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ABSTRACT

SUMMARY: Primary intraocular lymphoma (PIOL) is a rare form of primary central nervous system lymphoma that poses diagnostic challenges because of its nonspecific clinical features and complex imaging characteristics. This paper presents a focus case and 2 companion cases, highlighting the complexities in identifying and treating PIOL. In the focus case, a 66-year-old man experienced gradual painless vision loss with choroidal thickening on funduscopic examination and subsequent follow-up MRI. Transvitreal biopsy confirmed PIOL, and the patient was treated with intravitreal steroids and systemic rituximab without recurrence. Companion case 1 involved a 66-year-old woman with vision changes and choroidal thickening with episcleral extension on MRI suggestive of intraocular lymphoma and ultimately treated with radiation with the presumed diagnosis of PIOL. In the companion case 2, a 63-year-old man with ocular symptoms was diagnosed with chronic lymphocytic leukemia along with vitreoretinal Richter transformation. Enucleation was performed because of a lack of visual potential and failure of chemotherapy, which confirmed PIOL. Distinguishing PIOL from other ocular conditions is crucial, given its potential for CNS involvement. Imaging plays a vital role in corroborating clinical findings. While cytology remains the standard for diagnosis, supplementary tests, including cytokine analysis, immunohistochemistry, and flow cytometry, provide additional insights. PIOL treatment strategies are tailored to disease extent, ranging from locoregional chemotherapy to invasive enucleation. CNS involvement carries a poor prognosis and must evaluated and surveilled with MRI. In conclusion, this case series reviews the clinical and radiologic features of PIOL, emphasizing the significance of diagnostic imaging in determining disease extent and guiding treatments.

ABBREVIATIONS: CLL = chronic lymphocytic leukemia; FFA = fundus fluorescein angiography; IHC = cytometric immunohistochemistry; IOL = intraocular lymphoma; OCT = optical coherence tomography; PCNSL = primary central nervous system lymphoma; PIOL = primary intraocular lymphoma; RPE = retinal pigment epithelium; UBM = ultrasound biomicroscopy

Intraocular lymphomatous malignancies include a heterogeneous group of pathologies that involve the vitreous humor and retina or the uvea.[1] Primary intraocular lymphoma (PIOL) is a rare form of primary central nervous system lymphoma (PCNSL), usually originating from germinal center B-cells in the retina. PIOL is generally bilateral (in up to 90% of patients) and presents between the fourth and sixth decades of life.[2] There is remarkable variability in clinical presentation and sites of disease, which often extend into the cranium and CSF. At the initial presentation of PIOL, about 16%–34% of patients tend to have CNS involvement.[2]

PIOL often presents as bilateral, asymmetric vision changes, including blurry vision and floaters.[3] On direct funduscopic examination, the posterior segment may exhibit signs of vitritis with hypercellular inflammatory infiltrate. The retinal pigment epithelium (RPE) is a major site for lymphocytic proliferation. It can show patchy, yellow-orange infiltrate, more specific for PIOL.[4] Multimodal or ocular imaging plays an important role in establishing a diagnosis and monitoring response to treatment. These investigations include optical coherence tomography (OCT),[4] fundus fluorescein angiography (FFA),[5] indocyanine green angiography,[4] and fundus autofluorescence.[1] Funduscopic imaging modalities have shown positive and negative predictive values of 88.9% and 85%, respectively.[3] The diagnostic work-up often includes radiologic examination, primarily by using MRI. PIOL may show nonspecific, diffuse uveal contrast enhancement with retinal T2 iso-to-hypointensities, representing RPE infiltrate.[6-10] Imaging findings alone cannot reliably differentiate PIOL from uveitis and ocular melanoma and may yield false-negative results. Definitive diagnosis requires an anterior chamber tap, vitreous aspiration, or transscleral biopsy.[5] A summary of the common diagnostic modalities and their findings may be found in the Table.

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Review of common modalities used in the diagnosis and follow-up of PIOL with the associated findings and utility

Diagnostic Technique	Findings	Advantages	Limitations
Direct fundoscopy	• Anterior chamber cells • Keratin precipitates • Yellow white subretinal infiltrates	• Office-based examination	• Limited by opacities • Limited sensitivity to early or subtle changes
Ultrasound biomicroscopy	• Ciliary body thickening • Choroidal lesions	• High resolution of anterior segment • Useful in the setting of ocular opacities	• Anterior segment only • Specialized testing
Optical coherence tomography	• Vitreous cells • Vitreomacular interface abnormalities • RPE thickening, nodularity, and detachment	• High resolution, cross-sectional retinal imaging • Noninvasive and rapid test	• Limited by opacities • Coexisting conditions may limit interpretation
Fluorescein angiography	• Hypofluorescence at sites of tumor blockage • Hyperfluorescent spots indicating leakage • Late staining of RPE	• High-resolution visualization of retina and choroid vasculature • Differentiates PIOL from retinal vascular disease	• Invasive procedure requiring contrast • Cannot visualize subretinal lesions
Diagnostic vitrectomy	• Atypical lymphoid cells on cytologic examination • *IgH* gene rearrangements • *MYD88* gene mutations • CD20, CD3, CD79a, PAX5	• The standard diagnostic test • Samples can be sent for molecular and genetic testing	• Infection • RPE detachment
MRI with contrast	• Nodular enhancement • Choroidal thickening • Diffusion restriction	• Detailed soft-tissue contrast • Evaluates extraocular and CNS involvement	• Expensive
CT with contrast	• Intraocular mass lesions • Locoregional osseous lesions	• More readily available • Sensitive for calcific lesions	• Only if MRI is contraindicated • Poor tissue resolution • Radiation exposure
PET/CT	• Abnormal tracer uptake • Intraocular or systemic hypermetabolic lesions	• Disease staging • Evaluates for systemic involvement	• Significant radiation exposure • Low-grade lesions may be difficult to identify

The present report reviews a case of PIOL with 2 companion cases that highlight important aspects of its diagnosis and salient differentials. Cases were identified through a keyword search of our institution's PACS from January 2005 to December 2023 by using the following terms: "intraocular lymphoma," "primary vitreoretinal lymphoma," "primary intraocular lymphoma," "primary CNS lymphoma ocular," and "PCNSL–ocular." This yielded 3 cases of PIOL based on the diagnostic criteria demonstrating positive imaging findings.

Focus Case

A 66-year-old man presented with a history of painless, progressive vision loss that began in 2004. Despite regular ophthalmologic surveillance, the patient's vision continued to deteriorate, culminating in complete loss of central vision and only peripheral light perception in March 2011.

Imaging. An initial funduscopic examination in 2004 revealed choroidal thickening with late staining on FFA, suggesting a possible lymphoma diagnosis. Brain MRI and PET/CT were initially negative for intraocular, intracranial, and systemic lymphoma. Over time, the patient's vision progressively worsened, accompanied by choroidal thickening during ophthalmologic examinations. Subsequent brain MRI and PET/CT scans performed 7 years later revealed enhancing choroidal thickening along the lateral dorsal aspect of the left eye (Fig 1).

Operative Report and Pathology. A vitreous fine needle aspiration of the left eye mass revealed benign reactive lymphoid hyperplasia. Flow cytometry analysis identified a CD19 and CD20 positive B-cell population with κ immunoglobulin light chain restriction, consistent with malignant lymphoma (Fig 2). After a thorough evaluation with radiation-oncology and a negative bone marrow biopsy, the patient underwent treatment with intravitreal steroids and intravenous rituximab. Yearly surveillance imaging and funduscopic examinations showed no recurrence of lymphoma to date.

Companion Case 1

A 66-year-old woman presented with 7 months of vision changes, primarily affecting the right eye. Ophthalmologic assessment demonstrated choroidal thickening, subretinal fluid with wavy anterior surface on OCT, diffuse choroidal and ciliary body thickening on B-scan ultrasonography, and mild, diffuse ciliary body thickening on ultrasound biomicroscopy (UBM, Fig 3*E*–G). The vitreous inflammation could be secondary to autoimmune inflammatory or infectious uveitis processes; however, the significant ultrasonographic changes were concerning for lymphoma.

Imaging. Additional diagnostic studies were pursued including MRI and FDG PET/CT. Brain and orbit MRI depicted diffuse choroidal thickening with posteroinferior episcleral extension, exhibiting isointense signal on T1- and T2-weighted sequences, restricted diffusion on DWI, and homogeneous postcontrast

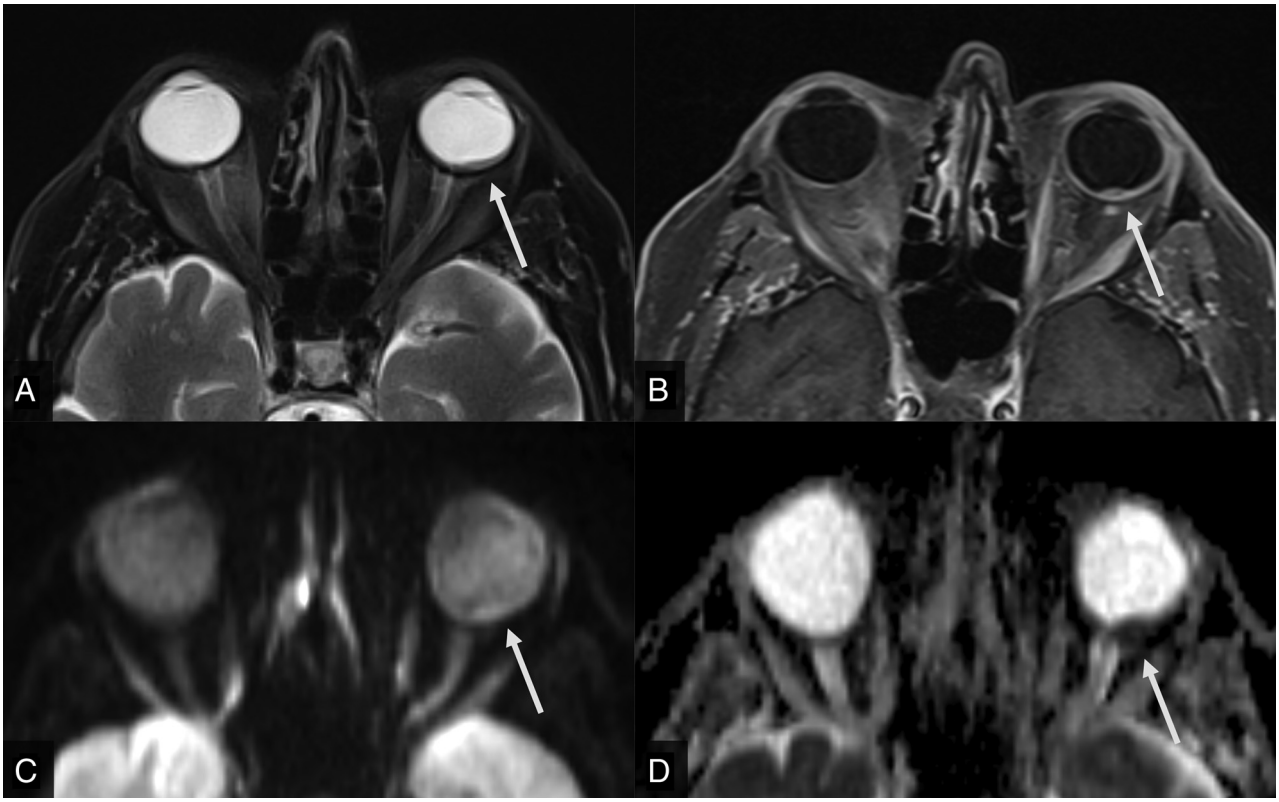

FIG 1. Axial T2-weighted (*A*) and postcontrast T1-weighted (*B*) images demonstrate focal enhancing choroidal thickening along the lateral dorsal aspect of the left eye (*arrow*) without retro-global extension, showing restriction on diffusion-weighted image (*C*) with a low corresponding apparent deficient coefficient (*D*).

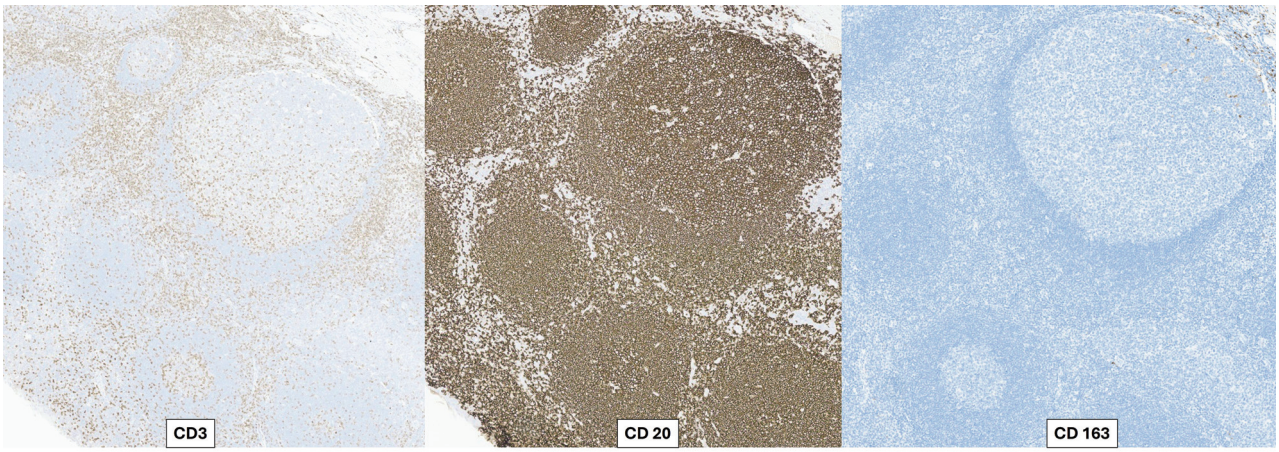

FIG 2. Histopathologic findings of intraocular lymphoma with the viterous fluid (cellblock) revealing moderate cellularity atypical B-cell positive lymphoid infiltrate, primarily consisting of large lymphocytes. The large lymphoid cell population is positive for CD20, and negative for CD3 and CD163. The background reactive T-cells are highlighted in the background on CD3 stains. Based on the cytomorphologic appearance of the atypical lymphoid cells and the immunohistochemical staining pattern, the findings are most consistent with diffuse large B-cell lymphoma.

enhancement with corresponding abnormal uptake on FDG-PET/CT (Fig 3*A*–*D*). The imaging differential diagnosis included neoplastic conditions, such as lymphoma or melanoma, alongside the possibility of an inflammatory process, such as uveitis.

Operative Report and Pathology. The patient underwent a lumbar puncture for CSF to evaluate for CNS involvement of the suspected neoplasm. Clinical pathologists performed a CSF cytometric immunohistochemistry (IHC), which exhibited normal surface antigens without monotypic B-cell population or blasts. At this juncture, the patient had the option of obtaining a biopsy to confirm the diagnosis or proceeding with low-dose radiation therapy to the eyes. Because of the biopsy risk and high likelihood of PIOL, the patient proceeded with radiation therapy by using 4

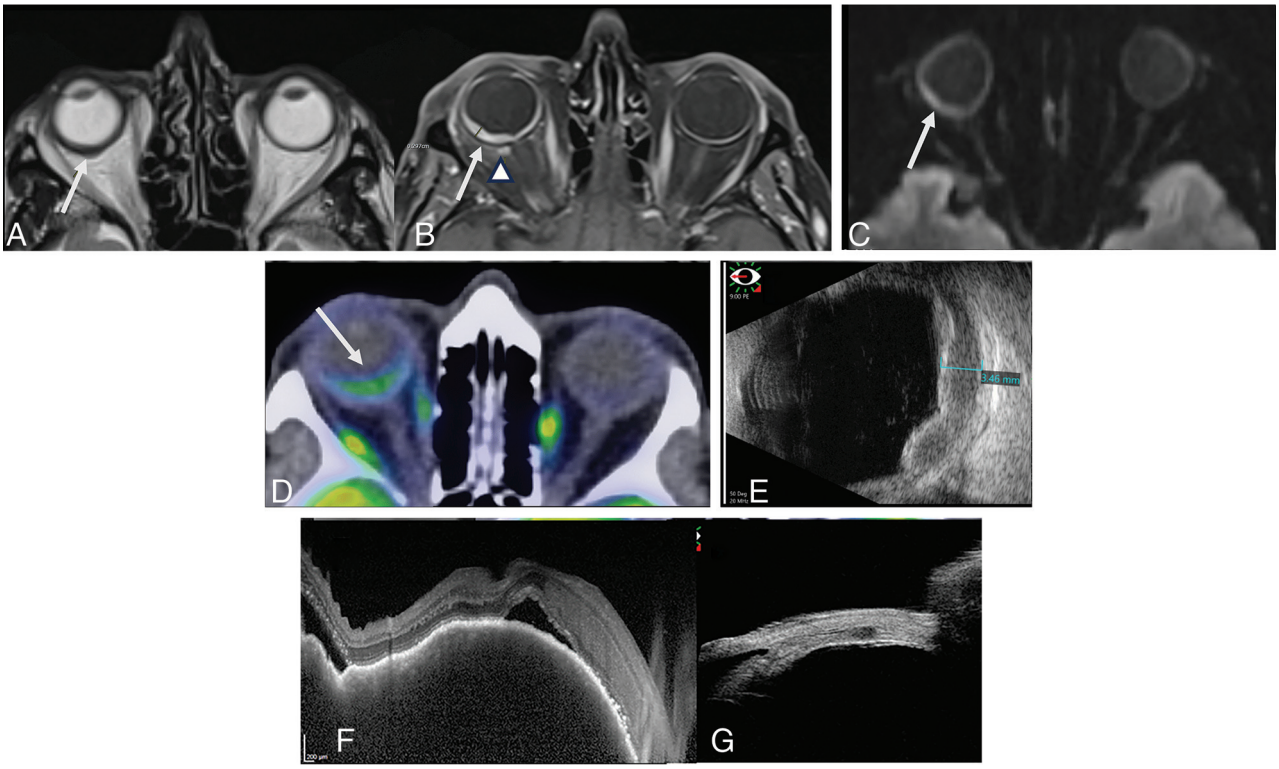

FIG 3. Axial T2-weighted (*A*) and postcontrast T1-weighted (*B*) images demonstrate abnormal, diffuse chorio-retinal thickening (*arrows*) of the right and central globe with episcleral extension (*arrowhead*). The lesion is isointense on T2 (*A*) and restricts on diffusion-weighted image (*C*). Axial FDG-PET/CT shows increased abnormal FDG uptake in the right and central choroid with trace thickening of the retina (*D*). B-scan ultrasound (*E*) shows diffuse choroidal thickening with an area of extraocular extension (*not shown*). The choroid measures up to 3.46 mm. Optical coherence tomography (*F*) demonstrates choroidal thickening with a wavy anterior surface and overlying pocket of subfoveal fluid. Mild diffuse ciliary body thickening on ultrasound biomicroscopy (*G*).

Gy in 2 fractions to both eyes instead of a confirmation biopsy. Yearly surveillance imaging and funduscopic examinations have shown no recurrence.

Companion Case 2

A 63-year-old man developed new left eye floaters and right visual blurring. Funduscopic examination showed vitreous hypercellularity, which is concerning for an autoimmune inflammatory or infectious process. Initial treatment with a course of corticosteroids failed to improve his symptoms. The patient returned a month later with a new pupillary defect and worsening left eye vision. Vitreous debris, disc edema, macular thickening, and a small lesion overlying the optic disc were observed on OCT examination (Fig 4*E*,-*F*). Laboratory results showed a significantly elevated lymphocyte count, raising concerns for lymphoma or leukemic infiltrate.

Imaging. A complete CT examination of the body was negative for systemic lymphoma or leukemia. A separate brain and orbit MRI identified a nodular soft tissue thickening near the left optic nerve head. Initial vitrectomy, including vitreous fluid and biopsy, yielded no malignancy on cytologic analysis, but a subsequent bone marrow biopsy was positive for chronic lymphocytic leukemia (CLL). A follow-up brain MRI 2 months later revealed significant growth of the left globe chorio-retinal mass (Fig 4*A*–*C*).

Operative Report and Pathology. A repeat vitreous aspiration with flow cytometry identified an ocular large B-cell lymphoma. Whole-body FDG-PET/CT showed increased tracer uptake in the left posterior globe without evidence of systemic lymphoma involvement (Fig 4*D*). Treatment involved several cycles of rituximab and methotrexate. Ultimately, chemotherapy did not improve symptoms, and because of worsening visual acuity and severe pain, enucleation was performed. Additional IHC following enucleation confirmed residual vitreoretinal B-cell lymphoma with CD3-positive lymphocytes and PAX5-positive, CD20-negative B-cells, indicating posttreatment residual disease. A final diagnosis of CLL with vitreoretinal Richter transformation to large B-cell lymphoma was made.

DISCUSSION

PIOL is an exceedingly rare disease, with an incidence of merely 0.27 per million patients, primarily affecting those over the age of 60 (72.1%) and more commonly in the white population (84.4%).[11] The typical clinical presentation involves the blurring of vision (72%), decreased visual acuity (63%), and floaters (60%).[5] Secondary intraocular lymphoma arises from lymphoma metastasis or recurrence in various body sites; however, this is comparatively rare.[8,12-14] The diagnosis of intraocular lymphoma primarily relies on clinical assessment during ophthalmologic examination, which may reveal findings such as posterior uveitis, vitreous haze, exudative retinal detachment, or hemorrhage.

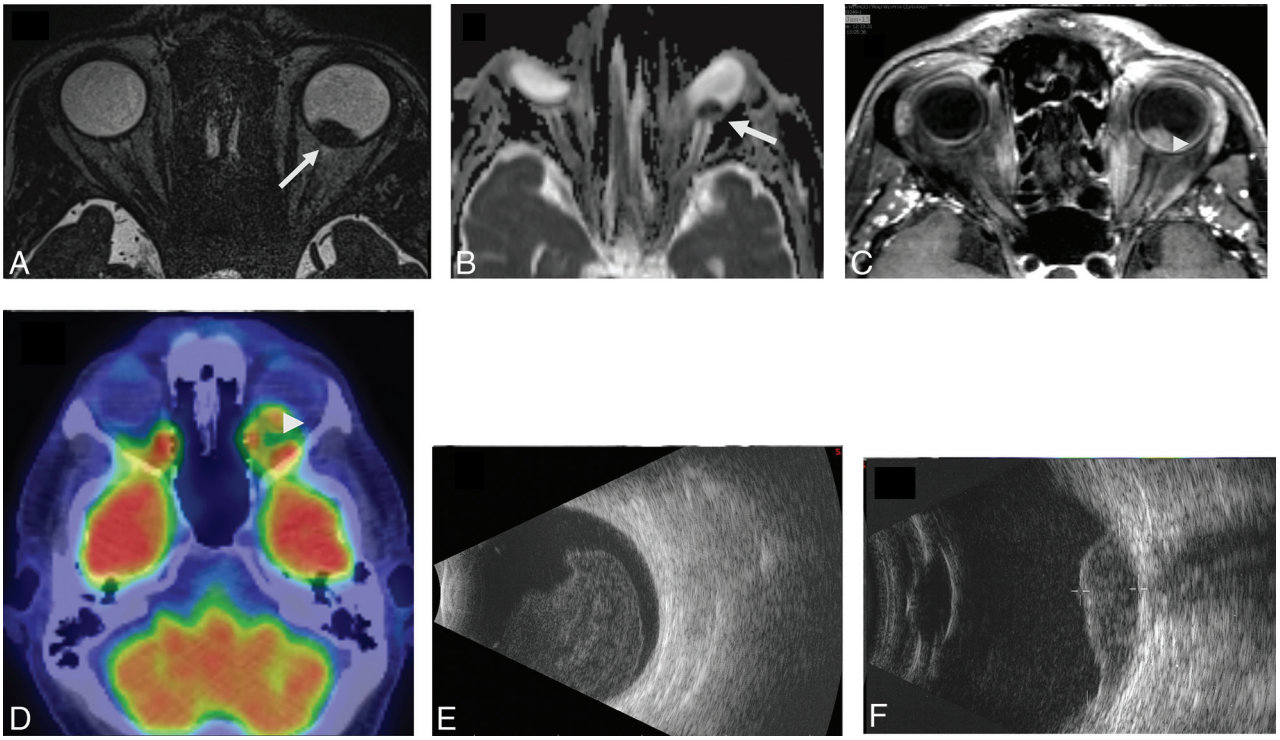

FIG 4. Axial T2-weighted (*A*) imaging demonstrates a chorio-retinal mass in the posterior central globe overlying the optic disc (*arrow*) with concomitant restriction and low apparent diffusion coefficient (*B*). The lesion appears homogeneously enhanced on postcontrast-T1 (*C*) and is FDG-avid on axial FDG-PET/CT (*D*) without retro-global extension. B-scan ultrasound (*E* and *F*) shows vitreous opacities (*E*) and a large lesion overlying the optic disc (*F*).

Investigative techniques often reveal distinctive patterns, such as hyper-reflective foci in posterior vitreous and creamy lesions with infiltration of the retina on OCT. FFA is often characterized by retinal pigment epithelial defects leading to a classic "leopard skin" pigmentation.[15,16] Ophthalmologic examination is generally followed by laboratory tests, radiologic imaging, and biopsies.

These 3 cases provide an opportunity to compare the extent of disease via cross-sectional imaging. The focus case exhibited an indolent growth pattern requiring years before the left eye-enhancing choroidal thickening was detected on MRI, whereas the first companion case demonstrated an obvious globe mass with noticeable growth over 2 months. Contrasted with the large intraocular growth, companion case 1 showed extension of PIOL through the sclera and invasion of the intraocular and extraocular areas; hence, the ciliary body thickening and episcleral extension. The subtle episcleral extension was evident on ultrasound and MRI but not on FDG-PET/CT due to the relatively small lesion size.

The findings in these cases show that the disease course varies widely and that timely diagnosis is critical. Currently, the time from symptom onset to cytopathological confirmation is nearly 14 months.[17] Patients with CLL are viewed as typically progressing slowly, but anywhere from 2%–10% of cases undergo Richter transformation, which is seen as a more aggressive malignancy.[18] As described in case 3, Richter syndrome affecting the eye is rare but has been reported with extrascleral extension.[19] Clinicians should have a high suspicion for intraocular lymphoma (IOL) in patients with CLL and new ocular inflammation.

Ocular lymphoma should be considered in cases of visual symptoms on cross-sectional imaging, especially when isolated ocular lesions are seen on brain MRI, as MRI can support clinical ophthalmologic findings. However, detailed MRI descriptions with images of intraocular lymphoma are limited, especially in cases without cerebral involvement. Due to the infrequency of PCNSL extensive imaging characterizations are challenging. Notably, among 2 studies involving approximately 100 patients, each dedicated to PIOL imaging, only 4 cases of isolated ocular lymphoma were documented.[7,10] Expected MRI patterns comprise restricted diffusion, iso-hypointense T2-weighted imaging, and avid enhancement. Lymphomas are considered a hypercellular malignancy and thus present with high signal on DWI and concurrent hypointensity on ADC.[6] Overall, however, it should be noted that these imaging findings are not pathognomonic for IOL, and imaging findings of IOL may mimic infections (uveitis, toxoplasmosis, syphilis, tuberculosis, viral retinitis, sarcoidosis, idiopathic uveitis, endophthalmitis), amelanotic melanoma, and metastasis. Recognizing isolated intraocular lymphoma is critical, as most cases (40%–92%) progress to CNS involvement in less than 30 months.[1,2,20] The mechanism underlying cerebral extension has not been proved, but theories include direct spread through the optic nerve, lymphatic spread, or simultaneous disease progression of concurrent PCNSL.[21]

A proposed IOL diagnostic algorithm by Lopes et al[22] highlights MRI's strength in assessing intraocular lymphoma to rule out CNS involvement. The diagnostic algorithm for IOL starts with a clinical examination and patient history. If IOL is suspected, MRI and lumbar puncture are used to check for CNS involvement and malignant cells in the CSF. Vitrectomy provides vitreous samples for further analysis. These tests confirm the

diagnosis and guide treatment plans like chemotherapy, radiation, or targeted therapies.[22] The utility of MRI is complemented by FDG-PET/CT. There are no large studies investigating the diagnostic role of FDG-PET/CT specific to PIOL. However, PET/CT is often used as disease staging for all lymphomas.[22,23] FDG-PET/CT allows for whole-body imaging, detection of increased metabolic activity before visible anatomic defects, and a precise anatomic overlay of the metabolic abnormalities. Furthermore, MRI provides valuable information for close follow-up, enabling the detection of delayed CNS progression and evaluation of therapeutic responses.[8,24] In the context of PIOL, the role of MRI has remained consistent over the past decades. However, considering that most imaging characterizations of PIOL were published in the early 2000s or focused on CNS involvement, the sensitivity and specificity of PIOL detection by using modern MRI scanners are unknown.[7,9,10] Despite this, the authors believe it is reasonable to anticipate these values to persist below those of a cytology work-up and eagerly anticipate future research developments in this area.

Cytology is the standard for diagnosis with a specificity of 98%–100%, albeit with a wide sensitivity range from 31%–87%.[25] Despite a heightened suspicion of PIOL, false-negatives do occur. Contributing factors include insufficient specimen volumes, extended delays from specimen collection to analysis (leading to apoptosis), or minimal neoplastic cells within the vitreous. Cytology, in concert with cytokine analysis, IHC, and flow cytometry, provides more thorough diagnostic information. Cytokine analysis, particularly the elevated interleukin-10 to interleukin-6 ratio ($>$ 1.0) within the vitreous, is associated with diffuse B-cell lymphoma.[25] However, this analysis is supplemental, as the ratio's accuracy can be influenced by corticosteroid treatment, which is often employed for the common misdiagnosis of uveitis. More recently, the diagnosis of PIOL involves detecting B-cell receptor clonality, myeloid differentiation primary response 88 gene (*MYD88*) mutations, and a B-cell lymphoma 2 translocation with polymerase chain reaction.[15,25]

Treatment approaches for PIOL are not standardized but rather tailored to the extent of disease. Hematologists/oncologists and ophthalmologists must work closely together for proper diagnosis. Unilateral cases typically involve vitrectomy and intraocular rituximab injection, with or without methotrexate. Bilateral involvement may necessitate a combination of intraocular injections, systemic chemotherapy, and radiation therapy.[5,15,26] The prognosis is closely linked to the presence or absence of CNS involvement. In a comprehensive meta-analysis involving 1484 patients, the 5-year survival rate was 97% for individuals lacking CNS involvement versus 54% for those with associated CNS pathology.[5]

CASE SUMMARY

PIOL is a rare diagnosis that has not been well characterized in the radiologic literature. The established MRI pattern follows the classic lymphoma description, namely, hypercellular lesions with diffusion restriction. Cross-sectional imaging, including PET/CT, is critical to determining disease staging, treatment planning, and response. Radiologic studies complement specialized ophthalmologic imaging and cytopathologic investigations, of which the latter remains the standard for diagnostic tests.

Disclosure forms provided by the authors are available with the full text and PDF of this article at www.ajnr.org.

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