Rare disease

Unilateral isolated extraocular muscle lymphoma

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Summary

The authors would like to present an unusual case of ocular adnexal, mucosa-associated lymphoid tissue lymphoma, isolated to a single extraocular muscle. A 59-year-old woman presented with a 3-month history of slowly progressive double vision, worse on elevation, for which her optometrist had given her prisms. A swollen left upper eyelid was present for 10 days. CT scan of the brain and orbits revealed a 3 cm × 1.5 cm mass arising from the region of the left superior rectus with no signs of bone erosion. Histology showed infiltration by small lymphoid cells. Stage 1AE low-grade marginal zone B cell lymphoma was diagnosed. Possible aetiologies included *Chlamydia psittaci* infection and the recently recognised IgG4-related sclerosing disease. After oral doxycycline 200 mg once a day failed to show improvement, localised radiotherapy 30 Gy resulted in excellent clinical and radiological resolution of this isolated lymphoma.

BACKGROUND

Isolated extraocular muscle infiltration by mucosa-associated lymphoid tissue (MALT) lymphoma is a rare, potentially life-threatening condition. Early diagnosis enables successful treatment, hence the importance of recognising potential presentations and conducting appropriate investigations. It has also been linked to the newly recognised IgG4-related sclerosing disease, a condition that warrants the attention of clinicians in all areas of medicine.

CASE PRESENTATION

A 59-year-old woman admitted to the ophthalmology department with a 3-month history of slowly progressive double vision, worse on elevation, for which her optometrist had given her prisms. A swollen left upper eyelid had been present for 10 days (figure 1). No pain or headache was reported and no medication was being taken. Her general health was otherwise good and her medical history was unremarkable. Physical examination was normal with no lymphadenopathy. Ophthalmic examination showed

visual acuity of 6/5 in each eye, normal pupillary reactions and healthy colour vision test with Ishihara plates. Fundus examination was normal and no proptosis was found when measured with an exophthalmometer. Direct examination of the superior rectus muscle showed enlargement and hyperaemia with restricted elevation of the left eye, confirmed by a Hess plot.

INVESTIGATIONS

Blood tests for thyroid function and myasthenia were normal.

CT scan of the brain and orbits revealed a 3 cm × 1.5 cm mass arising from the region of the left superior rectus with no signs of bone erosion.

CT scan of the neck, chest, abdomen and pelvis were normal.

MRI of the brain and orbit showed extensive soft tissue expansion of the superior rectus with tendon involvement (figures 2 and 3).

Biopsy: Histology showed infiltration by small lymphoid cells (figure 4). Immunohistochemistry demonstrated

Figure 1 Limited elevation of the left eye and swelling of the upper eyelid.

Figure 2 Sagittal view showing enlarged superior rectus muscle.

Figure 3 MRI axial view.

these cells to be CD20 CD5–, CD10–, CD23–, CD43–, Bcl-2+ Bcl-6– and cyclin D-1–.

Histology diagnosis: Stage 1AE low-grade marginal zone B cell lymphoma (MZBCL).

DIFFERENTIAL DIAGNOSIS

Muscle metastasis, myositis and lymphoma.

TREATMENT

Initial treatment was with oral doxycycline 200 mg once a day for 3 weeks. There was no detectable improvement. Localised radiotherapy 30 Gy was therefore conducted.

OUTCOME AND FOLLOW-UP

Localised radiotherapy 30 Gy resulted in excellent clinical and radiological resolution of this isolated lymphoma.

DISCUSSION

Orbital malignancies are commonly caused by lymphomas. The majority (50–78% in Western nations and ≥90% in Japan and Korea¹) are MZBCLs, a subtype of non-Hodgkin B-cell lymphoma (1–2%). They have distinct pathogenic, histological and clinical features and can be primary or secondary. It is unclear whether MALT naturally occurs in the eye or if it is the result of inflammation, reinforcing theories of *Chlamydia psittaci* (*Cp*) involvement proposed by Ferreri *et al*,² who detected *Cp*-DNA in 80% of their MZBCL cases, a finding supported and contested by later studies.^{3–4} This hypothesis may explain the rising incidence and marked geographical variation of MZBCL. Based on this (and the success of *Helicobacter pylori* eradication in gastric MALT lymphomas), antibiotic therapy has become a novel treatment.¹ Trials have shown

Figure 4 Histology: the infiltrate consists of small lymphoid cells with irregular and hyperchromatic nuclei.

conflicting results however. A meta-analysis by Husain *et al*⁵ showed inconsistent efficacy, advising against ‘blind’ treatment before utilising diagnostic PCR analysis.

IgG4-related sclerosing disease is a recently recognised multisystemic condition characterised by an increased number of IgG4-producing plasma cells in affected tissues. Inflammatory lesions usually include the pancreas, sub-mandibular glands, lacrimal glands and lymph nodes. Cheuk *et al*⁶ have reported cases of MZBCL arising in IgG4-related chronic sclerosing dacryoadenitis and IgG4-positive sclerosing inflammation present in MZBCLs, indicating a possible link between the two conditions. They propose that the lymphoid hyperplasia of IgG4-related sclerosing disease can provide a substrate for the emergence of lymphoma.

Learning points

- ▶ Lymphomas are the most common orbital malignancies and typically manifest in the form of a conjunctival mass and/or proptosis. However, one should not rule out the diagnosis in the absence of such findings and remain vigilant to much less common presentations such as diplopia alone.
- ▶ Marginal zone B-cell lymphomas, a subtype of non-Hodgkin B-cell lymphomas, are the most common histotype indicated in ocular adnexal lymphomas.
- ▶ *Chlamydia psittaci* may be involved in ocular adnexal lymphomas; therefore treatment with antibiotics such as doxycycline should be considered.

Competing interests None.

Patient consent Obtained.

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