Albini et al. show a fascinating video illustrating a unique finding—a nearly opaque posterior capsular plaque that developed in the eye of a pseudophakic woman who, several years previously, underwent a diagnostic vitrectomy in the same eye for suspected intraocular lymphoma. The earlier biopsy and brain MRI were negative for malignancy. This capsular biopsy was positive for B-cell lymphoma, and repeat MRI showed new central nervous system (CNS) lesions consistent with CNS lymphoma. Although the vitreous, subretinal space, sub-retinal pigment epithelium (RPE) space, and optic nerve are known sites for intraocular lymphoma cells to accumulate, the posterior capsule is highly unusual.

An initial negative vitreous biopsy in the setting of intraocular lymphoma is not unusual. Persistence by the ophthalmologist in the pursuit of the diagnosis of primary intraocular lymphoma is often necessary. Multimodal imaging consisting of fluorescein angiography and fundus autofluorescence looking for “leopard spots” and subtle arterial leakage, as well as optical coherence tomography to detect subretinal and sub-RPE abnormalities, can be helpful in cases where the fundus appears normal. Other confirmatory studies may include systemic evaluation, neuroimaging, lumbar puncture, and multiple sampling of intraocular tissue.

This video illustrates several important points: 1) Use of a 6-mm cannula when the view is suspect; 2) confirmation of the presence of the cannula in the vitreous cavity prior to beginning infusion; 3) obtaining an undiluted specimen by the infusion of air as opposed to saline to maintain intraocular pressure while not diluting the specimen; and 4) use of small-gauge vitrectomy instrumentation to obtain a cytologic specimen.

Since cytology remains the best way to diagnose intraocular lymphoma, proper handling of the biopsy specimen is critical. Discussing these details with an ocular pathologist prior to the biopsy is recommended. This case also illustrates that using small-gauge vitrectomy instrumentation does not result in untoward surgical trauma on the cells as some feared initially with the advent of the smaller port cutters and higher cutting rates.

In summary, primary vitreoretinal lymphoma is the ultimate cause of the masquerade syndrome and should be kept in mind as a potential diagnosis in the proper clinical setting, even when prior vitreous biopsies are negative. Persistence in obtaining the correct diagnosis is necessary. Although highly unusual, this case illustrates that lymphoma cells can accumulate as a posterior capsular plaque in the setting of a pseudophakic, prior vitrectomized eye.

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ABSTRACT: Primary vitreoretinal lymphoma (PVRL) can be a diagnostic challenge and commonly presents as a partially steroid-responsive vitritis or as subretinal cream-colored infiltrates. The authors present a patient with PVRL who initially presented with bilateral vitritis; however, after two non-diagnostic vitrectomy specimens and two unremarkable brain MRIs, she was lost to follow-up. She presented 2.5 years later with a white plaque on the posterior capsule of her left intraocular lens, though the vitreous cavity was free of infiltrate. Repeat biopsy revealed diffuse large B-cell lymphoma, and brain MRI demonstrated an enhancing lesion of the cerebellum, consistent with primary central nervous system lymphoma.
A 70-year-old woman presented with gradually worsening floaters bilaterally. Best-corrected visual acuity was 20/25 in the right eye and 20/20 in the left eye. Past ocular history was notable for uncomplicated cataract surgery in both eyes. She had been treated with sub-Tenon’s triamcinolone without resolution of symptoms. Past medical history was notable for breast cancer treated with mastectomy, hypertension, and depression. Anterior segment exam was unremarkable. Dilated fundus exam was notable for 2+ vitreous cells in both eyes, but the retina was otherwise unremarkable.

MRI of the brain and lumbar puncture were unremarkable. After diagnostic vitrectomy of the left eye to evaluate for primary vitreoretinal lymphoma (PVRL), the cytology showed atypical appearing lymphocytes. Polymerase chain reaction (PCR) did not indicate presence of a clonal population, and the sample was not sufficiently cellular for flow cytometry. Gram stain and cultures did not show evidence of an infectious process.

Due to the equivocal results, diagnostic pars plana vitrectomy (PPV) was performed in the right eye, which also showed atypical appearing lymphocytes, though the PCR and flow cytometry results did not indicate the presence of lymphoma. The microbiology workup was negative. The patient’s floaters resolved after surgery, and she was observed with serial exams and MRIs every 6 months.

After being lost to follow-up for 2.5 years, she then presented with deterioration of vision to hand motions in the left eye. She was noted to have a white plaque behind her intraocular lens (IOL) in the left eye, which hindered our view of the fundus. B-scan ultrasonography did not show a mass or retinal detachment. The patient underwent repeat PPV of the left eye to excise the
plaque for further analysis. Phakic IOL and infectious etiologies such as endogenous fungal endophtalmi-
titis were considered.

In the video (available at www.healio.com/ 
OSLIRetina), three 25-gauge ports were placed, and
a 6-mm infusion cannula was confirmed to be in the
vitreous cavity by pressing it against the posterior
lens capsule. With air infusing the vitreous cavity,
the plaque was easily aspirated with the vitrector to
obtain a non-dilute sample for pathology. Behind the
plaque, there was no evidence of lymphoma in the
vitreous cavity or retina. The pathologic specimen
confirmed the diagnosis of large B-cell lymphoma.
MRI of the brain now revealed enhancing lesions
within the cerebellum, consistent with primary cen-
tral nervous system lymphoma (PCNSL).

PVRL is commonly associated with PCNSL, and
both are most often of B-cell origin.1 Although 15%
to 25% of patients with PCNSL develop ophthal-
mic manifestations of lymphoma, 56% to 90% of patients
with PVRL have or will develop CNS manifestations.2
PVRL can be a diagnostic challenge, as it is a “mas-
quarade syndrome,” which is responsive to cortico-
steroids just like noninfectious uveitis. Posterior uve-
itis is the most common sign and presents frequently
with increased floaters and blurred vision, as in this
case.2 Patients may also present with cream-colored
retinal or retinal pigment epithelium infiltrates, with
a leopard-skin appearance on fluorescein angiogra-
phy.3 As far as we are aware, this case represents the
first report of PVRL manifesting as a plaque on the
posterior capsule of an IOL.

Specimens for histologic evaluation can be ob-
tained by either fine-needle vitreous aspiration or
vitrectomy. As in this case, multiple biopsies may be
necessary to obtain a definitive pathological diagno-
sis. Specimens should be carefully processed to pre-
vent cell degeneration that makes diagnosis difficult.
Obtaining only a sparse number of cells is the most
common reason for a negative vitrectomy specimen,
though vitreous specimens may also contain reac-
tive T-cell lymphocytes, necrotic cells, fibrin, and
inflammatory debris that can confound identification
of malignant cells.4 Retinal or chorioretinal biopsies
may be necessary in certain cases. Other useful diag-
nostic tools include vitreous cytokine analysis show-
ing IL-10:IL-6 ratio greater than 1.0, flow cytometry
to examine cell surface markers and demonstrate a
monoclonal B-cell population, and PCR to amplify
the immunoglobulin heavy chain DNA and detect
gene rearrangements.5

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