ABSTRACT

Objective
To report a case of inflammatory foveal mass and cystoid macular edema and the role of optical coherence tomography in its diagnosis and treatment monitoring.

Methods
This is a case report of a healthy, young female who experienced sudden onset of wavy, blurring of vision. Clinical examination revealed an idiopathic, yellow, foveal mass and cystoid macular edema.

Results
The condition resolved rapidly after treatment with oral corticosteroids suggesting an inflammatory nature. The vision returned to 20/20 after disease resolution.

Conclusion
Optical coherence tomography was useful in diagnosing this disease condition and monitoring treatment response.

CME is a common, nonspecific pathologic response to disturbance of the blood-retinal barrier (BRB). Disruption of the tight junctions between the endothelial cells of the retinal vasculature or retinal pigment epithelial (RPE) cells results in extravasation of fluid and plasma proteins into the extracellular spaces of the retina. CME develops when fluid accumulation forms cystic spaces in the outer plexiform and inner nuclear layers of the parafoveal retina. Over time, these cystic spaces may coalesce, enlarge, and produce macular defects or holes. Disruption of Muller, photoreceptor, and RPE cell function has been shown to occur in CME and result in diminished visual acuity.1,5

CME may develop from a variety of conditions, such as uveitis, diabetic and hypertensive retinopathy, venous occlusive disease, choroidal neovascularization, radiation retinopathy, vitreomacular-traction syndrome, epiretinal
membrane, retinitis pigmentosa, and ocular trauma or surgery. In the case of uveitis, inflammatory mediators are believed to promote leakage from the BRB. The treatment of uveitis-associated CME is directed toward stopping the inflammatory process using immunosuppressive medications to eliminate production of inflammatory mediators and reestablish normal BRB function.  

FA is the principal diagnostic method for confirming the presence of CME. Leakage of sodium fluorescein from disrupted BRB followed by dye accumulation in the cystic spaces result in the characteristic “petalloid” or honeycomb pattern seen in the angiogram. OCT is a relatively new imaging modality that analyzes reflected light from the retina to create highly detailed, cross-sectional images. In CME, hyporeflective intraretinal cystic spaces are visualized by OCT. The analytical software supplied with the OCT machine can perform quantitative measurements of retinal thickness which is helpful for monitoring response to treatment. In addition to being a sensitive method for detecting CME, OCT may demonstrate the presence of other pathologic conditions associated with CME, such as epiretinal membranes or vitreomacular traction. In this patient, OCT revealed the presence of a solid, hyperreflective foveal mass with overlying CME. OCT was also useful in monitoring the resolution of CME and disappearance of the foveal mass. The rapid disappearance of the foveal mass and CME after corticosteroid treatment strongly suggests that the
lesion was inflammatory in nature. Recurrence of the condition with premature cessation of antiinflammatory treatment added further evidence that an inflammatory process was occurring. We describe this condition as an idiopathic inflammatory foveal granuloma. The absence of concomitant signs of uveitis, such as anterior chamber or vitreous cells, snowbanking, and choroidal nodules, make it difficult to attribute this case to other established common causes of uveitis-induced CME, such as anterior uveitis, pars planitis, or ocular sarcoidosis. The rapid return of normal vision after treatment suggests that this is a relatively benign condition. In this case, OCT provided a noninvasive means of identifying the intraretinal pathologic processes and pointed to a treatment strategy. The value of performing OCT in the management of CME is clearly demonstrated in this case report.

References

Results
A 35-year-old Caucasian male was referred for evaluation of posterior uveitis. Vitritis, retinal vasculitis, and capillary dropout in the peripheral retina were observed. Fluorescein angiography confirmed the changes. Systemic and blood work-up revealed negative findings. Oral steroid combined with a systemic immunosuppressant had no effect on the disease course. The pattern of the disease and lack of response to immunosuppressives were consistent with a diagnosis of IRVAN syndrome. The patient remained on regular follow-up and maintained good visual acuity of 6/6 in the affected eye.

Conclusion
Clinicians should be aware of the existence of IRVAN syndrome to prevent injudicious use of steroids and systemic immunosuppressants in these patients.

IDIOPATHIC retinal vasculitis, aneurysms, and neuroretinitis (IRVAN) syndrome is diagnosed after normal systemic work-up and lack of treatment response to immunosuppressants.

A 35-year-old Caucasian was referred by his optician for evaluation of posterior uveitis. The patient experienced “cloudy” vision in his right eye for a few years, not severe enough to seek consultation. Otherwise, he was healthy. Systemic evaluation revealed no history of bowel, respiratory, or skin disorders. He had not travelled recently to the tropics, did not smoke, and had no family history of eye or systemic disease.

On examination, best-corrected visual acuity was 6/6 OU (OD: -1.25/+2.25 x 85; OS: –4.75/+0.50 x 80). Pupillary reactions, intraocular pressure (IOP), and color vision were normal in the right eye. Anterior-segment examination revealed no history of bowel, respiratory, or skin disorders. He had not travelled recently to the tropics, did not smoke, and had no family history of eye or systemic disease.

Examination of the right eye also revealed some fine dusting of keratic precipitates. There were 1+ cells in the anterior chamber and anterior vitreous. A mild posterior subcapsular cataract was present. The vitreous was syneretic and peripherally there were preretal vitreous opacities. Multiple aneurysmal dilatation of the arterioles and hard exudates associated with areas of capillary nonperfusion were noted at the peripheral retina. Fluorescein angiogram (FA) confirmed areas of capillary nonperfusion and aneurysmal dilatation of the arterioles (Figure 1). There was late disc staining, suggestive of neuroretinitis, with leakage from new vessels at the disc.

Extensive uveitis work-up, including ESR, C-reactive protein, antinuclear antibodies, rheumatoid factor, antcardiolipin antibodies, C3, C4, Bartonella antibodies, angiotensin converting enzyme level, fluorescent