DOI: 10.5603/OJ.2023.0014

Acute retinal pigment epithelitis in a 11-year-old child: case presentation and follow-up using optical coherence tomography angiography and static visual field

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ABSTRACT

We present a case of unilateral self-limiting acute retinal pigment epithelitis (ARPE) in an 11-year-old girl and 10 weeks of follow-up until complete recovery using spectral domain optical coherence tomography (SD-OCT), SD-OCT-angiography (SD-OCTA) and static visual field.

KEY WORDS: acute retinal pigment epithelitis; ERPE; acute retinal pigment epitheliitis; youngest

Ophthalmol J 2023; Vol. 8, 46–51

INTRODUCTION

Acute retinal pigment epithelitis (ARPE) was documented for the first time in 1972 by Krill and Deutman [1].

It is a rare, self-limiting idiopathic disease of the central retina. It typically manifests in young adults who are generally healthy [1].

The visual prognosis is good, and treatment is not required. Patients usually present with unilateral painless blurred vision, central/paracentral metamorphopsia, or scotoma. Examination of the fundus shows fine pigment stippling in the macula, bordered by areas of reduced pigmentation [1, 2]. ARPE occurs mostly unilaterally, but bilateral cases have been reported [3]. Electroretinogram studies are normal, whereas electrooculogram studies are subnormal, suggesting that RPE is the primary site of involvement in ARPE [4].

The most common spectral domain optical coherence tomography (SD-OCT) finding is disruption of the retinal pigment epithelium (RPE) inner layer (78%) and abnormal reflectivity of outer retinal layers [2]. In the literature review Raşit Kılıç reported that the interdigitation zone (IZ) was affected in all cases, the ellipsoid zone (EZ) in 95.6% of cases, the external limiting membrane (ELM) in 35.6% of cases, and the outer nuclear layer (ONL) in 26.7% of cases [5].

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CASE PRESENTATION

The patient, an 11-year-old Caucasian female, was examined due to a recent onset of central vision blurring in her left eye lasting for 4 days. She had no known ocular or medical disorders and denied any flu-like symptoms in the recent past.

COVID-19 history was unknown. Best-corrected visual acuity (BCVA) in the affected left eye was 9/20, whereas that in the right eye was 20/20. There were no notable findings in the intraocular pressure and anterior segment. However, an examination of the left eye fundus revealed the presence of focal pigmentary disturbance in the macula (Fig. 1). The right eye macula was unremarkable. Central blurring was observed in the left eye during the Amsler grid examination.

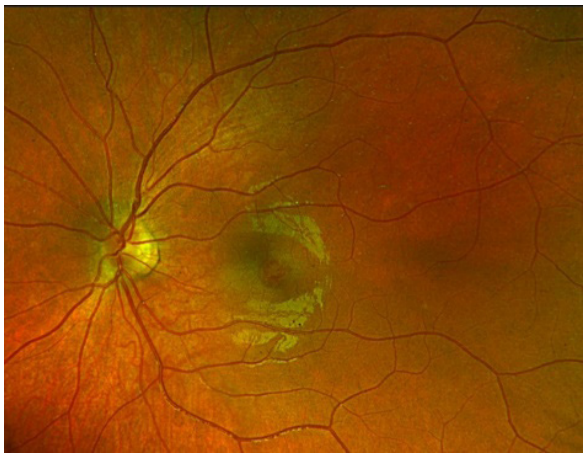

FIGURE 1. Fundus photography of the left eye

Spectral-domain optical coherence tomography (AvantiTM Widefield OCT) of the affected eye showed a subfoveal hyper-reflective lesion in the outer neurosensory retina with disruption of the ellipsoid zone (EZ) and external limiting membrane (ELM). The retinal pigment epithelium (RPE) below the hyper-reflective lesion was thickened. There were no cysts within the retina or fluid underneath it.

The outer nuclear layer (ONL) contained hyper-reflective lesions. Central retinal thickness was 258 um *vs.* 241 in the fellow eye. Central superficial vessel density in OCT-angiography (AngioVueTM OCTA Angiography) was 25% higher than in the second eye (Fig. 2). Fundus autofluorescence showed no difference between the eye.

Central visual field static 10-2 threshold test reveals central scotoma (pattern deviation 4 points < 5% and 4 points < 2%) in the left eye (Fig. 3).

We considered retrobulbar optic nerve inflammation, trauma, laser-induced ocular injury, or even amblyopia as the differential diagnosis.

Toxoplasmosis antibodies blood test was negative. Inflammatory blood parameters were normal. The patient recognized colors and did not report retrobulbar pain or any neurological symptoms. Her family history was negative. We diagnosed her with ARPE and observed her without treatment. 12 days after presentation BCVA was 14/20 and 15 days later achieved 20/20.

Ten weeks from the beginning, the hyper-reflective lesion on OCT resolved. The displaced ELM

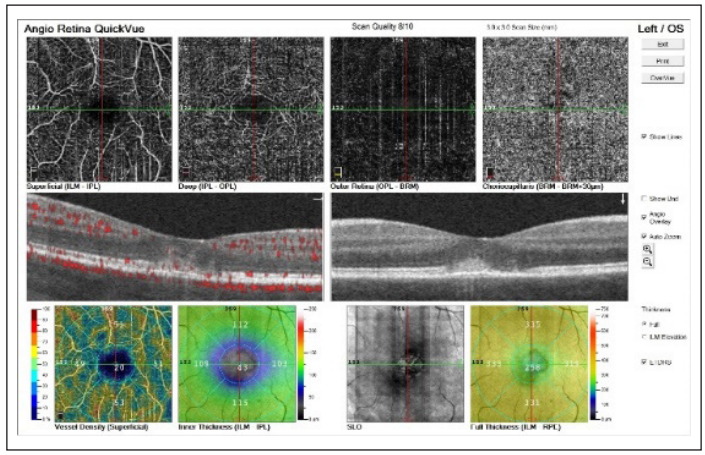

FIGURE 2. Spectral-domain optical coherence tomography (AvantiTM Widefield OCT) of the affected eye showed a subfoveal hyper-reflective lesion in the outer neurosensory retina. The ellipsoid zone (EZ) and external limiting membrane (ELM) was interrupted. The retinal pigment epithelium (RPE) below the hyper-reflective lesion was thickened

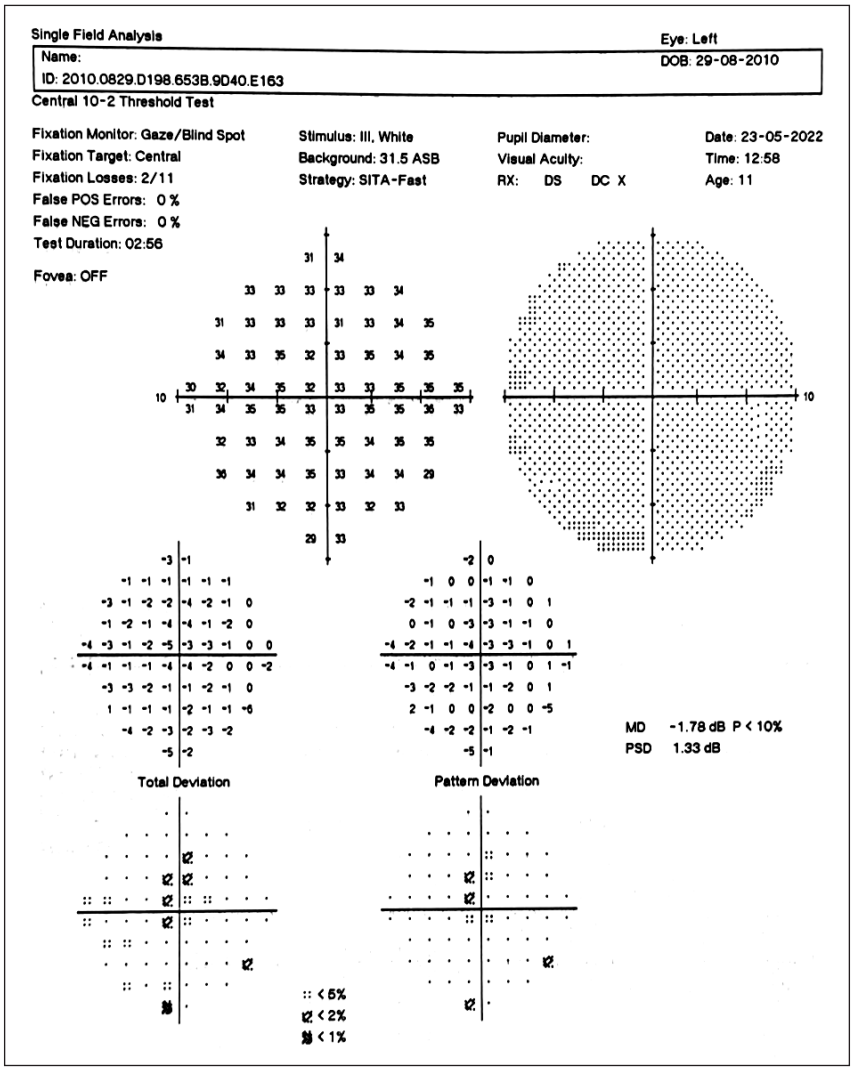

FIGURE 3. Central visual field static 10-2 threshold test of the left eye reveal central scotoma (pattern deviation 4 points < 5% and 4 points < 2%)

was restored. Central retinal thickness was 240 um *vs*. 244 in the second eye. Central superficial vessel density in OCT-Angiography was 7% lower than in the the second eye (Fig. 4).

Central visual field static 10-2 Threshold test of the left eye revealed paracentral scotoma (pattern deviation 3 points < 5%, 2 points < 2%, and 1 point < 1% (Fig. 5).

Fundus findings mostly disappeared (Fig. 6).

DISCUSSION

To the best of our knowledge, this is the youngest patient described with an acute phase of ARPE. ARPE is an acute, self-limiting disease of unknown etiology. The pathogenesis and etiology of the disease remain unclear [1, 6].

Certain studies have suggested that there could be a correlation between a viral infection and ARPE. Raşit Kılıç found viral infection in 25.9% of cases [5]. Our patient did not present general symptoms. The laboratory test results were normal. The patient did not report any viral infection in the recent past, but many viral infections can be asymptomatic. Tests for all possible pathogens are not widely available.

In the differential diagnosis, other retinal diseases should be considered, such as white dot syndromes, including acute posterior multifocal placoid pigment epitheliopathy (APMPPE), birdshot chorioretinopathy, serpiginous choroiditis and multiple evanescent white dot syndrome (MEWDS), which can present with lightly colored lesions within the macular deep retina and RPE. While the clin-

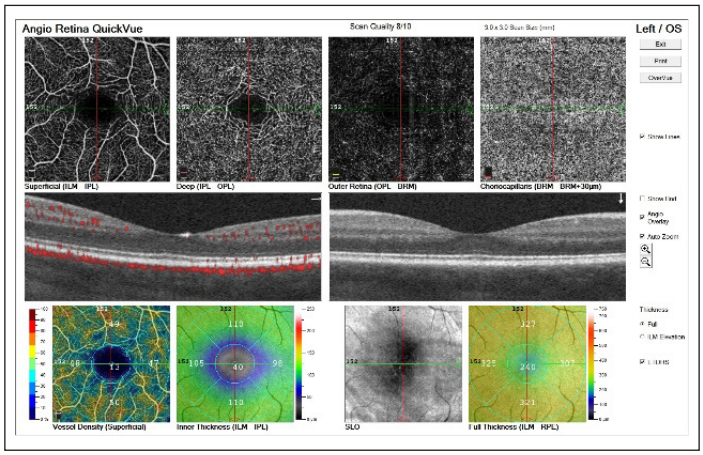

FIGURE 4. Spectral-domain optical coherence tomography (AvantiTM Widefield OCT) of the affected eye after 10 weeks of observation showed almost complete restoration of displaced external limiting membrane (ELM), a reduction of the central retinal thickness, and central superficial vessel density

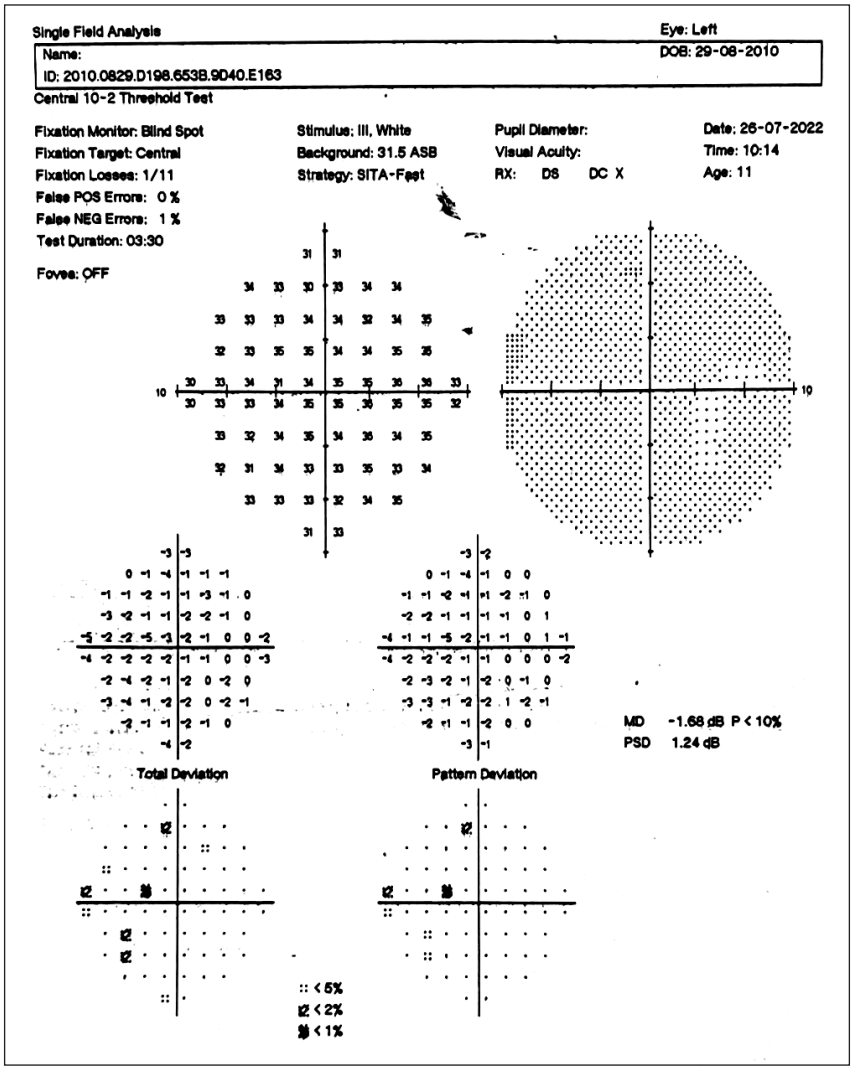

FIGURE 5. Central visual field static 10-2 threshold test of the left eye after 10 weeks of observation

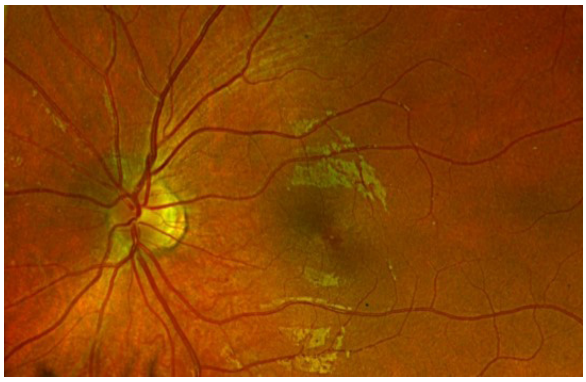

FIGURE 6. Fundus photography of the left eye after 10 weeks of observation showed almost regular fovea.

ical course and prognosis can be different, they can be very similar in the early stages [7]. The differences are sometimes discrete, like choroidal thickening, perifoveal dots, or very mild vitritis associated with MEWDS [8].

It is essential to inquire about using psychoactive substances, such as poppers, which can be linked with disruption of the foveal cone outer segments [9].

The pathophysiology of ARPE is still not fully understood. Hyper-reflective lesions seen on OCT in the outer neurosensory retina in ARPE suggest transient dysfunction or inflammation at the interface between the photoreceptor outer segments and the apical side of the RPE cells [2, 10].

Analogous morphological alterations can be observed in animal models with specific genetic photoreceptors outer segments (POS) phagocytosis deficiency. The undigested POS may accumulate at the fovea.

It is believed that an acute and transitory phagocytosis dysfunction of RPE may be responsible for ARPE symptoms. The temporary nature of this disease suggests that irreversible alterations do not occur in photoreceptors, which is associated with good recovery [6]. According to Puche et al. reports, there may be a connection between the pathogenesis of ARPE and Mer tyrosine kinase (MerTK) deficiency [6]. MerTK functions as a negative regulator of $\alpha v\beta 5$ integrin-dependent POS binding. An insufficiency of MerTK leads to an excess POS binding by RPE cells [11]. Animal models with insufficient MerTK expression display rapid and total retina degeneration. In humans, some MerTK mutations lead to retinitis pigmentosa [12].

Because of the inflammatory nature of ARPE, when an active case is encountered, treatment is sometimes initiated to prevent further vision loss. A short course of systemic non-steroidal anti-inflammatory drugs (NSAIDs) or corticosteroids has been shown in the literature to shorten visual recovery in patients with the acute phase of ARPE. In actual practice, therapy is often not implemented considering the short, self-limiting course of the disease, relatively good prognosis, and possible side effects of systemic therapy [13]. Our experience has shown that recovery is relatively fast, and no treatment is necessary. According to some other observations, 89% of affected eyes have complete recovery of visual acuity within 2 months with no treatment [2].

ARPE typically affects healthy young adults [1]. The prevalence of ARPE in the child population is still unknown. Cases of patients under 18-year-old are sporadic. Raşit Kılıç, in the literature review, found 61 patients with 67 involved eyes. The mean patient age was 30,6 years. The youngest patient was 16-year-old and the oldest 55-year-old [5].

Our 11-year-old patient can be the youngest described individual with an acute phase of ARPE with perspectives for further very long follow-up.

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