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ORIGINAL ARTICLE

Intraocular T-cell Lymphoma: Clinical Presentation, Diagnosis, Treatment, and Outcome

Florence Chaput, MD[1], Radgonde Amer, MD[2], Edoardo Baglivo, MD[3], Valerie Touitou, MD, PhD[4], Alexandra Kozyreff, MD[5], Dominique Bron, MD, PhD[6], Bahram Bodaghi, MD, PhD[4], Phuc LeHoang, MD, PhD[4], Chris Bergstrom, MD[7], Hans E. Grossniklaus, MD[7], Chi-Chao Chan, MD[8], Jacob Pe'er, MD[2], and Laure E. Caspers, MD[1]

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ABSTRACT

Purpose: To report on the clinical data of seven patients with T-cell intraocular lymphoma (IOL).
Methods: Retrospective case series.
Results: Seven immunocompetent patients, 12 eyes, 6 women, with T-cell-IOL were included from five countries. Mean age was 53.5 years (range: 25–82). Four patients had systemic-ocular lymphoma, two had CNS-ocular lymphoma, and one had systemic-CNS- ocular lymphoma. Vitritis was the most frequent clinical sign, followed by anterior uveitis and serous retinal detachment. Cytopathologic examination was performed on all ocular specimens (vitreous in six and iris mass biopsy in one patient). Adjunctive diagnostic procedures included immunohistochemistry, molecular tests, and cytokine profiling of vitreous samples. Treatment modalities included systemic chemotherapy (five patients), intravitreal methotrexate (three patients), globe radiotherapy, and intrathecal chemotherapy. Mean survival from diagnosis was 21.7 months (range: 2–69). Two patients are still alive.
Conclusions: T-cell IOL has variable clinical manifestations and prognosis. Systemic involvement, SRD, and vitreoretinal involvement were frequently observed.

Keywords: Intraocular lymphoma, primary vitreoretinal lymphoma, T-lymphoma, uveitis

Intraocular lymphomas (IOLs) are heterogeneous malignant neoplasms. The most common group is constituted by vitreoretinal lymphomas, which are mostly high-grade B-cell malignancies (diffuse large B-cell lymphoma) with poor prognosis because of associated central nervous system (CNS) lymphomas. Other groups of IOL are less common. They include the primary choroidal lymphomas, which are typically low-grade B-cell lymphomas that are similar to mucosa associated lymphoid tissue, extranodal marginal zone B-cell lymphomas elsewhere in the body, and the secondary, metastatic, uveal lymphomas.[1] T-cell IOL is a very rare disease. Case reports and small series of T-cell-IOL have been previously reported.[2–28] Unlike metastatic B-cell IOL, which involves the uvea, metastatic T-cell IOL predominantly

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presents with vitreoretinal involvement.[26] The most frequently reported metastatic T-cell IOL is the cutaneous peripheral T-cell lymphoma (mainly the mycosis fungoides subtype), the NK-T cell lymphoma and the adult T-cell lymphoma/leukemia. The aim of this article is to report on the clinical features, diagnosis, treatment, and outcome of seven immunocompetent patients diagnosed with T-cell IOL.

MATERIALS AND METHODS

This retrospective study included seven patients from five tertiary referral centers in Belgium, Switzerland, France, Israel, and USA, treated between 2000 and 2014. Diagnosis of T-cell IOL was based on vitreous specimens, iris biopsy, and extrascleral masses. We collected data from ophthalmic, medical, and pathology charts, including: demographic data; clinical ophthalmologic findings (laterality, location, initial, and final visual acuity, VA – Snellen charts); initial ocular symptoms; duration of symptoms prior to treatment; type of ocular involvement; diagnostic modalities; tumor type; initial lymphoma location; ocular treatment and response to treatment; systemic therapy; response to treatment and adverse effects of treatment; length of survival; and duration of follow-up. Incomplete data from patients 1, 3, and 5 have already been reported.[21,26,28] but charts of these patients were retrieved and complementary data were collected and included in the present study.

RESULTS

Demographic and Clinical Features

Seven immunocompetent Caucasian patients (six women, one man) with a mean age of 54 years (range: 25–82) were included. Clinical data are detailed in Table 1.

Four patients (57.1%) had systemic lymphoma in addition to IOL. The organs affected included the skin in the form of mycosis fungoides in two patients (pnts 5, 7), bone marrow in one patient (pnt 3) and liver in one patient (pnt 6). Two patients (28.6%) had CNS lymphoma in addition to IOL (pnts 2, 4) and one patient (14.3%) had both systemic (adrenal gland) and CNS lymphoma in addition to IOL (pnt 1).

In two patients, the diagnosis of IOL was concomitant with the diagnosis of the systemic and/or CNS lymphoma (pnts 1, 3). Two patients presented with primary IOL (pnt 2 subsequently developed CNS lymphoma 3 years later and pnt 5 developed mycosis fungoides 8 months later). Three patients developed IOL subsequent to the diagnosis of the systemic or CNS lymphoma (pnts 4, 6, 7 at 4, 5 and 24 months later, respectively).

All patients presented with blurred vision, two patients in addition complained of ocular discomfort or pain, one patient had floaters, and one patient had eye redness.

IOL was bilateral in five patients (71%, pnts 3–7) and unilateral in two patients (29%, pnts 1, 2).

Initial VA ranged from 10/10 to no light perception. Most eyes preserved good vision except for four eyes that sustained profound visual loss (pnts 1, 2, 6, 7).

Six patients (86%) (10 eyes) presented with vitreous cells at the time of diagnosis and one patient (pnt 5, 1 eye) developed vitreous cells during vitreoretinal recurrence (Figure 1). Five patients (71%) (8 eyes) had anterior chamber cells masquerading as anterior uveitis: three patients (4 eyes) with non-granulomatous and two patients (4 eyes) with granulomatous anterior uveitis. Iris involvement occurred in one patient (pnt 5, 2 eyes). Marked cystoid macular edema (CME) was present in one patient (pnt 3, 2 eyes) (Figures 2 and 3),creamy white retinal lesions were described in four patients (57%) (7 eyes), and serous retinal detachment (SRD) was present in five patients (71%) (7 eyes). Optic neuropathy and retinal venous occlusion were both observed in one eye (pnts 1, 2). One patient had extrascleral extension into the orbit (pnt 1, 1 eye) and another patient with bilateral IOL had unilateral subconjunctival extension (pnt 6). The mean interval between the first symptoms and the beginning of the treatment was 8.5 months (range: 6 weeks to 29 months).

Diagnostic Techniques

The results of the diagnostic procedures performed are summarized in Table 2. All ocular samples (vitreous sample in six patients and iris mass biopsy in one patient, pnt 5) underwent cytopathologic analysis and anaplastic lymphoid cells were observed in all cases. In patient 3, they were small sized cells and in patient 7, they were pleomorphic cells, otherwise in the rest of the cases they were large lymphoid cells.

Immunohistochemistry was performed in six cases and it confirmed the presence of T-cell surface markers. Polymerase chain reaction (PCR) was performed in four cases (Figures 4–6)and it showed T-cell receptor (TCR) gene rearrangement. In patient 3, the same TCR gene rearrangement was detected in the bone marrow, blood, and vitreous and in patient 7, the same gene rearrangement was detected in the skin and vitreous. Flow cytometry was additionally performed on the vitreous sample of the latter patient. Cytokine profiling of the vitreous sample was performed in three patients and the IL-10:IL-6 ratio was performed in patients 1 and 2, and it was <1.

Figure 4 shows the lymphoid cells with large or small sizes and variable irregular nucleus, finely dispersed chromatin and inconspicuous nucleoli mixed with necrotic and degenerative cells and monoclonal T-cell gene rearrangement. Figure 5 shows the monoclonal gene rearrangement of patient 2.

TABLE 1. Clinical characteristics.

Pnt. no.	Gender	Age	Symptoms	Initial VA	Laterality	Clinical findings	Location at diagnosis	Chronology
1.	F	57	Acute visual loss	LE:LP	Unilateral	Anterior hypertensive uveitis Hypopyon, mild vitritis Creamy white retinal lesions Serous retinal detachment Optic neuropathy, secondary orbital infiltration	Adrenal gland, eye CNS (CSF)	Concomitant
2.	F	82	Acute visual loss	RE:LP	Unilateral	Anterior uveitis Vitritis Serous retinal detachment Retinal vein occlusion Vitreous hemorrhage	Eye was involved first CNS involvement developed 36 months later	PIOL
3.	F	71	Visual loss Pain Redness	RE:6/10 LE:5/10	Bilateral	Anterior uveitis Vitritis Cystoid macular edema Serous retinal detachment	Bone marrow and eye	Concomitant
4.	F	40	Blurred vision	RE:9/10 LE:10/10	Bilateral	Vitritis Creamy white retinal lesions	CNS (brain and meningeal involvement: pia mater and subarachnoid space)	CNS was involved first and eye involvement developed 4 months later
5.	M	31	Unilateral visual decrease Discomfort	LE:20/20 RE:20/60	Bilateral	Granulomatous anterior uveitis Involvement of iris Hypopyon Vitritis	Eye was involved first, skin involvement developed 8 months later	PIOL
6.	F	25	Not reported	RE:7/10 LE:LP+	Bilateral	Vitritis Creamy white retinal lesions Serous retinal detachment Secondary extrascleral unilateral subconjunctival infiltration	Liver	Liver was involved first and eye involvement developed 5 months later
7.	F	68	Blurred vision Floaters	RE:CF LE:20/70	Bilateral	Anterior uveitis Vitritis Serous + rhegmatogenous retinal detachment Creamy white retinal lesions Retinal pigment epithelial changes	Skin (mycosis fungoides)	Skin was involved first and eye involvement developed 2 years later

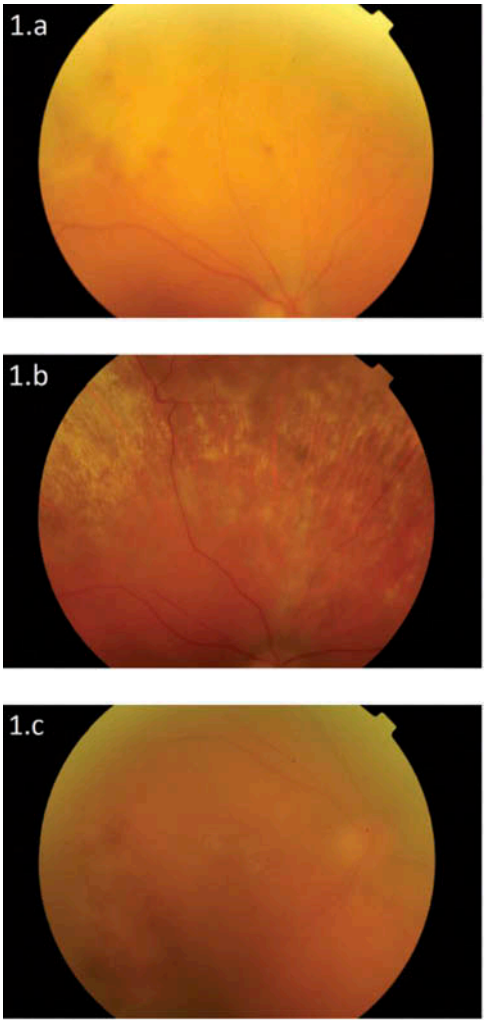

FIGURE 1 Patient 2 (a) initial retinal involvement with deep diffuse retinal infiltrates and few hemorrhages; (b) scarring of the retina 3 months after initiation of treatment; (c) recurrence in the form of diffuse deep retinal yellow-white lesions and hemorrhages 9 months later.

Therapeutic Modalities

The different treatment modalities and prognosis are reported in Table 3.

FIGURE 2. Patient 3 : Granulomatous keratic precipitates.

Five patients (71%) received systemic chemotherapy: MTX+CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisone) in two patients and MTX alone in two patients. In one patient (pnt 6) it was not reported, as it was performed in a different hospital. Patient 2 received only globe radiotherapy, as this patient had IOL for a period of 3 years before developing CNS lymphoma when she declined further treatment. Patient 7 received topical chemotherapy in the form of Targretin because of mycosis fungoides.

Ocular treatment consisted of either intravitreal MTX injections (400 µg/0.1 mL) in four patients or globe radiotherapy in four patients (at a dose of 30 Gy in two patients and 20 Gy in one patient, and planned for pnt 7). Patient 6 received globe radiation in another hospital and treatment was continued with intravitreal MTX.

Three patients responded fully to intravitreal treatment and two of them achieved remission of IOL following eight and nine injections (pnts 6 and 4, respectively). Patient 5 sustained ocular recurrence 2 weeks following complete regression of IOL. MTX intravitreal injections were discontinued in patient 3

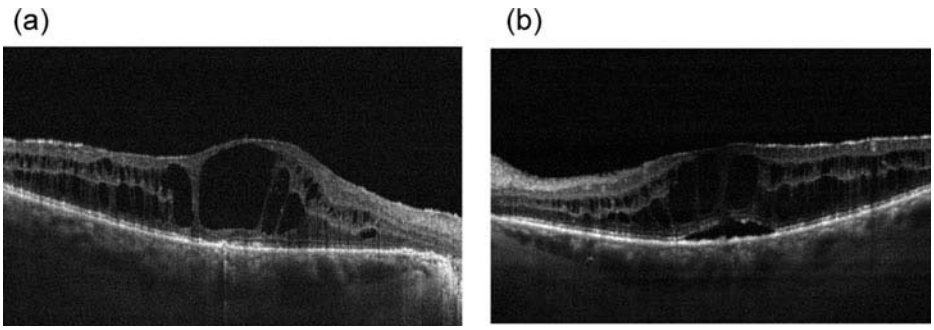

FIGURE 3. Patient 3: Spectral-domain optical coherence tomography showing bilateral cystoid macular edema and subretinal fluid in a patient with T-cell intraocular lymphoma secondary to bone marrow lymphoma.

TABLE 2. Diagnostic methods of T-cell lymphoma.

Pnt. no.	Histology of ocular specimen	Immunohistochemistry	PCR	ELISA
1.	Anaplastic large cell lymphoma: large anaplastic irregular multinucleated lymphoid cells, mitosis, granular basophilic cytoplasm	CD30+, ALK 1–, Alk P80–, CD56–, CD2+, CD 5–, CD3 3 –, CD7–	Polyclonal TCR gene rearrangement, (PCR EBV+, hybridization EBV (EBER) –)	IL-10: 493 pg/mL, IL-6: 1184 pg/mL Ratio IL-10:IL-6 = 0.4164
2.	Lymphoid cells and necrotic cells, poor morphology	–	TCRγ, TCR, CDR3	IL-10: 113.1 pg/mL, IL-6: 189.9 pg/mL Ratio IL-10:IL6 = 0.5956
3.	Small-sized lymphoid cells	CD3+, CD4 + CD8 +	Monoclonal TCR-β rearrangement	IL-10: 81.57 pg/mL
4.	Atypical lymphoid cells	CD3+ and leucocyte common antigen	Not done	Not done
5.	Large atypical lymphoid cells Apoptotic bodies, mitotic figures	CD3+ and leucocyte common antigen CD20–, CD79α–, CD30– Terminal deoxynucleotidyl transferase Myeloperoxidase Neuron specific antigen HMB-45	Not done	Not done
6.	Large anaplastic lymphoid cells, hyperchromatic nuclei, many mitosis	CD3+	Not done	Not done
7.	Pleomorphic lymphoid cells, large irregular nuclei, basophilic cytoplasm	CD4+, CD8+	Monoclonal TCR gene rearrangement	Not done

TCR, T-cell receptor.

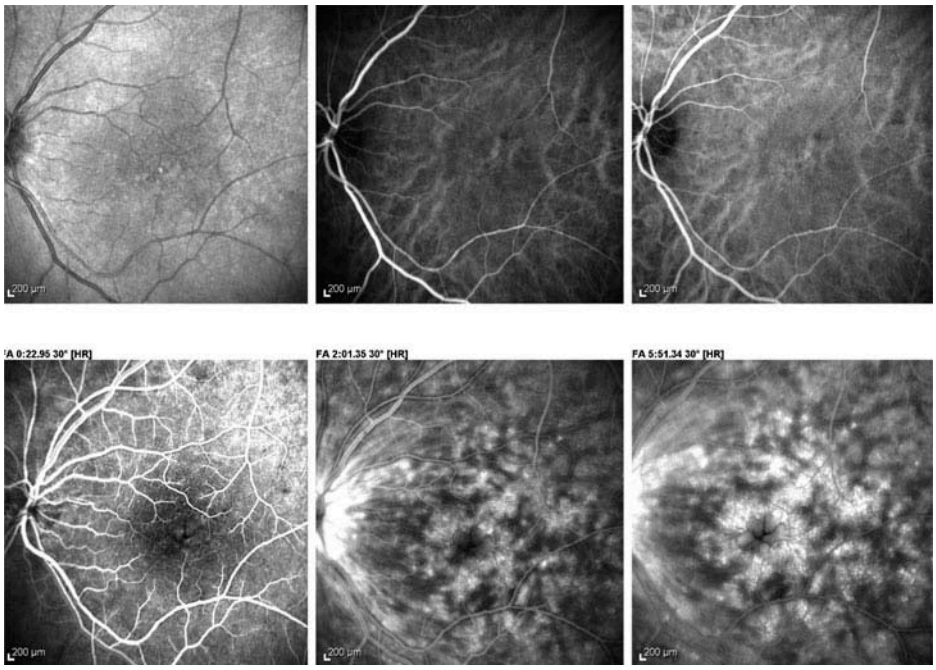

FIGURE 4. Patient 3: T-cell IOL secondary to bone marrow lymphoma. Red free photography (top left) .Indocyanine green angiography (ICGA) showing no choroidal involvement (top right). Fluorescein angiography (FA) (bottom): showing diffuse leakage of dye remaining after a good response of the retinal infiltration to the intravitreal injections of methotrexate.

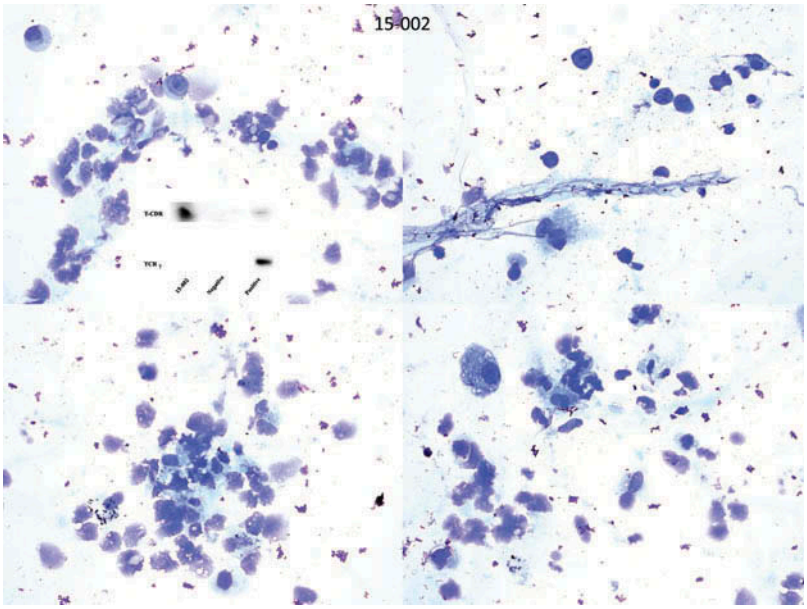

FIGURE 5. PCR performed on microdissection of vitreous cells of patient 2. TCR gene rearrangement using primer pairs TCRγ and TCR-CDR3. IgH gene rearrangement was not detected using primer pairs of FR3A, FR2A, and CDR3CDR.

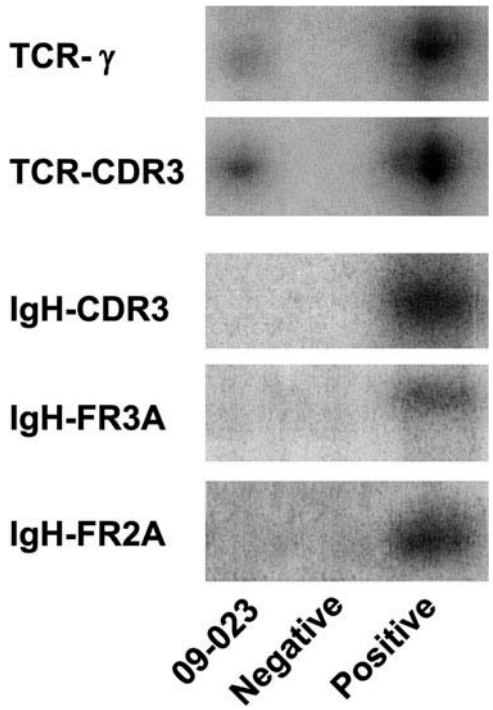

FIGURE 6. PCR were performed on microdissected atypical lymphoid cells from patient 6 using primers of TCR-CDR3, as well as three primers of IgH gene (CDR3, FR2A, and FR3A)

due to severe toxic keratopathy. The patient is still alive, with a persistent low activity in her eye and bone marrow. She is still treated with oral methotrexate (50 mg/week) in addition to repeated intravitreal injections of triamcinolone acetonide because of CME.

Patients 1 and 4 who also suffered from CNS lymphoma received intrathecal chemotherapy (methylprednisolone, MTX, cytarabine in pnt 1 and MTX in pnt 4). Patient 5, despite not suffering from CNS involvement, also received intrathecal MTX as prophylactic treatment.

The mean survival time from diagnosis of IOL was 21.7 months (range: 2–69 months). Recurrence of systemic or CNS lymphoma was the cause of death in four patients. Two patients are still alive (pnts 3 and 7, with a follow-up of 69 and 4 months, respectively). Patient 2 with unilateral primary intraocular lymphoma (PIOL) was in remission for 36 months when she developed CNS lymphoma but died from cardiac arrest.

DISCUSSION

In this study, we report the clinical presentation, diagnosis, treatment, and outcome of seven patients with T-cell IOL confirmed by ocular biopsy. Most of the clinical findings are similar between T-cell IOL and B-cell IOL, un-enabling differentiation between the two entities based on ocular signs.

All patients however suffered from vitreoretinal involvement. This is in contrast to metastatic B-cell lymphoma that usually presents with uveal involvement but similar to PVRL of the B-cell type. In our study, compared with the main T-cell IOL series,[26] we found a higher rate of anterior uveitis (71%) and subretinal/retinal infiltrates (57%), as in PVRL, and a lower rate of vasculitis/perivascular infiltrates.[26] We also found a very high rate of SRD (71%). We report four new cases of SRD while, to our knowledge, only three other cases have been previously reported on

TABLE 3. Treatment modalities.

Pnt. no.	Treatment		Ocular side-effects	Ocular remission/recurrence	Survival from diagnosis
1.	Systemic chemotherapy	Yes[a,b,c]		No response, no remission	2 months
	Intrathecal chemotherapy	Yes[a,b,c]			DOD
2.	Globe radiotherapy	Yes	None	Total remission	36 months
				No recurrence	DUC
3.	Oral chemotherapy	MTX			Still alive (69 months)
	IVT	MTX (>30 IVT)	Toxic keratitis	Partial response, no remission	AWD
4.	IV systemic chemotherapy	MTX		Remission	9 months
	Intrathecal chemotherapy	MTX		No local recurrence	DOD
	IVT	MTX	None		
5.	IV systemic chemotherapy	Yes[d,e]		Remission	18 months
	Intrathecal chemotherapy	MTX	Toxic keratitis	Recurrence: 2 weeks later	DOD
	IVT	MTX (twice weekly, 7 times)		Systemic relapse: 8 months later	
6.	Systemic chemotherapy	Yes (not reported)		Remission	7 months
	Globe radiotherapy	Yes		Recurrence: 2 months later	DOD
	IVT	MTX	Toxic keratitis Cataract		
7.	Local chemotherapy	Topical cutaneous Targretin and Clobetasol	None	Still active	Still alive (4 months)
	Globe radiotherapy	Yes			AWD

[a]IV: CHOP 14 = CHOP + MTX + IT: methylprednisolone, MTX, cytarabine.
[b]IV: cytarabine, asparaginase (1 course) + IT: cytarabine.
[c]IV: CYVE: cytarabine (J1–4), etoposide (J2–J5) + IT: cytarabine.
[d]IV: CHOP.
[e]IV: CHOP + MTX + leucovorin calcium + IT: MTX+ IVT: MTX 400 µg
DOD, Death of disease; AWD, alive with disease; DUC, death of unrelated cause; IV, intravenous; IVT, intravitreous; MTX, methotrexate; IT, intrathecal; CHOP, cyclophosphamide; doxorubicin; vincristine; prednisone.

T-cell IOL in the past[19,22,25] and two SRD cases were recently described on T-cell IOL.[29,30] A low rate of SRD was previously reported in B-cell lymphoma (10–28%).[5,31–38] Furthermore, CME is an unusual manifestation of IOL. Few cases were described in B-cell IOL[31,37,39] and T-cell IOL.[3] Severe bilateral CME was present in one of our patients. These differences in the occurrence of SRD and CME might be attributed to the use of optical coherence tomographic exams, which have enhanced our understanding of retinal pathologic conditions.

Four patterns have been reported in T-cell IOL: (1) PIOL: occurred in 29% of patients in our series compared with 14% in the T-cell review,[26] which did not differ from B-cell IOL (17–30%).[39–41] (2) Ocular-CNS lymphoma: occurred in 29% of our patients. It is reported in around 30% of patients in series of T-cell IOL,[26] and similarly in the series of B-cell IOL reported by Fardeau et al.[42] However, a much higher rate (50–85%) was found in other series of B-cell IOL (50–85%).[39–41,43,44] (3) Ocular-systemic lymphoma: occurred in four patients (57%); and (4) ocular-systemic-CNS lymphoma: seen in one patient. The high rate of systemic involvement (57%) is in accordance with the bigger series of Levy-Clarke et al. (69%).[26] Two of our patients (29%) had skin involvement (mycosis fungoides), which is similar to the series of T-cell IOL of Levy-Clarke in 27.6%.[26] All our patients with systemic lymphoma had vitreoretinal involvement and none had choroidal location. This is in accordance with the series of Levy-Clarke showing that T-cell IOL corresponding to metastasis from systemic T-cell lymphoma predominantly involves vitreous and retina.[26] This differs from B-cell IOL secondary to metastasis from systemic B-cell lymphoma, which mostly affects the choroid following hematogenous spread,[45] while vitreoretinal involvement is typically observed in CNS B-cell lymphoma associated with IOL (Table 4).[55] In our study, the diagnosis was based on cytopathologic analysis of the ocular specimen. Adjunctive modalities included immunochemistry, molecular tests, and cytokine profiling of the vitreous sample. The diagnosis of T-cell IOL is difficult, usually made by examination of vitreous specimen, and repeated vitreous biopsies are often needed.[5,11,44] Diagnostic cellular yield is superior with full pars plana vitrectomy compared with core vitreous biopsy.[56] The gold standard remains the cytopathologic examination. However,

TABLE 4. Comparison between the current study and other studies of T-cell IOL and B-cell IOL.

Clinical features	T-cell IOL (current study)	T-cell IOL (Levy-Clarke's study)	B-cell IOL (other studies)	
Bilateral involvement	71%	54%	60–90%	Freeman et al. 1987,[40] Coupland et al. 2009.[43]
Anterior uveitis	71%	45%	21–55.5%	Freeman et al. 1987,[40] Char et al. 1988,[5] Siegel et al. 1989,[31] Whitcup et al. 1993,[44] Velez et al. 2000,[46] Hoffman et al. 2003.[23]
Vitritis	86%	65%	50–100%	Freeman et al. 1987,[40] Char et al. 1988,[5] Siegel et al. 1989,[31] Whitcup et al. 1993,[44] Velez et al. 2000,[46] Hoffman et al. 2003,[23] Fardeau et al. 2009.[42]
Subretinal/retinal infiltrates	71%	28%	19–73%	Freeman et al. 1987,[40] Char et al. 1988,[5] Siegel et al. 1989,[31] Velez et al. 2000,[46] Fardeau et al. 2009.[42]
Serous retinal detachment	30%	10%	9–28.5%	Siegel et al. 1989,[31] Char et al. 1988,[5] Ursea et al. 1997,[47] Hoffman et al. 2003.[23]
Papillitis	14%	20%	11–18%	Velez et al. 2000,[46] Hoffman et al. 2003,[23] Fardeau et al. 2009.[42]
Macular edema	14%	3.5%	7–25%	Siegel et al. 1989,[31] Whitcup et al. 1993,[44] Fardeau et al. 2009.[42]
Iris involvement	14%	14%	3%	Velez et al. 2000.[46]
Vasculitis/perivascular infiltrates	0%	10%	7.5–18%	Whitcup et al. 1993,[44] Velez et al. 2000,[46] Hoffman et al. 2003,[23] Fardeau et al. 2009.[42]
Mean age (years)	53	57	53–63	Freeman et al. 1987,[40] Whitcup et al. 1993,[44] Surawicz et al. 1999,[48] Grimm et al. 2007,[49] Fardeau et al. 2009,[42] Hong et al. 2011.[50]
Mean survival	21.7 months	2 weeks to 101 months	20–58 months	Freeman et al. 1987,[40] DeAngelis et al. 1992,[51] Peterson et al. 1993,[52] Grimm et al. 2007,[49] Hong et al. 2011,[50] Kimura et al. 2012.[53]
PIOL	29%	14%	17–30%	Freeman et al. 1987,[40] Gill & Jampol 2001,[39] Mochizuki & Singh 2009.[41]
CNS involvement	29%	31%	33–85%	Freeman et al. 1987,[40] Whitcup et al. 1993,[44] Gill & Jampol 2001,[39] Mochizuki & Singh 2009,[41] Coupland et al. 2009,[43] Fardeau et al. 2009.[42]
Systemic involvement	57%	69%	9–22%	Freeman et al. 1987,[40] Mochizuki & Singh 2009,[41] Cao et al. 2011.[54]

IOL, intraocular lymphoma.

immunophenotyping, flow cytometry, and molecular analysis are helpful to improve the diagnostic accuracy.[57] Flow cytometry allows a rapid and objective evaluation of a larger number of cells than immunohistochemical staining. Flow cytometry was performed in only one of our patients. T-cell lymphomas fail to express specific marker that can be stained to indicate a monoclonal origin. In T-cell IOL, Davis[57] used several markers: CD2 (pan T-cell), CD3 (pan T-cell), CD4 (T helper/Inducer), CD5 (pan T-cell, some B-cell), CD7 (pan T-cell), CD8 (T suppressors-cytotoxic), and T-cell receptor (TCR, alpha and beta).[57] PCR has been used for years to confirm the diagnosis in B-cell lymphomas. It appears to be highly sensitive and has been adapted to T-cell lymphomas by detection of T-cell receptor gene rearrangements.[16,57] The diagnosis in four of our patients was confirmed by PCR. Moreover, a high level of interleukin-10 (IL-10) and high IL-10/IL-6 ratio support the diagnosis of B-cell IOL.[58,59] As IL-10 is a cytokine related to B-lymphocytes,[58] high level of IL-10 is not expected in the vitreous of patients with T-cell IOL. IL-10:IL-6 ratio was <1, as expected in two of our patients.

The mean survival after diagnosis of T-cell IOL in our patients was 21.7 months, with a range of 2–69 months. Levy-Clarke et al. also reported a variable rate of survival in T-cell IOL (ranging from 2 weeks to 101 months, mean not reported).[26] In vitreoretinal lymphomas, Kimura et al. found a mean survival of 32 months and a 5-year survival rate of 61% in Japan.[53] Overall survival duration was found to be highly variable in T-cell IOL but rarely more than 1 year. This might be related to the frequent systemic involvement in T-cell IOL. Metastatic T-cell IOL has poor prognosis. Indeed intravitreal MTX as a single modality of treatment of T-cell IOL appears less effective compared with the published results in B-cell PIOL.[60.]

The treatment of T-cell IOL is not well established. In our group of patients, systemic MTX ± CHOP was administered in five patients and intravitreal (MTX) chemotherapy in four patients. In accordance with Levy-Clarke et al.[26] we observed

poor and variable prognosis in our patients with only one patient achieving complete remission and one patient with long survival over 5 years after the diagnosis of the IOL. The extension of the systemic lymphoma was the cause of death in four patients within 18 months after diagnosis. Three of our patients had ocular radiotherapy. In Levy-Clarke's review of T-cell IOL, radiotherapy was the first choice of treatment, usually combined with chemotherapy. B-cell primary vitreoretinal lymphomas are highly chemo- and radiosensitive. However, radiotherapy had minimal impact on the overall survival because CNS relapse occurs in almost all patients with median survival of 12–20 months, limiting the 5-year overall survival rate after radiotherapy to 10–29%.[52.]

In conclusion, T-cell IOL is a very rare entity and its diagnosis remains difficult. In our patients, clinical presentation and prognosis were highly variable. SRD and systemic involvement were observed in the majority of our patients. All but one patient had presented with vitreoretinal involvement, while none of them had choroidal involvement, even when the lymphoma was secondary to systemic T-cell lymphoma. The diagnosis of T-cell IOL is more difficult than B-cell IOL and no guidelines exist regarding the treatment of these rare cases of T-cell IOL. An international registry is needed to enhance our knowledge of T-cell IOL.

DECLARATION OF INTEREST

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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