OCT shows further thinning of the outer retina, but also further recovery, especially of the outer nuclear layer and retinal pigment epithelium.

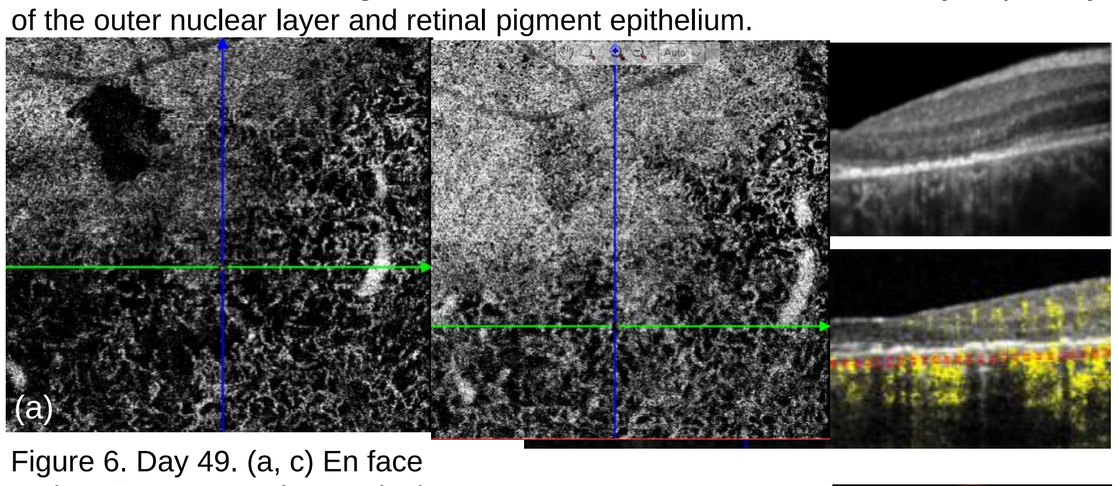
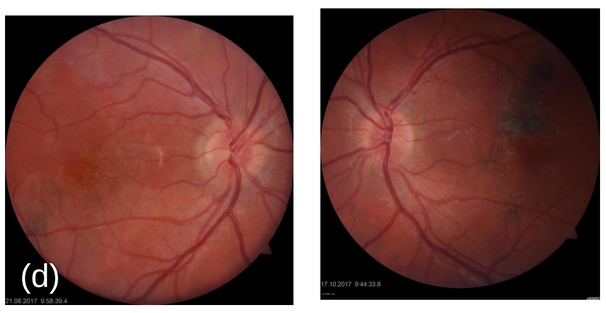


Figure 6. Day 49. (a, c) En face and B-Scan of optical coherence tomography angiography (OCTA) with clear improvement of the area of hypoperfusion at the level of choriocapillaris compared to day one (Figure 2). (b) OCT shows partial recovery in the outer retinal layers in the area of the placoid lesion in resolution (d).



Conclusion

Our APMPPE case monitored by OCTA provides further evidence of choriocapillaris non-perfusion in active lesions, but with preservation of the Sattler's layer and partial reperfusion in healed inactive lesions, which is consistent with previous studies supporting the etiopathological hypothesis of inner choroidal origin of the disease with decreased blood flow leading to ischemic changes of the overlying outer retina and RPE [1,2,3]. Moreover, we demonstrate RPE and photoreceptor-layer recovery during the transition from active to inactive disease, likely due to preservation of blood flow in the Sattler's layer. OCTA provides a deeper insight into the pathogenesis of APMPPE and is a non-invasive, readily available and promising imaging tool to diagnose and monitor APMPPE. However, it should be noted that especially with dense hyperreflectivity at the level of the outer retina or RPE in acute disease stage it might be difficult to identify the choroicapillaris flow deficiency because of low resolution and artifacts.

References
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