

Optical Coherence Tomography Findings in Relentless Placoid Chorioretinitis: Discussing a Case with Previous Reports

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Abstract

A 24-year-old man presented with blurry vision in the left eye started 1 weeks prior. At initial visit his best-corrected visual acuity (VA) was 1.0 in both eyes. Dilated fundus examination revealed hypo and hyperpigmented flat retinal lesions in both eyes. The hypopigmented lesions were multifocal, placoid and creamy-white at the level of RPE involving both the posterior pole and mid-far peripheral retina. Optical coherence tomography (OCT) scans showed disruption/defects in the outer retina and RPE hyperplasia. The laboratory work-up was negative for microbiological agents and rheumatological markers. Initial diagnosis was acute posterior multifocal placoid pigment epitheliopathy (APMPPE). Three weeks later the patient presented with newly formed far periphery lesions with a decreased VA. OCT revealed septated subretinal fluid and hyperreflective humps in the outer retina and RPE. Both OCT, fluorescein angiography (FA), fundus autofluorescence (FAF) findings and clinical features pointed out the diagnosis of relentless placoid chorioretinitis (RPC). The patient is treated with systemic and subtenon steroids and resulted with 0.2 Snellen equivalent vision. To conclude, OCT showed hyperreflectivity in the outer retina and RPE at active stage of RPC.

Keywords: Relentless Placoid Chorioretinitis; Optical Coherence Tomography; Subfoveal Fluid

Introduction

Jones and colleagues coined the term "Relentless Placoid Chorioretinitis (RPC)" after reviewing the charts of six patients demonstrating similar features to those of both acute posterior multifocal placoid pigment epitheliopathy (APMPPE) and serpiginous chorioretinitis (SC), but not typical for either of the diseases [1]. APMPPE is mostly a self-limited disease with characteristic creamy white plaques at the level of retina pigment epithelium and inner choroid, and with a benign course of 1 or 2 months. Recurrences and visual loss are not expected [2]. However, SC has a more long-term clinical course with a guarded visual prognosis when the macula is involved [3,4]. The inflammatory lesions radiate from the optic disk to the periphery in a serpentine pattern. Choroidal neovascularization may complicate the course of the disease [5].

The optical coherence tomography (OCT) has enabled us to evaluate both retinal and choroidal morphology in different types of posterior uveitis. However, there is scarce literature about the OCT findings in RPC. In the original paper by Jones and colleagues there was no OCT evaluation. In 2008 Amer&Florescu described OCT findings of active RPC as subfoveal fluid accumulation associated with pigment epithelial detachment and hyperreflectivity of both inner and outer retinal layers [6,7]. Baxter and Opremcak reported HLA-A3

and HLA-C7 association in 3 consecutive cases with a OCT finding of thickened retinal pigment epithelium (RPE) [7]. In both reports low resolution OCT images were taken. Veronese et al. described a “cockade pattern” on fundus autofluorescence (FAF) in active stage of the disease, and reported spectral domain OCT correlates with fundus autofluorescence [8]. We herein wanted to present a RPC case, and discuss our findings with the previous reports.

Case Report

A 24-year-old man presented with a 1-week history of blurry vision in the left eye. His medical history revealed that he had similar symptoms in both eyes 6 years ago, and received topical and oral corticosteroid therapy, and single intraocular Bevacizumab injection in his left eye.

At initial visit his best-corrected visual acuity (VA) was 1.0 in both eyes. Anterior segment and vitreous examination was unremarkable in both eyes. Intraocular pressure was 14 mm Hg in both eyes. Dilated fundus examination revealed pigmented chorioretinal atrophic areas in both macula and mid-peripheral retina bilaterally, which was more evident in the right eye (Figure 1). Fundus autofluorescence showed hypoautofluorescent patches more evident in the right eye. Fluorescein angiography showed hyperfluorescence of the lesions related to window defect, and hypofluorescence related to blockade by pigmentation within the lesions. OCT scans passing through the lesions showed disruption/defects in the outer retina (external limiting membrane, ellipsoid zone and interdigitation zone) and RPE hyperplasia.

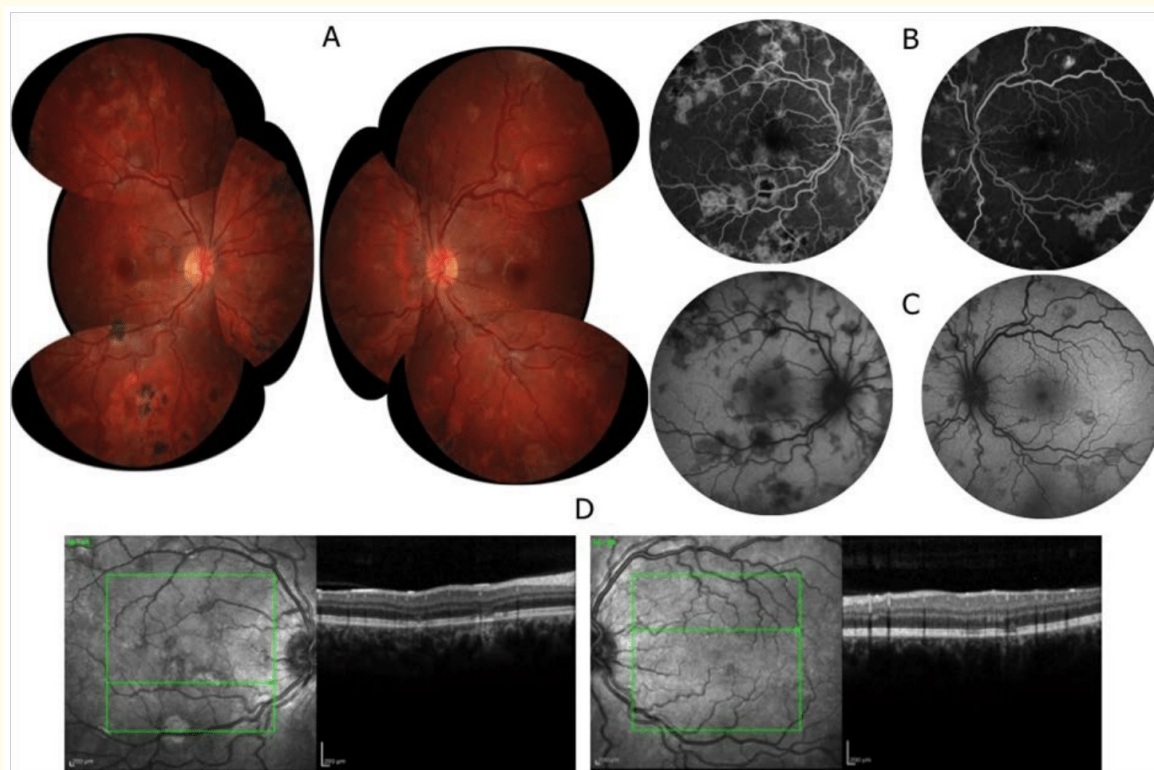


Figure 1: (A) Color pictures of both eyes showing chorioretinal atrophic lesions, some of which are pigmented (B) Fluorescein angiograms of both eyes demonstrating “window defect” hyperfluorescence and “blockade” hypofluorescence by pigment (C) Fundus autofluorescence images of both eyes showing multiple hypoautofluorescent patches (D) Optical coherence tomography scans through lesions showing outer retinal atrophic areas.

The laboratory work-up revealed normal values for white blood cells (8800/mm³), erythrocyte sedimentation rate (10mm, 1 hour) (0-15). Testing was negative for syphilis, human immunodeficiency virus, and toxoplasmosis. We performed QuantiFERON-TB Gold (QFT®) test to exclude *Mycobacterium tuberculosis*, and the result was negative. Rheumatoid factor, antinuclear antibodies were not detected. The angiotensin converting enzyme level and chest X-ray were normal. Pathergy testing was negative for Behcet’s disease. Rheumatology consultation found no clue of systemic lupus erythematosus, sarcoidosis, or other auto-immune diseases. Neurology consultation with magnetic resonance imaging of the brain to identify central nervous system vasculitis was unrevealing. At this stage we diagnosed the patient as APMPE, and scheduled for 1-month visit without any treatment. Three weeks later, he presented with VA deteriorated to counting fingers at 1 meter in the left eye. Fundoscopy revealed creamy-white new retinal lesions between vascular arcades in the left eye, however, fundoscopic appearance and other findings were unchanged in the right eye.

In the left eye FAF revealed new hypoautofluorescent spots with hyperautofluorescent borders, which were not present at the initial visit. FA displayed early hypofluorescence and late staining of the new lesions. There was also leakage from retinal venules and optic disk. OCT scan through the fovea revealed some intraretinal fluid, septated subretinal fluid and RPE undulations. Moreover, OCT scans passing through the active lesions showed hyperreflectivity in the outer retina and RPE. Indocyanine Green Angiography (ICG) showed typical hypoautofluorescence in both early and late frames (Figure 2). We revised our diagnosis as RPC, and initiated 1 mg/kg/day oral prednisolone acetate therapy. We also performed sub-Tenon’s 40 mg methylprednisolone and 3 mg betamethasone injection for macular edema in the left eye. Two weeks later fundus lesions appeared to be flattened, more grayish in color and confluent on color picture. VA was improved to 0.2. Macular edema was resolved and there was disruptions in the outer retina and RPE hyperplasia on OCT.

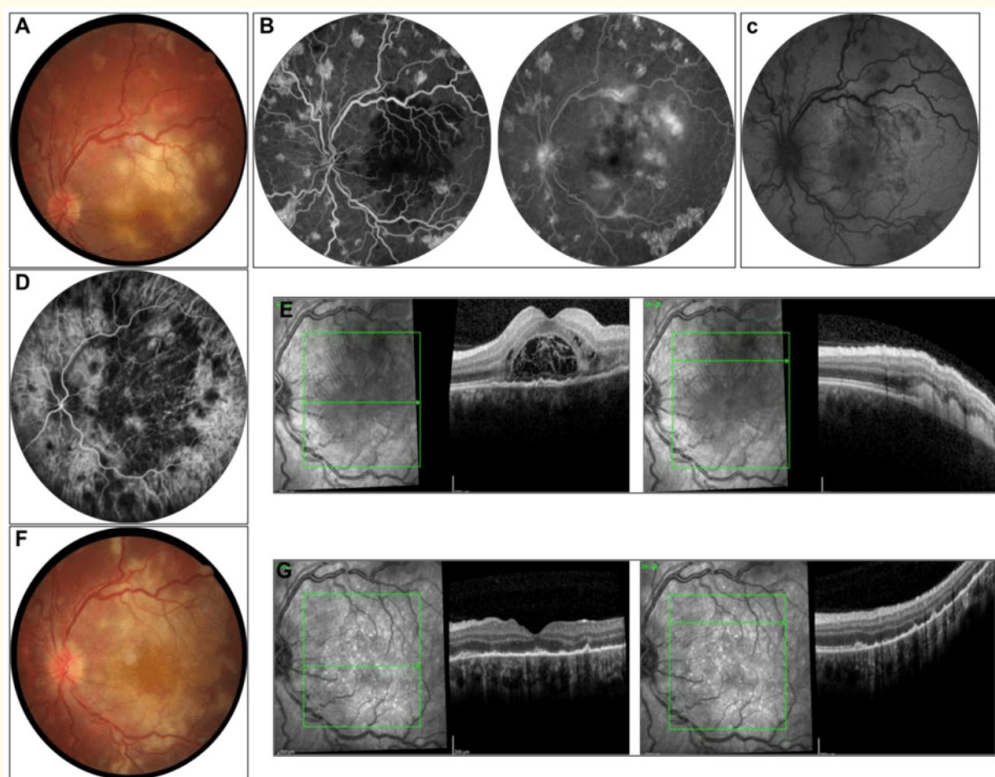


Figure 2: (A) Color picture of the left eye showing new creamy-white retinal lesions in the macula in addition to old faint lesions outside upper vascular arcade (B) Fluorescein angiography showed early hypofluorescence and late hyperfluorescence of the new lesions, hyperfluorescence of the optic disk and perivascular leakage were also observed (C) Fundus autofluorescence demonstrated new hypoautofluorescent patches with hyperautofluorescent borders in the left macula (D) Indocyanine green angiography revealed hypofluorescence of the lesions during testing (E) Optical coherence tomography scan through fovea showing septated subretinal fluid associated with some intraretinal fluid and RPE undulations (left), another scan through an active lesion showing hyperreflectivity in the outer retina and RPE (right) (F) Color picture taken two weeks later showing active lesions turned to be more grayish in color (G) Optical coherence tomography scans showing dry macula but outer retinal defects and RPE hyperplasia.

Discussion

Our case has similar features to APMPE and serpiginous choroiditis in that the creamy-white retinal lesions showed early hypofluorescence and late hyperfluorescence on FA [2,4]. Furthermore, the placoid shape of the retinal lesions was similar to that seen in APMPE, while the longer clinical course and being active at a time reminds SC. However, the present case and those described by Jones differed from typical APMPE or SC, because the retinal lesions were extremely numerous, developed in the mid- and far periphery, and also did not follow a helicoid pattern. Therefore, we believe that our case is consistent with the description given for RPC. Due to its rarity there is no standard therapy for RPC. Steroids and other immunosuppressive agents have been tried with varying results [1,9]. We also tried sub-Tenon's steroid injection for macular edema in the current case, which seemed to dry the macula effectively. However, we are not sure if this is the pure effect of sub-Tenon's steroid injection.

OCT imaging studies are quite scarce in RPC. The most detailed one by Veronese et al presented FAF correlates with OCT findings at different stages of the disease [8]. In parallel to their findings we observed atrophic changes supported by both OCT and FAF in the convalescence phase of the disease. However, our findings at active stage contrast with theirs. In their original description of cockade pattern on FAF they observed three concentric zones of different autofluorescence in the vicinity of active lesion as more hypo in the center, hyper pericentrically and less hypo in the periphery. Corresponding OCT scans revealed subretinal fluid and outer retinal defect in the center. Contrary to this we observed a central hypoautofluorescence surrounded by hyperautofluorescent border rather than three distinct zones of autofluorescence. OCT showed retinal thickening with hyperreflectivity in the outer retina and RPE. We think that at active stage of the disease such an early atrophy of the photoreceptors is not highly expectable. Supporting other reports of OCT findings in RPC we observed transretinal or outer retinal hyperreflectivity during the active stage [6,7]. This was also shown in other uveitic entities such as APMPE, multifocal choroiditis and SC [10-12]. Veronese et al did not give a color picture for the active stage we were not able to compare fundoscopic appearances between cases [8]. Similar to their case we observed atrophic changes in the outer retina after 2 weeks. A study reported choroidal sparing in RPC, however, we did not investigate this in the current case [13]. Interestingly we observed septated subfoveal fluid which was described in Vogt-Koyanagi-Harada disease and neovascular age related macular degeneration treated with photodynamic therapy. The septations were likely fibrin, and the subretinal hyper-reflective material could be inflammatory cells and debris. Recently Stronbehn and Sohn reported a similar appearance in a case with choroidal metastasis of lung cancer [14]. Subretinal fluid accumulation is a well-known finding for APMPE, SC and RPC as well. However, we could not find such an appearance for RPC during literature search.

Conclusion

In conclusion, at active stage of RPC OCT showed hyperreflectivity in the outer retina and RPE. There then ensue the photoreceptor atrophy and RPE hyperplasia during the convalescence phase of the disease. Associated sub-foveal fluid may have a "septated" appearance.

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