Mikulicz disease is within the clinical and pathological spectrum of IgG4-related disease irrespective of the organ of origin. However, hyalized dense fibrosis as observed in cases 1 and 2 is not typical for IgG4-related diseases to our knowledge. We speculated that this difference might depend on anatomical characteristics of the lacrimal gland (eg, tightly surrounded by muscles and bone) or the duration of this disease. In addition, it is interesting that acinar subunits were considerably atrophied, although no patients showed lacrimal dysfunction such as keratoconjunctivitis sicca. Similarly, pancreas with autoimmune pancreatitis usually shows normal exocrine pancreatic function irrespective of acinar atrophy. This discrepancy has not been well documented until now, and further pathophysiological examinations are mandatory for this issue.

In all of the cases, we performed unilateral resection of the lacrimal gland to make a diagnosis. However, surgery was also intended as part of the treatment in case 2 because steroid therapy was undesirable for the patient’s diabetes mellitus. The left eyelid swelling, his chief symptom, improved after surgery. Thus, excisions of enlarged lacrimal glands (occasionally bilateral) may be an alternative treatment for Mikulicz disease when steroid treatment is undesirable.

In conclusion, the 4 cases of Mikulicz disease reported here had chronic sclerosing inflammation of the lacrimal gland with IgG4-stained plasma cell infiltration. These findings support the theory that Mikulicz disease is within the clinical spectrum of IgG4-related sclerosing diseases.

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Financial Disclosure: None reported.


Spontaneous Filtration Bleb as a Consequence of Scleritis

Report of a Case. A 40-year-old woman with systemic lupus erythematosus was seen for evaluation of persistent redness and discomfort of variable intensity in the left eye during the previous 2 years. Her best-corrected visual acuity was 20/30 OD and 20/40 OS, with intraocular pressure of 10 mm Hg in the right eye and 11 mm Hg in the left eye. Slitlamp biomicroscopy showed mild dilatation of conjunctival and episcleral vessels of the right eye. The left eye demonstrated diffusely dilated episcleral vessels with a flat, supertemporal perilimbal avascular region with focal surrounding conjunctival edema. Magnetic resonance imaging performed to assess for orbital venous outflow disturbances was unrevealing. Topical corticosteroid therapy (1% prednisolone acetate) with subsequent tapering resulted in limited clinical improvement bilaterally.

At follow-up examination 2 years later, the patient reported substantial left eye pain. Visual acuity in both eyes was unchanged. Examination of the right eye demonstrated engorgement of both superficial vessels and the deeper scleral vascular plexus with a supertemporal perilimbal avascular patch (Figure 1). The left eye demonstrated apparent underly-
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Financial Disclosure: None reported.


Comment. Scleral inflammatory disease is a deep and destructive inflammation presumed to be incited by autoimmune system dysregulation and is often classified on the basis of the site of pathologic findings and severity of inflammation. Scleritis is characterized by edema and inflammatory cell infiltration of the sclera and is commonly associated with identifiable systemic disease and often with ocular complications. The development of a spontaneous filtering bleb with consequential hypotony is an unusual complication of anterior scleritis, and, to our knowledge, our case is the first reported.

Thinning secondary to scleral inflammation, with a resultant focal aqueous shunt and collection external to the subconjunctival or sub-Tenon space, analogous to a deliberate guarded filtration procedure, is the presumed underlying mechanism. Ultrasound biomicroscopy to quantify scleral thinning was unavailable; however, its potential role in the assessment of scleritis subtypes, disclosing disease progression and judging treatment efficacy, has been demonstrated. This unusual complication emphasizes the destructive nature of scleral inflammatory processes and a management dilemma between the risks of infection and hypotony-related complications with observation compared with the potential to incite further, more substantial scleral inflammation with a surgical approach.

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Photoreceptor Disruption Secondary to Posterior Vitreous Detachment as Visualized Using High-Speed Ultrahigh-Resolution Optical Coherence Tomography

Optical coherence tomography (OCT) has been shown to be beneficial in the diagnosis of posterior vitreous detachment (PVD) and vitreomacular traction. In 2001, ultrahigh-resolution OCT (UHR-OCT), capable of 3-µm axial resolution in the human eye, has demonstrated refined visualization of outer retinal layers. Dramatic advances in the imaging speed of OCT enable high pixel density, high-definition imaging with further improved image quality. The following is a case of bilateral photoreceptor disruption secondary to PVD, imaged using high-speed UHR-OCT.

Report of a Case. A 66-year-old man underwent cataract extraction and placement of a posterior chamber intraocular lens (PCIOl) in the left eye. One day after surgery, his visual acuity returned to 20/20 OS. One week after surgery, he reported a decline in vision in the left eye associated with a floater. Best-corrected visual acuity was 20/25 OD and 20/40 OS. Anterior ocular examination findings revealed moderate nuclear sclerosis in the right eye and a well-placed PCIOL in the left eye. Dilated fundus examination revealed a Weiss ring in both eyes. In the asymptomatic right eye, high-speed UHR-OCT demonstrated vitreofoveal attachment (seen in some OCT images; image not shown herein), slight foveal thickening, irregular fovea, and minimal interruption of the photoreceptor outer segment layer (Figure, A). In the symptomatic left eye, there was a detached posterior hyaloid with an associated pseudopapillorum, interruption of the foveal photoreceptor outer segment layer, and an irregular fovea (Figure, B).

Four months later, visual symptoms had improved in the left eye, although the floater persisted. No ocular symptoms were noted in the right eye. On examination, best-corrected visual acuity was 20/25 OU. Anterior and posterior ocular examination findings remained unchanged in both eyes. High-speed UHR-OCT imaging was again performed, which revealed progression to complete vitreomacular separation in the right eye with return of normal foveal contour, as well as greater interruption of the foveal photoreceptor outer segment layer (Figure, C). In the left eye, the photoreceptor outer segment layer abnormality had decreased, and foveal contour had returned to normal (Figure, D).

Comment. This case demonstrates bilateral lucencies within the foveal photoreceptor outer segment layer secondary to PVD as visualized using high-speed UHR-OCT. In the asymptomatic right eye, an increase in photoreceptor disruption was observed on detachment of the posterior hyaloid. In the symptomatic left eye, a larger interruption in the photoreceptor outer segment layer, associated with foveal posterior hyaloid separation, was evident at the initial examination. Four months later, foveal photoreceptors had returned to normal, and visual acuity had improved.

Zambarakji et al reported similar OCT findings in patients with macular microholes. Their patients had small foveal lesions evident on biomicroscopy. On standard resolution OCT 3 imaging, most of the patients had small outer retinal defects, and many had partial PVD. In contrast, our patient had no evidence of foveal abnormalities on biomicroscopy. The initial manifestation of our patient’s right eye is also similar to cases reported by myself and colleagues. However, those patients had vitreofoveal traction without evidence of photoreceptor disruption and with the added symptom of metamorphopsia, which our patient did not have.