Photo Essay

Ultra-wide Field Fluorescein Angiography in Retinitis Pigmentosa with Intermediate Uveitis

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PRESENTATION

An 18-year-old man presented with a history of night blindness and decreased vision in both eyes. There was no family history of retinitis pigmentosa (RP). Best corrected visual acuity was 6/18 in his right eye and 6/24 in the left eye. On slit lamp examination, few pigmented keratic precipitates were seen in both eyes, with occasional anterior chamber cells. There were no cataracts, but retrolental cells were seen in both eyes. Intraocular pressure in both eyes was 16 mmHg. Both eyes showed mild disc pallor, pigment spicules around the equator, and inferior preretinal exudates [Figure 1].

Disc leakage and petaloid leakage in the macula were noted on ultra-widefield fluorescein angiograms (UWFA, Figure 2). Central macular thickness (CMT) based on optical coherence tomography (Cirrus HD-OCT, Carl Zeiss Meditec, Dublin, CA) was 500 microns in the right eye and 433 microns in the left eye [Figure 3] with intraretinal cystoid changes. Scotopic electroretinogram was extinguished, and the patient had a small visual field of 10° in both eyes. Chest X-ray, hemogram, and angiotensin converting enzyme levels were normal, and VDRL and Mantoux tests were negative. A diagnosis of non-syndromic RP with intermediate uveitis and cystoid macular edema (CME) in both eyes was made.

The patient was started on oral prednisolone 1 mg/kg daily. Two weeks after therapy, the visual acuity improved to 6/9 in both eyes with resolution of CME. CMT was reduced to 282 microns in the right eye and 273 microns in the left eye, with marked decrease in cystic spaces.

DISCUSSION

Ultra-widefield imaging and fluorescein angiography (UWFI and UWFA) have improved imaging of peripheral retinal disorders and uveitis cases.[1,2] The conventional fundus camera has a limited field of view, capturing 30° and 50° of the fundus in one frame. Using various montaging techniques, one can portray up to a maximum of 140°. UWFI (Optos Inc., Marlborough, MA, USA) employs dual wavelength scanning laser ophthalmoscopy and can give a single image covering up to 82% or 200 degrees of the retina. This system takes advantage of the optics of an ellipsoid mirror which contains two focal points. The laser beam of the UWFI camera is directed through one of the focal points, while the patient's eye is positioned such that the second focal

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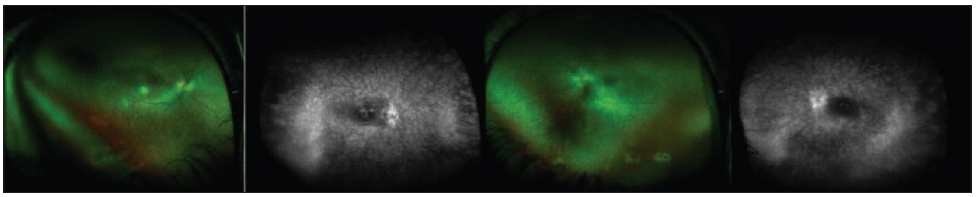

Figure 1. Fundus photographs and fluorescein angiograms of the both eyes shows typical bony spicule pigmentation, and preretinal exudates inferiorly suggestive of retinitis pigmentosa with intermediate uveitis.

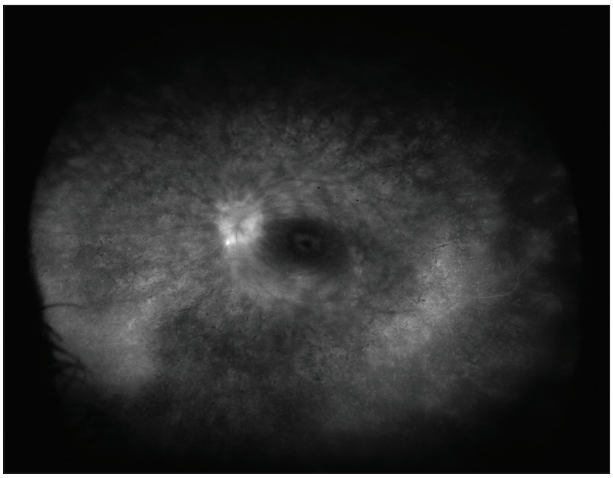

Figure 2. Ultra-wide field fundus fluorescein angiogram of the left eye shows disc leak and petaloid leakage at the macula.

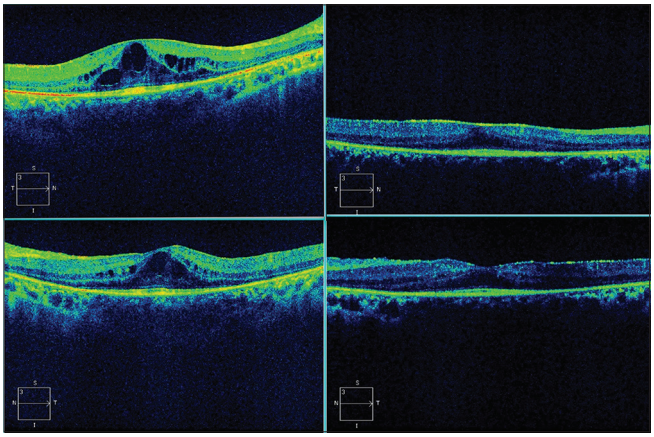

Figure 3. Pre- and post-treatment fundus imaging of the right (up) and left eyes (down). At presentation, central macular thickness (CMT) was 433 microns in the left eye with cystic spaces visible on optical coherence tomography.

point is located inside the patient's eye creating a 200 degree image.

CME has been reported in 11-70% of patients with RP.[3] However, reports on intermediate uveitis coexisting with RP are rare.[4] The presence of peripheral vasculitis, vascular leakage, and CME could masquerade as uveitis in RP. The exact cause of CME in RP is unclear but is presumed to be due to breakdown of the blood retinal barrier and decreased pumping efficiency of the retinal pigment epithelium. Intermediate uveitis which is characterized by disc leakage, CME on fluorescein angiogram and preretinal exudates have rarely been reported in RP.[4] There is limited data in the literature regarding the role of UWFA in RP. Kaufman et al showed that UWFA revealed peripheral vascular leakage indicative of vasculitis in 15 of 25 patients (60%) in RP.[5] Miller and colleagues showed that 31% of RP patients showed peripheral vascular leakage, and 63% patients showed CME without clinical signs of Coats'-like response.[6]

In the patient described herein, UWFI and UWFA were able to simultaneously pick up peripheral pre-retinal exudates, clinical features of RP, central CME and disc leakage. Thus, a single investigation helped us identify and document both pathologies. The OCT showed significant resolution of CME after 2 weeks of therapy and visual acuity of the patient was also improved. This is the evidence supporting that CME in our patient was inflammatory in nature. Thus, although rare, one should also rule out the presence of intermediate uveitis patients with RP with CME. UWFI/UWFA may prove to be a promising tool for simultaneous imaging of central and peripheral retinal pathologies.

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Conflicts of Interest

There are no conflicts of interest.

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