Case Report

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Optic coherence tomography angiography follow-up in a case of Purtscher-like retinopathy due to atypical hemolytic uremic syndrome

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Abstract

Purpose: To report a case of Purtscher-like retinopathy due to atypical hemolytic uremic syndrome and the changes seen in the optical coherence tomography angiography before and after treatment with eculizumab.

Case description: A 22-year-old man with an unremarkable medical history presented with acute, bilateral blurred vision and headache of 1-week duration. Best corrected visual acuity of 20/50 and 20/40, respectively, in the patient's right eye and left eye. Funduscopy revealed multiple cotton-wool spots associated with intrarretinal fluid. Swept source optical coherence tomography revealed multifocal retinal detachments with increased choroidal thickness. Optical coherence tomography angiography showed areas of ischemia in both capillary plexus. Due to concurrent symptoms and laboratory analysis, he was diagnosed with atypical hemolytic uremic syndrome and secondary Purtscher-like retinopathy; therefore, treatment with eculizumab was initiated. After 2 months revascularization of the previous ischemic areas was seen in the optical coherence tomography angiography that were correlated with best corrected visual acuity improvement.

Conclusion: Our findings suggest that evaluation of the macular capillary plexus revascularization by optical coherence tomography angiography during the disease could help to predict an improvement of best corrected visual acuity in these patients and the measurement of choroidal thickness could give us information about the resolution of the pathologic process.

Keywords

Optic coherence tomography angiography, Purtscher-like retinopathy, eculizumab

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Introduction

Purtscher's retinopathy is characterized by acute visual loss and retinal findings such as cotton-wool exudates, intraretinal hemorrhages, retinal whitening, and serous retinal detachment following head or chest trauma. When the etiology is not trauma related, it is called Purtscher-like retinopathy. Associated conditions include connective tissue and autoimmune disorders, thrombotic microangiopathic diseases such as hemolytic uremic syndrome (HUS), disseminated intravascular coagulation (DIC), thrombotic thrombocytopenic purpura (TTP), and HELLP (hemolysis, elevated liver enzymes, and low platelet count) syndrome.[1–4]

Atypical hemolytic uremic syndrome (aHUS) is a rare and potentially life-threatening entity characterized by microangiopathic hemolytic anemia, thrombocytopenia, and acute kidney failure. This syndrome is differentiated from HUS by the absence of diarrhea and Shiga toxin–induced infection.[1,2] The pathological action of aHUS is

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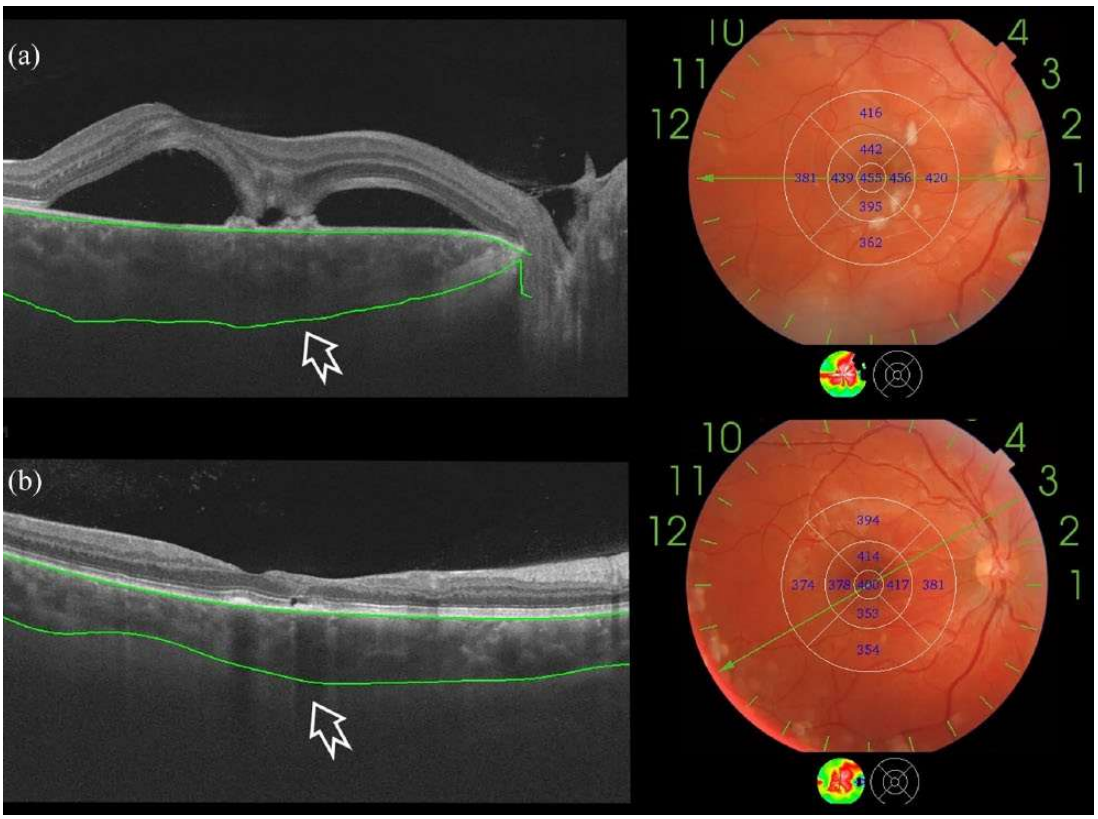

Figure 1. Color fundus photographs and swept source optic coherence tomography (SS-OCT) images of the right eye (Figure 1) and left eye (Figure 2) at initial presentation (a) and 4 months after initiating eculizumab treatment (b). Subfoveal choroidal thickness pointed with arrow and measured on fundus photographs (455 and 400 µm in the right eye; 355 and 397 µm in the left eye).

the dysregulation of the complement system leading to endothelial damage and complement aggregates. Several genetic anomalies have been associated with this syndrome, including mutations in complement factor H (CFH) and complement factor I (CFI), membrane cofactor protein (MCP), and factor B.[1–3]

Here we present an unusual case of a patient who developed Purtscher-like retinopathy due to aHUS. In this case, optical coherence tomography angiography (OCTA) was used to assess changes in the superficial capillary plexus (SCP) and deep capillary plexus (DCP) throughout the entire disease process. To our knowledge, this is the first such case reported to date.

Case report

A 22-year-old man with an unremarkable medical history presented with acute, bilateral blurred vision, dizziness, and headache of 1-week duration. The ophthalmoscopic examination revealed best corrected visual acuity (BCVA) of 20/50 and 20/40, respectively, in the patient's right eye (RE) and left eye (LE). Anterior segment examination was unremarkable.

Funduscopy revealed multiple cotton-wool spots associated with intrarretinal fluid. Given these findings, we performed fluorescein angiography (FA) and swept source optical coherence tomography (SS-OCT) with OCTA. FA showed bilateral peripapillary hyperfluorescent spots. SS-OCT revealed multifocal serous retinal detachment (SRD) in both eyes associated with increased choroidal thickness, the last being more noticeable in the RE (Figures 1 and 2). OCTA showed areas of perifoveal ischemia in both capillary plexus with a secondary enlargement of the foveal avascular zone (FAZ) (Figure 3).

Due to concurrent symptoms (dizziness, headache), the patient was referred to the internal medicine department. Findings from laboratory showed a slight anemia (hemoglobin: 10.1 mg/dl) and signs of kidney failure (creatinine levels of 13.1 mg/dl). Platelet count and coagulation were within normal limits.

Because of laboratory test compatible with acute kidney failure, the patient was hospitalized. A subsequent biopsy showed signs compatible with thrombotic microangiopathy. An autoimmunity profile test was performed before plasmapheresis, hemodialysis, and corticoid therapy were initiated. During hospitalization,

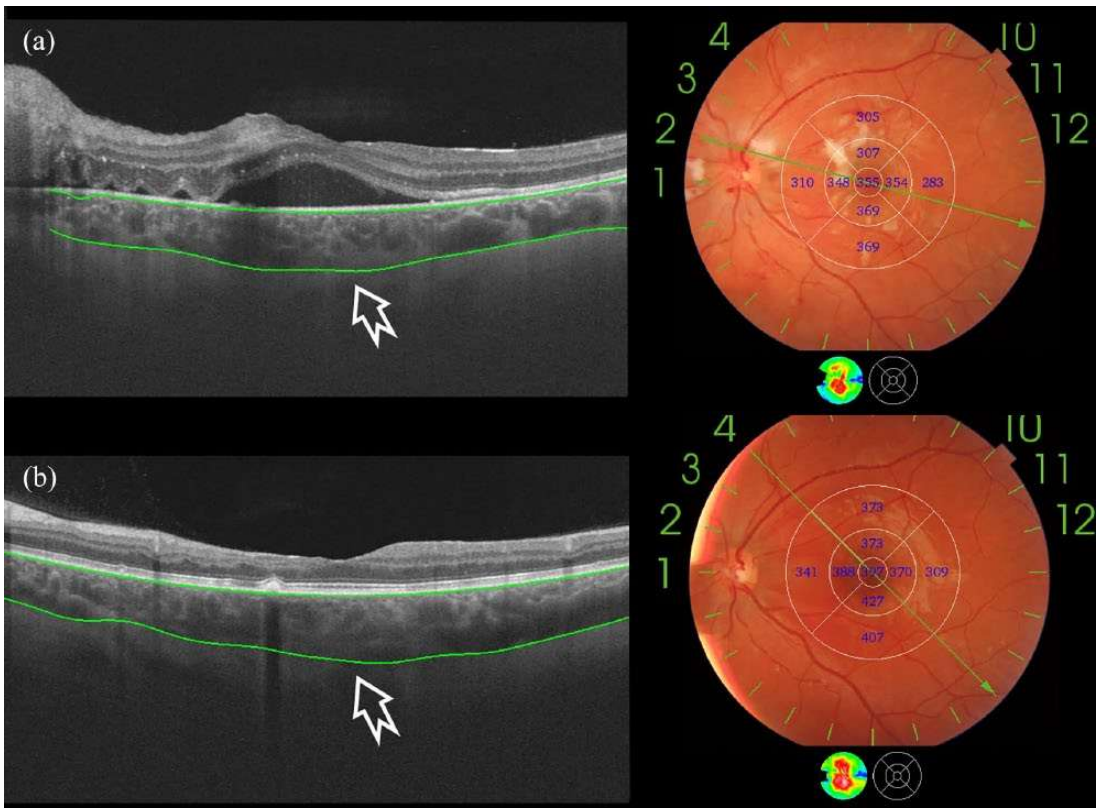

Figure 2. (a) Fundus photographs showing microhemorrhages and cotton-wool spots, SS-OCT focal serous retinal detachments with increased choroidal thickness. (b) Fundus photographs showing resolution of microhemorrhages and cotton-wool spots, SS-OCT resolution of focal serous retinal detachments with focal retinal atrophy.

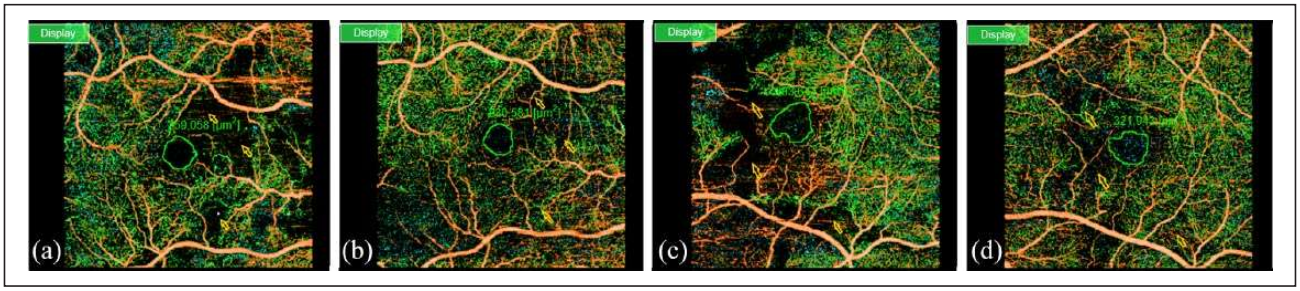

Figure 3. Optic coherence tomography angiography (OCTA) of the right eye and left eye at diagnosis (a and c) and after eculizumab treatment (b and d). It is noticeable in both eyes, revascularization of the initial perifoveal ischemic areas (yellow arrows) and changes in the size of the FAZ (259–230 µm in the right eye and 383–321 µm in the left eye).

the patient developed elevated lactate dehydrogenase (LDH) levels, schistocytes were detected, anemia and platelet counts worsened. The ophthalmoscopic examination revealed optic disk swelling, with an increase of the SRDs.

The diagnostic suspicion was a thrombotic microangiopathy. Based on the laboratory findings, the patient was diagnosed with aHUS. Eculizumab—a humanized monoclonal antibody that blocks the complement activity by cleavage of the complement protein C5—was prescribed at a dose of 900 mg weekly for 4 weeks and 1200 mg biweekly for 1 month.[1] After initiation of eculizumab treatment, hemoglobin increased and LDH levels decreased. However, renal function did not improve, and the patient was scheduled for dialysis after hospital discharge. At the ophthalmological follow-up visit, 4 months after discharge, the patient had a BCVA of 20/25 in his RE and 20/20 in his LE. Fundoscopic exam revealed resolution of the SRD and the cotton-wool spots with reactive hyperplasia of the retinal pigment epithelium, choroidal

thickness measures improved, but still were over the mean (Figures 1 and 2). OCTA showed revascularization in both eyes with only a few remaining areas of perifoveal ischemia (Figure 3).

Discussion

Purtscher-like retinopathy is an occlusive microvasculopathy with an estimated incidence of 0.24 persons per million per year. The most widely accepted theory to explain the cause of this condition is microembolization of C5 and leukocyte aggregates, causing arteriolar precapillary occlusion and microvascular infarct of the retinal nerve fiber layer and hemorrhages.[2]

Atypical HUS is seen in 5%–10% of HUS cases. Prognosis is poor, with a high mortality and morbidity rate in the acute phase. Progression to end-stage renal disease occurs in up to 50% of cases.[3] Unopposed activation of the complement system results in formation of the membrane attack complex on cell surfaces, especially endothelial cells of the microcirculation such as the kidney or retina. This leads to obstruction by fibrin-platelet thrombi and systemic multi-organ disfunction.[2–4]

We report a case of Purtscher-like retinopathy in which OCTA was used to assess the course of the macular vascular perfusion during the entire disease process. To our knowledge, this is the first case of its kind reported to date. Initially, we observed areas of perifoveal ischemia with an increase of the FAZ due to the microvascular obstruction. Initiation of eculizumab treatment halted the thrombotic microangiopathy process and, consequently, both systemic and ophthalmologic variables improved. OCTA showed revascularization of the previous perifoveal ischemic areas with a secondary reduction of the previous FAZ measures, while there was also an improvement in the SS-OCT initial findings.

Furthermore, a reduction in choroidal thickness after the introduction of eculizumab treatment was noticed; however, these values were still over the known mean in the last visit. This phenomenon can also be seen in other inflammatory diseases and could be used in the follow-up of these patients.[5]

Miguel et al.[6] performed a systematic review of Purtscher-like retinopathy, finding that the only prognostic factors for improved BCVA after 6 months of follow-up were the absence of macular edema and of the pseudo cherry red spot. Our findings suggest that evaluation of the macular capillary plexus revascularization by OCTA could help to predict an improvement of BCVA in these patients and the measurement of choroidal thickness could give us information about the resolution of the pathologic process.

Declaration of conflicting interests

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