CASE REPORT

Case of Orbital Mucosa-Associated Lymphoid Tissue Lymphoma Associated with Serous Retinal Detachment

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ABSTRACT

We present a case of orbital mucosa-associated lymphoid tissue (MALT) lymphoma accompanied by a serous retinal detachment (SRD). A 44-year-old man who developed uveitis accompanied by a SRD in the right eye was referred to the Chiba National Hospital. B-mode ultrasonography and magnetic resonance imaging (MRI) showed a thickened choroidal membrane and a retrobulbar mass. Although topical steroid treatments improved the inflammation, the SRD did not improve. He was then referred to the Chiba University Hospital. Two months later, the retrobulbar mass had increased, and a partial resection biopsy combined with flow cytometry was performed. The final diagnosis was MALT lymphoma. After R-CHOP (rituximab–cyclophosphamide, doxorubicin, vincristine, and prednisolone) chemotherapy, signs of the tumour could not be detected and the SRD was reduced. We conclude that the SRD was most likely due to the infiltration of lymphoma cells and also to choroidal circulatory disturbances. Retrobulbar malignant lymphomas associated with a SRD are very rare and difficult to diagnosis without biopsy, but because early diagnosis is helpful in deciding the appropriate therapy, we recommend partial biopsy.

Keywords: Chemotherapy, mucosa-associated lymphoid tissue lymphoma, retrobulbar, serous retinal detachment

INTRODUCTION

Orbital mucosa-associated lymphoid tissue (MALT) lymphomas accompanied by a serous retinal detachment (SRD) are very rare, and only a few cases have been reported. Panda et al. reported a single case of orbital MALT lymphoma with a SRD that presented as panuveitis.¹ Gaucher et al. presented three cases of ocular MALT lymphoma associated with the secondary uveal effusion syndrome or panuveitis accompanied by a SRD.² We present a case of orbital MALT lymphoma accompanied by a SRD that was refractory to steroids.

CASE REPORT

A 44-year-old man who developed uveitis in his right eye was referred to the Chiba National Hospital. The onset of the uveitis was uncertain. His visual acuity was 20/40 OD and 20/20 OS, and the intraocular pressure (IOP) was 32 mm Hg OD and 15 mm Hg OS. Keratic precipitates and inflammatory cells were observed in the anterior chamber. Fundus examinations showed a serous retinal detachment (SRD) and a hyperaemic disc but no vitreous opacities (Figure 1). Optical coherence tomography (OCT) showed a SRD, and B-mode ultrasonography showed a thickened

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FIGURE 1 Initial fundus findings in a patient with a retrobulbar MALT lymphoma. (A) Fundus photograph (upper panel) and optical coherence tomographic (OCT) image (lower panel) show a serous retinal detachment (SRD). (B) B-mode ultrasonography shows a thickening of the choroidal membrane. (C) Fluorescein angiogram (upper panels) and indocyanine green angiogram (lower panels) show hyperfluorescence in the region of the SRD.

choroidal membrane and a retrobulbar mass (Figure 1). Fluorescein angiography and indocyanine green angiography showed hyperfluorescence at the area of the SRD (Figure 1). The magnetic resonance imaging (MRI) findings showed that the mass had T1- and T2-low intensities (Figure 2). Laboratory investigations were unremarkable, although topical steroids improved the degree of inflammation, but the SRD was not improved. Because the oral steroids did not reduce the size of the retrobulbar mass, he was referred to the Chiba University Hospital.

It was difficult to perform surgical biopsy at the first visit to Chiba University Hospital because of the location of the mass. Because the intraocular inflammation had already improved at the time of the first visit to our hospital and no vitreous opacities were found, we did not perform intraorbital surgical biopsy but only examined intraocular cytokines in the anterior chamber. The intraocular interleukin (IL)-10 was not detectable. The SRD most likely developed because of choroidal circulatory disturbances. Thus,

we had expected that it would take a long time to resolve the SRD and continued the oral steroid therapies as an idiopathic orbital inflammatory syndrome accompanied with a SRD. However, 2 months later, MRI performed in the Chiba University Hospital showed that the retrobulbar mass had increased despite the continued oral steroid treatment, and a malignant lymphoma was suspected (Figure 2). But because of the increased size of the retrobulbar mass, a transconjunctival surgical biopsy could be performed. A partial resection biopsy was performed, and histopathological examinations identified small-to moderate-size atypical lymphoid cells. The cells were CD20-immunopositive but were CD3-, CD5-, and CD10-negative (Figure 3). Flow cytometry showed a light chain restriction, and an immunoglobulin H (IgH) gene rearrangement that was confirmed by Southern blot analysis. Fluorescence in situ hybridization indicated a chromosomal break at the MALT1 gene locus. Positron emission tomography excluded metastasis but showed a strong

FIGURE 2 Magnetic resonance image (MRI) before chemotherapy in a patient with a retrobulbar MALT lymphoma. (A) T1-low (left panel) and T2-low (right panel) retrobulbar mass can be seen. (B) Two months later, the size of the retrobulbar mass increased and a transconjunctival surgical biopsy could be performed.

FIGURE 3 Representative histopathological findings of the surgical specimen of the retrobulbar mass. H-E staining identified atypical small- to moderate-size lymphocytes in the tissue ($\times 200$). Immunohistochemical findings indicate that these cells are CD20-positive but CD5- and CD10-negative ($\times 200$).

FIGURE 4 Fundus photograph (A), CT image (B), and an OCT image (C) after chemotherapy. Six months after chemotherapy, the SRD is reduced, with residual secondary retinal degeneration (A and C). A CT image shows that the retrobulbar lymphoma is absent (B).

accumulation of signal in the retrobulbar tumour. Taken together, the final diagnosis was primary orbital retrobulbar MALT lymphoma.

The patient underwent eight sessions of R-CHOP (rituximab–cyclophosphamide, doxorubicin, vincristine, and prednisolone) chemotherapy with a “cool off” protocol. Six months later, the SRD improved, and 18 months later the tumour was completely absent (Figure 4). Although the optic disc was not pale, the retina had degenerated secondarily and the cataract had progressed. Thus, the final visual acuity was 20/400. The IOP was 11 mm Hg without any glaucoma medications. Neither ocular inflammation nor recurrence was observed after the chemotherapy.

DISCUSSION

The clinical, histopathological, and imaging findings indicated that the mass was a retrobulbar MALT lymphoma accompanied by a SRD. The mass was refractory to topical steroids. A thickening of the choroidal membrane and retinal and choroidal

hyperpermeability to fluorescein and indocyanine green angiography suggested that there was not only lymphoma cell infiltration but also choroidal circulatory disturbances that led to the SRD.

Recent studies have shown that eyes with an idiopathic orbital inflammatory syndrome accompanied with a SRD were successfully treated with oral corticosteroids.^{3,4} Because surgery is difficult to perform on a retrobulbar tumour surrounding the optic nerve, our patient had oral corticosteroid treatment as an idiopathic orbital inflammatory syndrome accompanied with SRD. Because the intraocular IL-10 was not detectable and the vitreous was clear, the masquerade syndrome due to malignant lymphoma was not suspected at first, although it was refractory to the steroids.

After the diagnosis of MALT lymphoma, chemotherapy was selected because radiotherapy had a risk of relapse and some large cells in the biopsy specimens were Ki-67-positive, suspected of being caused by large cell transformation. After the R-CHOP therapy, no recurrence was observed. However, the retina was secondarily degenerated because the extensive SRD had persisted for a long time.

In conclusion, retrobulbar MALT lymphomas accompanied by refractory SRD are very rare and difficult to diagnose without biopsy. But early diagnosis is helpful in deciding the appropriate therapy, and if possible, we recommend partial biopsy of the mass.

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Note: Figures 1, 3, and 4 of this article are available in colour online at <http://informahealthcare.com/oph>.

REFERENCES

- [1] Panda P, Forooghian F, Goodglick T, Chan CC, Nussenblatt R, Sen HN. Orbital lymphoma masquerading as panuveitis. *Ocul Immunol Inflamm* 2010;18:181–183.
- [2] Gaucher D, Bodaghi B, Charlotte F, Schneider C, Cassoux N, Lemaitre C, Leblond V, Rao N, LeHoang P. MALT-type B-cell lymphoma masquerading as scleritis or posterior uveitis. *J Fr Ophthalmol* 2005;28:31–38.
- [3] Slagle WS, Boothe KH, Musick AN, Eckermann DR, Slagle AM. Idiopathic orbital inflammatory syndrome without pain: a case report. *Optometry* 2010;81:146–152.
- [4] Saeed F. Nonsimultaneous bilateral nonrhegmatogenous retinal detachments presenting as a complication of panuveitis in the setting of idiopathic orbital inflammatory syndrome. *Optometry* 2011;82:728–738.