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CASE REPORT

Primary central nervous system lymphoma presenting as a single choroidal lesion mimicking metastasis: A case report

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Abstract

BACKGROUND

Primary choroidal lymphoma is usually an indolent B-cell lymphoma and rarely progresses to extraocular sites. Herein, we report a case of primary choroidal lymphoma diagnosed as diffuse large B-cell lymphoma (DLBL), which progressed to the brain parenchyma after 4 mo.

CASE SUMMARY

A 78-year-old man presented with diminution of vision in his right eye. A choroidal lesion suspected of metastatic lesion was observed in the right eye by ophthalmologic examination. To discover the primary tumor, imaging investigations were performed but no malignant lesion was detected. After 4 mo, the patient returned to the clinic presenting with neurological symptoms. Brain magnetic resonance imaging revealed an abnormal contrast-enhancing mass in the left cerebellum. A stereotactic biopsy was performed, and DLBL was confirmed. The patient received the high dose methotrexate-based chemotherapy and he achieved complete remission.

CONCLUSION

Primary choroidal lymphoma is usually known to have a benign clinical course without systemic involvement. We present a rare case of primary choroidal lymphoma diagnosed as DLBL that progressed to the brain parenchyma within months.

Key Words: Primary choroidal lymphoma; Diffuse large B-cell lymphoma; Primary central nervous system lymphoma; Case report

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Core Tip: Primary choroidal lymphoma is a rare subset of primary intraocular lymphoma and shows a benign clinical course with no systemic involvement. Our case report involves a primary choroidal lymphoma demonstrating rare extraocular progression within months.

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INTRODUCTION

Primary intraocular lymphoma (PIOL) is a rare subset of primary central nervous system lymphoma (PCNSL) and includes vitreoretinal, choroidal, and iridal lymphomas [1]. Among PIOLs, primary choroidal lymphoma is a very rare disease, and only a small number of cases have been reported in the literature. Previously reported primary choroidal lymphomas were usually low-grade B-cell lymphomas, which rarely progressed to the central nervous system[2,3]. Herein, we report a very rare case of primary choroidal lymphoma diagnosed as diffuse large B-cell lymphoma (DLBL), which initially presented as a unilateral choroidal lesion mimicking metastasis.

CASE PRESENTATION

Chief complaints

A 78-year-old male came to the hospital presenting with a diminution of vision in his right eye.

History of present illness

His blurred vision had been aggravated for several weeks.

History of past illness

The patient had no relevant medical history, such as that of diabetes mellitus or hypertension. In the past, the patient had undergone short-term treatment for occupational pneumoconiosis resulting from his work as a coal miner for > 10 years.

Personal and family history

No special history of personal and family.

Physical examination

An ophthalmological examination revealed exudative subretinal detachment with a choroidal lesion suspicious of metastasis in the right eye (Figure 1A).

Laboratory examinations

Lactate dehydrogenase was moderately elevated at 387 U/L (normal range: 119-229 U/L).

Imaging examinations

To identify the primary tumor, the patient was referred to the Department of Hemato-oncology at our institute. Brain magnetic resonance imaging (MRI) (Figure 2A) and computed tomography (CT) of the neck, chest, and abdomen-pelvis showed no malignant lesions.

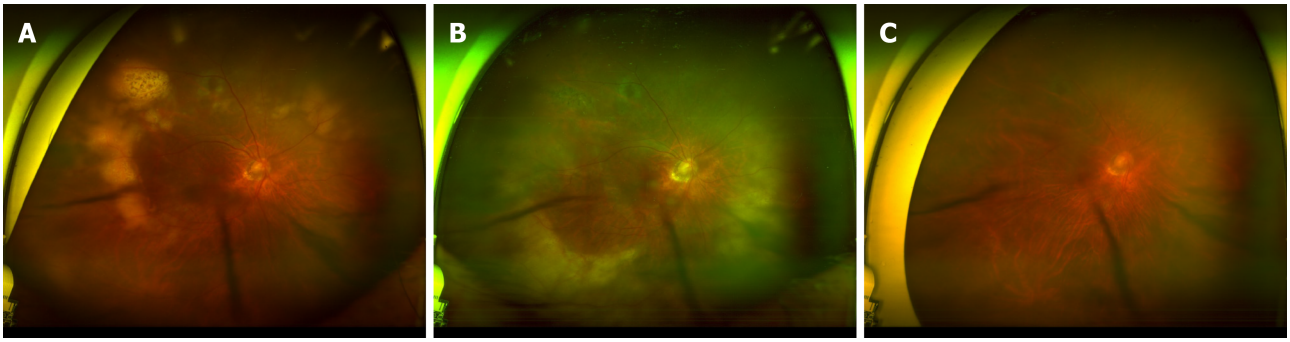

Figure 1 Course of ophthalmoscopy examinations. A: Image showing multifocal, creamy yellow, subretinal infiltrates during the first clinical visit; B: Image showing the spread of the creamy yellow, subretinal infiltrates toward the mid-periphery of the fundus. The image was taken 4 mo after the first clinical visit; C: Subretinal infiltrates appear to be markedly decreased after six cycles of systemic chemotherapy.

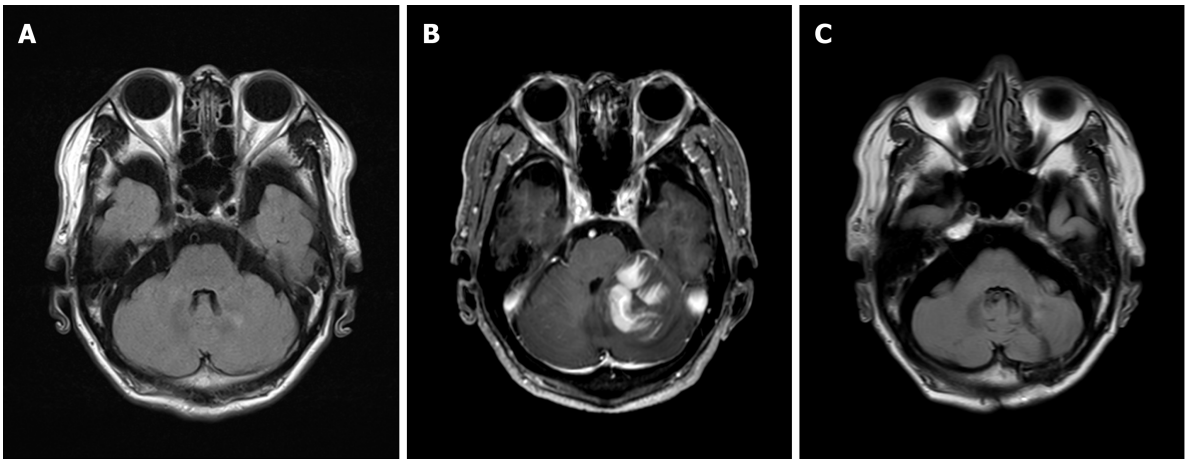

Figure 2 Findings of brain magnetic resonance imaging. A: Image showing no evidence of space-occupying intracranial or diffusion-restricted lesions. The image was taken at the first clinical visit; B: Image showing an irregular contrast-enhancing lesion, measuring approximately 3.8 cm × 3.8 cm in size, with diffusion restriction in the left cerebellum. The image was taken 4 mon after the first clinical visit; C: Image showing no evidence of abnormal contrast-enhancing lesion in the cerebellum after six cycles of systemic chemotherapy.

FINAL DIAGNOSIS

Four months later, the patient revisited the clinic because of dysarthria, headache, and right-sided weakness. No apparent aggravation of visual acuity was observed. The same findings of ophthalmoscopy as at the first visit were confirmed (Figure 1B). Brain MRI showed a single, irregular, contrast-enhancing mass in the left cerebellum (Figure 2B). The patient underwent a stereotactic biopsy of the mass and was diagnosed with DLBL (Figure 3). Further imaging, including CT and positron emission tomography/CT, revealed no systemic involvement. There was no evidence of lymphoma involvement in the cerebrospinal fluid and bone marrow.

TREATMENT

The patient received high-dose methotrexate (HD-MTX)-based chemotherapy, which comprised methotrexate (3.5 g/m^2), vincristine (1.4 mg/m^2, capped at 2 mg), and prednisolone (100 mg/d), with the addition of procarbazine (100 mg/m^2) in the first, third, and fifth cycles. Intrathecal injection was not given in this patient because there was no evidence of lymphoma involvement in the CSF test before and after the first cycle of HD-MTX based chemotherapy.

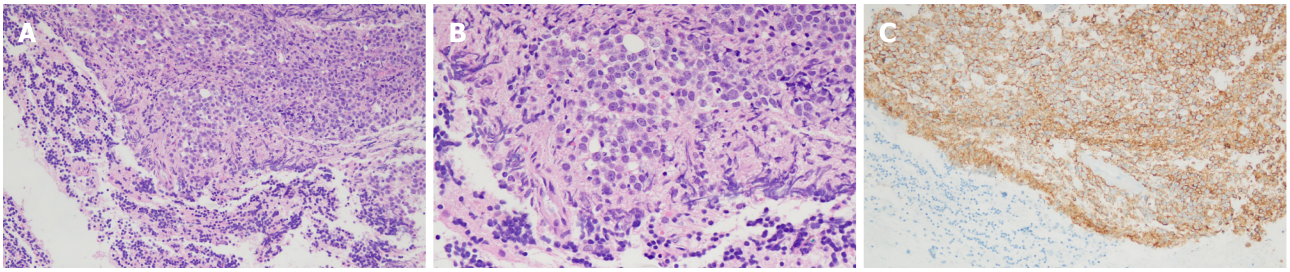

Figure 3 Representative microscopic images of diffuse large B-cell lymphoma in the brain. A: Infiltration of large, atypical cells with prominent nucleoli around the granular layer of the cerebellum (hematoxylin and eosin stain, magnification: 200 ×); B: Infiltration of large, atypical cells with prominent nucleoli around the granular layer of the cerebellum (hematoxylin and eosin stain, magnification: 400 ×); C: Tumor cells show strong positivity for the B-cell surface marker CD20 on immunohistochemistry (Immunohistochemical stain, magnification: 200 ×).

OUTCOME AND FOLLOW-UP

After completing six cycles of HD-MTX-based chemotherapy, the patient's neurological symptoms, such as right-sided weakness, dysarthria, and blurred vision, markedly improved. The abnormal, contrast-enhancing mass in the left cerebellum disappeared on brain MRI (Figure 2C), and the creamy yellow, subretinal infiltrates were markedly decreased (Figure 1C); these findings were compatible with complete remission. Maintenance therapy was recommended, but the patient and caregiver refused the treatment due to concerns about the side effects of additional chemotherapy and radiotherapy. The patient and his main caregiver were seriously concerned about the risk of cognitive decline that can be caused by additional treatments. Complete remission was maintained for approximately 17 mo without the maintenance chemotherapy.

DISCUSSION

Intraocular lymphoma is extremely rare and accounts for approximately 1.86% of intraocular malignancies[4]. Intraocular lymphoma is a heterogeneous group of malignancies located in different tissues, including the vitreous, retina, choroid, ciliary body, and iris, within the eye. It also refers to forms of primary or secondary to central nervous system lymphoma or disseminated systemic disease[3,5].

Primary intraocular lymphoma is considered a subset of PCNSL, and it progresses to the central nervous system in 15%-25% of PCNSL cases[3]. Primary vitreoretinal lymphoma is the most common intraocular lymphoma, followed by uveal lymphoma.

Choroidal lymphoma is a subset of uveal lymphoma. It can be subdivided into primary and secondary lymphoma based on the presence of systemic lymphoma at the time of ocular presentation. Primary choroidal lymphomas are defined as the absence of prior systemic lymphomas or concurrent extraocular lymphomas [6]. Several studies have reported clinical differences between primary and secondary choroidal lymphomas[2,7].

Primary choroidal lymphomas are mainly low grade B-cell lymphomas such as extranodal marginal zone B-cell lymphoma, are usually unilateral, and typically do not progress to the central nervous system parenchyma. Secondary choroidal lymphoma is characterized by the presence of previously known cancer or concurrent systemic lymphomas at the initial ocular presentation.

In contrast to primary choroidal lymphomas, secondary choroidal lymphomas are more likely to demonstrate bilateral involvement and preexistent lymphomas. More than half of secondary choroidal lymphomas have been confirmed as high-grade B-cell lymphomas, such as DLBL[2,8].

Unlike the previously reported cases of primary choroidal lymphoma, this case was characterized by the pathological findings of DLBL and disease progression to the brain parenchyma within a few months. In most of the previous cases of primary choroidal lymphoma, management involved local treatment or observation, whereas in our case, HD-MTX-based chemotherapy was administered, and the treatment response was complete remission.

CONCLUSION

Primary choroidal lymphoma is generally known to have a benign clinical course without systemic involvement. We reported a rare case of primary choroidal lymphoma diagnosed as DLBL, characterized by an aggressive clinical course that progressed to the brain parenchyma within a few months.

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