Systemic diffuse large B-cell lymphoma with bilateral ciliary body involvement

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To the Editor: Primary intraocular lymphoma (PIOL) is rare with an approximate incidence of 4.8/million and about 300 new cases annually in the United States.[1] Compared with PIOL, the prevalence of secondary intraocular lymphoma (SIOL) is significantly lower. Spreading to the eye from a distant site via the circulation, SIOLs occur within the uvea, with most cases confined to the choroid. Ciliary body involvement is rare in SIOL, while secondary diffuse large B-cell lymphoma with bilateral ciliary body involvement has, to our knowledge, been reported rarely.

A 54-year-old man presented to the Department of Ophthalmology at Peking Union Medical College Hospital on July 15, 2019 with redness, pain, and blurred vision in both eyes. During the previous 3 months, he noticed progressively worsening vision in both eyes, which was diagnosed as bilateral anterior uveitis and secondary glaucoma. He was given 1% prednisolone eye drops, 2% carteolol hydrochloride eye drops, and 1% brinzolamide eye drops. Unfortunately, both eyes further deteriorated, and the patient presented with hypopyon and increased ocular pressure (IOP) 3 months later. Based on immunohistochemical analysis of an axillary lymph node biopsy, he was diagnosed with systemic diffuse large B-cell lymphoma and treated with six courses of chemotherapy (standard rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone). On admission, his best-corrected visual acuity (BCVA) was 20/100 in the right eye and 20/200 in the left, while intraocular pressure was 42 mmHg (1 mmHg = 0.133 kPa) in the right eye and 45 mmHg in the left. Both eyes showed severe anterior chamber inflammation with 3+ large keratic precipitates, 1+ flare, 4+ cells, and a pseudohypopyon in the inferior angle [Figure 1A and 1B]; the bilateral peripheral anterior chamber was obliterated and the iris appeared thickened with nodular areas of infiltration; dilation examination revealed invisible fundus. B-scan ultrasound demonstrated a clear vitreous cavity and absence of retino-choroidal anomalies [Figure 1C and 1D]. Ultrasound biomicroscopy (UBM) revealed 360° thickening of the iris and ciliary body, plus peripheral angle closure [Figure 1E and 1F]. Paracentesis of the anterior chamber and diagnostic vitrectomy were performed; cytology of aqueous and vitreous specimens revealed atypical lymphoid cells [Figure 1G], shown to be IgK and IgH on gene rearrangement analysis. Brain magnetic resonance imaging was insignificant. Given his medical history, secondary ciliary body lymphoma (diffuse large B-cell) was diagnosed. Both eyes were treated with intravitreal methotrexate (MTX) at a dose of 0.4 mg in 0.1 mL twice a week for 4 weeks, weekly for 4 weeks, and monthly for 10 months. After single MTX injection in each eye, the BCVA improved to 20/63 in the right and 20/100 in the left, intraocular pressure decreased to 13 mmHg in the right and 19 mmHg in the left, and the pseudohypopyon disappeared [Figure 1H and 1I]. The patient continued with systemic lymphoma chemotherapy.

Reported cases with ciliary body involvement usually present with mild anterior inflammation.[2] However, Ahmed *et al*[3] proposed that anterior reaction and keratic precipitates may be present, especially in SIOL cases. Consistent with previous reports, the patient presented with an intense anterior segment reaction and a pseudohypopyon in the inferior angle. The patient denied oral ulcers, vulval ulcers, and rashes, which can be distinguished from Behcet's uveitis. Secondary glaucoma is also a common clinical finding in patients with intraocular lymphoma.[4] We believe the dense tumor infiltration observed in the chamber angle, as well as thickened iris and ciliary body, may have been the main causes of secondary glaucoma. After one intravitreal injection of MTX in each eye, intraocular pressure was controlled.

UBM of the anterior segment is helpful for detecting iris and ciliary body. It can produce high-resolution images of

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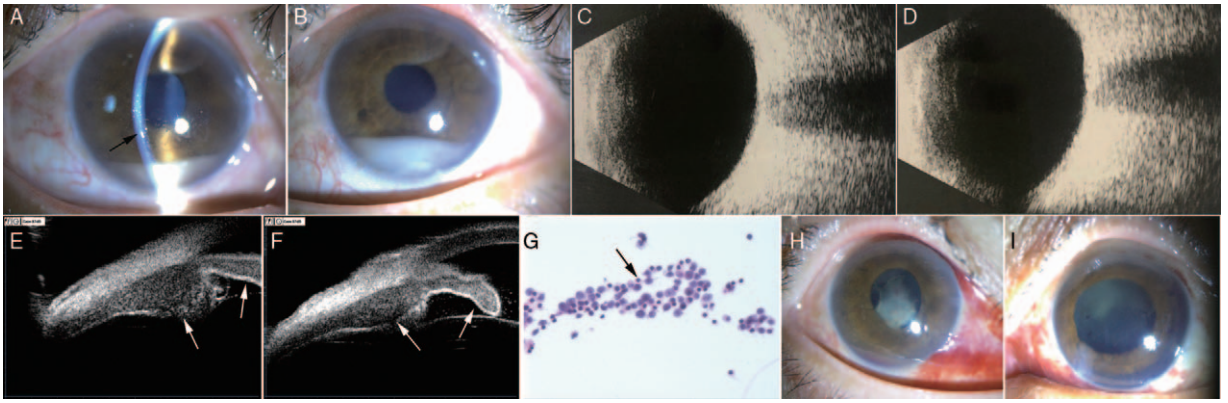

Figure 1: Representative image of the patient. (A and B) Anterior segment photography of both eyes showing severe anterior chamber inflammation with 3+ large keratic precipitates (black arrow), 1+ flare, 4+ cells, and a pseudohypopyon in the inferior angle. (C and D) B-scan ultrasound of both eyes demonstrated a clear vitreous cavity and an absence of retino-choroidal anomalies. (E and F) Ultrasound biomicroscopy of both eyes showing thickening of the iris and adjacent ciliary body with acoustic solidity but low internal reflectivity (white arrows). (G) Liquid-based cytology test of the aqueous and vitreous humor showing large atypical lymphoid cells (black arrow, hematoxylin and eosin, original magnification ×40). (H and I) Anterior segment photography of both eyes showing disappearance of pseudohypopyon, right eye with nasal subconjunctival hemorrhage, and pupillary organization membrane.

the ciliary body, providing visualization of tumor location and accurate tumor measurements.[2] In the patient, UBM revealed 360° thickening of the iris and ciliary body with acoustic solidity but low internal reflectivity in both eyes, a useful finding for differentiating anterior uveal lymphoma from uveitis.

The presence of tumor cells in anterior uveal structures makes it possible to achieve a diagnosis of intraocular lymphoma. However, aqueous samples are frequently non-confirmatory due to scant sample cellularity.[5] In this case, paracentesis of the anterior chamber and diagnostic vitrectomy were performed at the same time in his left eye to improve the diagnostic rate; intraocular involvement of diffuse large B-cell lymphoma was confirmed by liquid-based cytology and gene rearrangement analysis of aqueous and vitreous humor cells.

In conclusion, ciliary body recurrence of lymphoma should be considered in SIOL patients presenting with pseudohypopyon and elevated IOP accompanied by systemic lymphoma. Vitreous or tissue biopsy should be considered as it is the gold standard for intraocular lymphoma diagnosis. In addition, chemotherapy or radiotherapy regimens might preserve visual acuity, while the use of multiple-disciplinary teams could provide ciliary body lymphoma patients more individualized treatment.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given their consent for their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Conflicts of interest

None.

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