Isolated lateral rectus muscle large B cell lymphoma: A rare case report and review of the literature

Seyed Mohsen Rafizadeh a, Zohreh Nozarian b, Seyed Ali Sonbolestan a,∗

a Department of Oculo-facial Plastic and Reconstructive Surgery, Farabi Hospital, Eye Research Center, Tehran University of Medical Sciences, Tehran, Iran
b Department of Pathology, Farabi Hospital, Tehran University of Medical Sciences, Tehran, Iran

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ABSTRACT

Purpose: To report a 65-year-old male patient with primary lateral rectus large B cell lymphoma.

Observations: The patient had been referred because of progressive proptosis and limitation of ductions, especially abduction (ortho position at primary gaze) and conjunctival injection. Computerized tomography of the orbit and paranasal sinuses depicted a massive lateral rectus muscle enlargement without any other orbital involvement. Lateral orbitotomy and lateral rectus belly incisional biopsy was done, and histopathologic and immunohistochemical staining and systemic evaluations revealed the diagnosis of primary orbital large B-cell lymphoma.

Conclusions and importance: This case indicated that, though rare, extraocular muscle enlargement could be the main finding of primary orbital lymphoma. Large B-cell lymphoma could involve only the orbital tissues, although it is more prevalent with systemic involvement.

1. Introduction

Ocular adnexal lymphoma (OAL) can involve different structures such as the conjunctiva, eyelids, or orbital tissue, especially the lacrimal gland. OAL, which encompasses 5–10% of all extranodal lymphomas, is the most common malignancy of the ocular adnexa, from which 50–60% are orbital lymphomas.

Several infiltrative, inflammatory, infectious, and neoplastic etiologies can involve the extraocular muscles. Differentiating between these entities could be challenging due to the possible similarity of their clinical and imaging findings. In patients with painless enlargement of extraocular muscles and proptosis who do not show the characteristic features of thyroid eye disease a clinical suspicion for neoplasm should be kept in mind. However, the involvement of single extraocular muscle with lymphoma is notably rare.

Large B-cell lymphoma is the second most common B-cell lymphoma of the orbit which involves the orbital tissues mostly secondary to systemic disease. In this case report, we describe the clinical, imaging and pathologic findings of a primary orbital large B-cell lymphoma involving the lateral rectus muscle in a 65-year-old male patient.

2. Case report

A 65-year-old male presented to the orbit clinic complaining of progressive proptosis, redness, periocular swelling and decreased vision in his left eye in the last three months. His past medical history was positive only for hypertension.

Complete ophthalmic examination revealed a visual acuity of 20/20 (right eye, OD) and 20/50 (left eye, OS). The afferent pupillary defect (APD) OS was noted. His left eye proptosis was clear and exophthalmometry measurements at base 120 were 18 mm (OD) and 23 mm (OS). Prominent conjunctival injection, especially over the lateral rectus muscle, was seen. Regarding the oculomotor movements, minus 2 ocular motility limitation particularly in abduction was noted, but the patient’s eyes were in ortho position at primary gaze (Fig. 1A, B).

Other slit lamp examination findings were within normal limits, but on fundus evaluation 2+ optic disc edema and marginal disc blurring (OS) were documented. On palpation, no regional lymphadenopathy was found.

Computerized tomography (CT) scan of the orbit and paranasal sinuses showed an isolated enlargement of the left lateral rectus (LR) muscle (height up to 28 mm and anteroposterior length up to about 40 mm).

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mm) which partially extended to the superior orbital fissure (SOF). The fusiform enlargement involved only the LR muscle belly, and the medial margin of the mass partially encased the optic nerve sheath complex (ONSC), superior rectus muscle and inferior rectus muscle (Fig. 1, C, D).

Laboratory tests including complete blood count (CBC), thyroid function test, angiotensin-converting enzyme, and autoimmune serology were within normal limits.

Incisional biopsy under general anesthesia was scheduled. After lateral orbitotomy and exploration, the left LR muscle belly was found to be enlarged and inflamed, so a biopsy was taken from the muscle belly.

The histopathologic sections showed complete effacement of striated muscle tissue by a diffuse and cohesive infiltrate of large size atypical cells. Laboratory tests including complete blood count (CBC), thyroid function test, angiotensin-converting enzyme, and autoimmune serology were within normal limits.

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mononuclear cells. Most cells showed large, atypical nuclei (nucleus size ≥ histiocyte nucleus or > 2 lymphocyte nuclei) with vesicular chromatin and prominent nucleoli. For definitive diagnosis, a primary immunohistochemistry (IHC) panel was done. Tumor cells were positive for LCA and Vimentin and negative for S100, Melan A, HMWCK and EMA. A second panel for the diagnosis of lymphoma was done in which atypical cells were positive for CD20 and negative for CD3, CD99, CD30, TDT and BCL6. Ki67 was high also (positive in 60–70% of tumoral cells). Finally, the definitive diagnosis was large B-cell lymphoma (Fig. 2).

After incisional biopsy, because of optic disc edema and the patient’s pain and decreased vision, methylprednisolone pulse therapy (500 mg per day for three days) was initiated immediately, followed by 50 mg oral prednisolone per day for ten days. The patient’s visual acuity improved to 20/25, and his periorbital pain subsided.

After the final diagnosis, the patient was referred to an expert oncologist for complete workup and management. The systemic evaluation included chest, abdomen, and pelvic CT scan. No tumoral involvement or lymphadenopathy was recorded. Additionally, fluordeoxyglucose (FDG)-positron emission tomography (PET) reported no metabolic evidence of malignancy in any other portions of the body (Fig. 1, E, F). Due to its high-grade nature, the patient’s disease was chosen to be treated with R–CHOP regimen including rituximab, cyclophosphamide, doxorubicin (hydroxydaunorubicin/Adriamycin®), vincristine (Oncovin®), and prednisone in combination with radiotherapy.

### 3. Discussion

Isolated lymphomatous lesions of the extraocular muscles are rare and account for only 0.17% of all orbital lymphomas.7,8 These malignant lesions are often low–grade, and their prognosis is good. Systemic involvement is seen in a minority of patients.9

In this study, a case of lymphoma involving the lateral rectus muscle is described. Although rare, orbital lymphoma can arise discretely in extraocular muscles, but most of the reported lymphomatous lesions in this orbital structure are low grade disorders. The most common B-cell lymphoma is extranodal marginal zone B-cell lymphoma (EMZL) followed by large B-cell lymphoma (LBCL), which was diagnosed in this case.8 LBCL has a higher incidence among older patients, and according to previous studies 70% of patients were older than 50 years of age.

The most common symptom of orbital lymphoma, particularly B-cell lymphoma, is proptosis. Patients presenting with proptosis, specifically unilateral proptosis, should always be referred for orbital imaging such as a CT scan or magnetic resonance imaging (MRI) as soon as possible.2 Currently, the diagnosis of orbital lymphoma consists of CT or MRI followed by biopsy-proven histological confirmation.5

Several other symptoms such as limited ocular movements, swelling, pain, ptosis, changes in visual acuity, diplopia, chemosis and edema are also reported, some of which were seen in our case as well. LBCL often occurs as a unilateral lesion which also occurred in our case.5

According to previous reports, most B-cell lymphomas are situated in the extraconal space (72%), and the most common involved structure is the lacrimal gland (51% of all orbital B-cell lymphomas), while 8% of them are located in the intraconal area and 9% involve the extraocular muscles.6

Primary isolated involvement of extraocular muscle in LBCL is truly rare and, as far as we know, has been reported previously in three cases (Table 1).7,9 These patients reported diplopia, low vision or periorbital pain and swelling. All of them underwent incisional biopsy and chemotherapy with or without radiotherapy was the main treatment method. On the other hand, compressive optic neuropathy (CON) due to extraocular muscle lymphomatous involvement was reported rarely in the literature. According to our search, only four cases were reported previously.5,7,9,10 Our case suffered from optic neuropathy which required us to implement a steroid pulse treatment. Steroid pulse therapy after surgical biopsy may lead to a medical decompression, and prednisolone itself is included in chemotherapy regimens.11 The prognosis of CON-induced visual impairment depends on the duration of symptoms.12 Short-term use of steroids after biopsy can protect the optic nerve from irreversible damage. The incisional biopsy itself may not decrease the severity of CON, because only a small part of the tumor is resected and the surgery itself may worsen the condition from tissue swelling or even limited intraorbital hemorrhage.

The main feature of our case was its primary form. Primary orbital lymphoma is demarcated as a biopsy-confirmed orbital lymphoma with no evidence of concurrent systemic lymphoma after a complete clinical assessment or no previous history of lymphoma disease.7 Histopathologic diagnosis of orbital lymphoma is based on the tissue morphology. Of course, other complementary evaluations such as IHC and gene rearrangement can help confirm the diagnosis and help to differentiate types of lymphomas.13,14 In this case, histopathologic and IHC studies confirmed the type of lymphoma to be LBCL.

### 4. Conclusion

In conclusion, despite its rarity, ocular adnexal lymphoma, even its less common forms, should be included in the differential diagnosis of enlarged extraocular muscles. Imaging and, if needed, biopsy should be done for exact diagnosis and appropriate treatment.

### Patient consent

Written informed consent was obtained from the patient for participation in the study and publication of this report and related photos.

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### Declaration of competing interest

The authors declare that there is no conflict of interest.

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### Table 1

Primary Large B-cell lymphoma of the extraocular muscles cases.

| Case No | Gender | Age | Eye | Presenting symptom | Involved extraocular muscle | Treatment
|---------|--------|-----|-----|--------------------|-----------------------------|---------|
| 1       | Female | 62  | Right | Ptosis and mild ocular pain | Superior rectus-levator muscle complex | Chemotherapy |
| 2       | Male   | 82  | Left  | Vertical diplopia and periorbital swelling | Superior and medial rectus muscles | Chemotherapy and radiotherapy |
| 3       | Male   | 67  | Right | Diplopia and blurred vision | Inferior and medial rectus muscles | Intravenous prednisolone and chemotherapy |
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