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Case Report

Isolated superior oblique muscle extranodal marginal zone B cell lymphoma: case report

Fatimah Alhammad [a,*]; Azza Maktabi [a]; Hind M. Alkatan [b]; Sahar M. Elkhamary [c]; Enmar Almazyad [a]; Osama Al-Sheikh [a]

Abstract

We describe a rare case of isolated extraocular muscle ocular adnexal lymphoma of a middle-aged female who presented with redness in the left eye associated with progressive proptosis over one year. Magnetic resonance imaging of the orbit indicated isolated enlargement of the left superior oblique (SO) muscle with an apparent diffusion coefficient (ADC) of ($0.77 \pm 0.11 \times 10^{-3}$ mm^2/s). Histopathology with immunohistochemical staining of the incisional biopsy from the SO muscle belly confirmed the diagnosis of extranodal marginal zone B cell lymphoma.

Keywords: Ocular adnexal lymphoma, Proptosis, Superior oblique

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Introduction

Extraocular muscles can be affected by various etiologies; including infiltrative, inflammatory, infectious, and neoplastic processes. These etiologies may share similar, clinical and radiological features making diagnosis challenging.

High level of clinical suspicion for neoplasm should be considered in any painless enlargement of extraocular muscles that lack the characteristics of thyroid eye disease, especially when the superior component is involved.[1]

Orbital lymphoma is the most common malignant tumor encountered in the orbit.[2] However, the involvement of single extraocular muscle with this tumor is considered extremely rare.[3] Previous studies have reported greater involvement of the rectus muscles compared other ocular muscles[3]

In this case report, we describe the clinical, radiological and pathological characteristics of a biopsy-proven ocular adnexal lymphoma involving the superior oblique muscle in a middle-aged female.

Case report

A 42-year-old female presented to the oculoplastic clinic complaining of redness in the left eye associated with progressive proptosis over one year. She is known case of diabetes mellitus, bronchial asthma, and cardiac disease.

Ophthalmic examination indicated a visual acuity of 20/40 and intraocular pressure of 19 mmHg bilaterally. The left eye showed proptosis (exophthalmometry measurements at base 105 were 16 mm in the right eye (OD) and 20 mm in the left eye (OS)). She had a conjunctival injection, upper lid ptosis

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[a] King Khaled Eye Specialist Hospital, Riyadh, Saudi Arabia
[b] Departments of Ophthalmology and Pathology, College of Medicine, King Saud University, Riyadh, Saudi Arabia
[c] Department of Diagnostic Radiology, Mansoura Faculty of Medicine, Egypt

* Corresponding author at: King Khaled Eye Specialist Hospital, PO Box 7191, Al Aroubah Road, Riyadh 11642, Saudi Arabia.
e-mail address: dr.alhammad@ymail.com (F. Alhammad).

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(MRD1 4 mm OD and 2 mm OS) and ocular motility limitation. Other aspects of the examination including the anterior segment, optic disc and fundus were within normal limits in both eyes. There was no regional lymphadenopathy.

Magnetic resonance imaging (MRI) studies of the orbit indicated an isolated enlargement of the left superior oblique (SO) muscle, that appeared as a fusiform muscle enlargement from its tendinous origin just before its attachment to the trochlear fossa of the frontal bone in the upper medial aspect of the orbit along with the muscle belly. The mass lesion had an intermediate signal intensity on T1 and T2-weighted images and intense enhancement following intravenous injection of contrast media with diffusion-weighted Image on b = 1000 and b = 0 sequences. The apparent diffusion coefficient (ADC) was ($0.77 \pm 0.11 \times 10^{-3}$ mm^2/s) determined by the manual region-of-interest intensity with diffusion pattern relatively low signal intensity within the lesion compared to that of brain parenchyma. There was no evidence of infiltration of the adjacent superior rectus and medial rectus muscles with normal surrounding fat (Fig. 1).

Laboratory tests were normal including complete blood count (CBC), thyroid function test, angiotensin-converting enzyme, and autoimmune serology. However, the Mantoux test (purified protein derivative; PPD) was positive.

Incisional biopsy under general anaesthesia was planned. On exploration, left SO muscle belly was enlarged and inflamed with dilated and tortuous engorged blood vessels were along its tendon. A biopsy was taken from SO muscle belly.

The histopathologic section showed muscle fibers separated by diffuse proliferation of small round cells with dark nuclei and minimal cytoplasm representing mature lymphocytes. Occasional larger lymphoid cells and some plasma cells were also present (Fig. 2). Immunohistochemical staining showed a predominant infiltrate of B-lymphocytes that was CD20, CD43 and Bcl-2 positive and CD10, CD30 and Bcl-6 negative (Fig. 3). In addition, immunoglobulin heavy chain gene rearrangement was positive. Hence the pathological diagnosis was extranodal marginal zone B cell lymphoma.

After reaching the final diagnosis, the patient was referred to a general hospital with an oncology center for a full metastatic workup to exclude systemic disease and management.

Discussion

Ocular adnexal lymphomas (OALs) are the most common orbital neoplasms in adults. It may arise from different origins include the conjunctiva, lacrimal gland, eyelids, orbital soft tissue, and extraocular muscles.[4] Extraocular muscles involvement is considered rare and accounts for only 0.17% of all orbital lymphomas.[3]

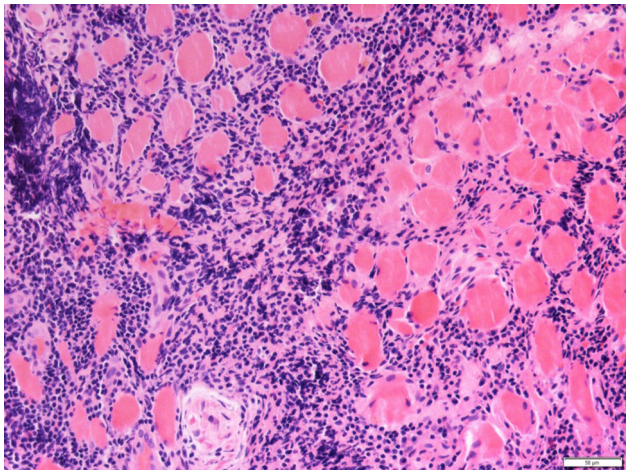

Fig. 2. Microphotograph of the SO muscle biopsy showing the muscle fibers separated by diffuse proliferation of small round cells with dark nuclei and minimal cytoplasm representing mature lymphocytes (Original magnification ×200. Hematoxylin and eosin).

We describe here in a case of lymphoma involving the superior oblique muscle. Although the most commonly infiltrated structures by lymphoid tumors are found within the superio-lateral quadrant, including the superior rectus muscle, lateral rectus muscle and lacrimal gland,[5] the involvement of superior oblique muscle has been reported in only a few cases in the literature. In a major review extraocular muscle lymphoma, rectus muscles were found to be more involved than the obliques and levator muscles with 73%, 17%, and 11% respectively.[3] Isolated involvement of SO in OALs is extremely rare and has been reported previously in one case only.[3]

The SO muscle is rarely involved in different pathological entities that affect extraocular muscles.[6] Isolated SO muscle involvement in thyroid eye disease (TED) has not been reported and early involvement of the SO muscle make other differentials ahead of TED including malignancy which should be ruled out.[7] In the current report, the patient had a positive purified protein derivative (PPD) tuberculosis test. Hence we considered the possibility of tuberculosis myositis in the differential diagnosis and the aim was to get a biopsy-proven diagnosis before any empirical treatment.

Imaging can help differentiate lymphoma from other etiologies. Diffusion-weighted imaging (DWI) has been

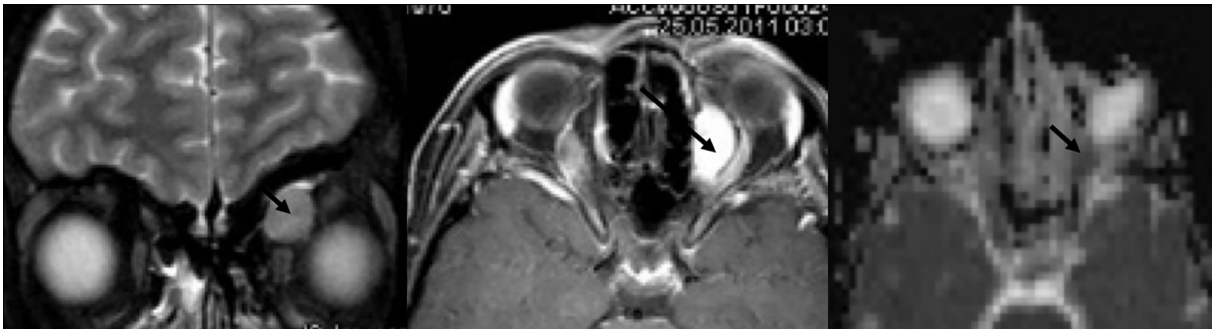

Fig. 1. Magnetic resonance images of the left superior oblique (SO) muscle isolated enlargement (Black arrow) in 3 different consecutive images: T2-weighted coronal image with fat suppression, Post-contrast T1-weighted axial image with fat suppression, and diffusion-weighted axial image.

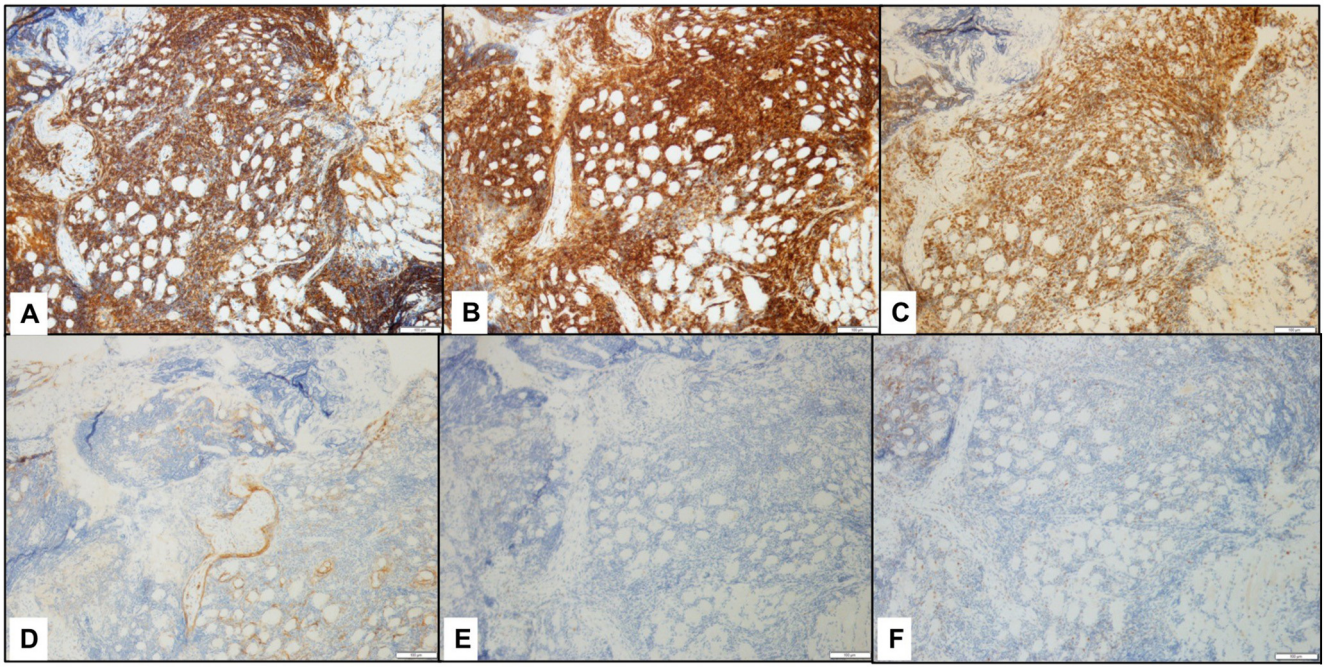

Fig. 3. The immunohistochemical profile of the lymphocytic proliferation showing expression of cells to CD20 in A, CD43 in B, and Bcl-2 in C. Note the absence of expression of the following markers: CD10, CD30, and Bcl-6 in D, E, and F confirming the diagnosis of extra-nodal marginal-zone lymphoma (Original magnification ×100).

increasingly used as a non-invasive imaging modality to characterize soft-tissue masses using apparent diffusion coefficient (ADC) measurements to differentiate malignant tumors from other benign etiologies.[8] In the current study, the lesion had an ADC value of $(0.77 \pm 0.11 \times 10^{-3}\ mm^2/s)$. Lesions with an ADC less than $0.93 \times 10^{-3}\ mm^2/s$ are 90% specific for malignancy.[9] Furthermore, the high cellularity of lymphoma which exceeds the cellularity of other malignant tumors[10] makes the ADC values for lymphomas low (mean, $0.55 \pm 0.06\ 10^{-3}\ mm^2/s$) compared to other malignant tumors and idiopathic orbital inflammation.

Histopathologic diagnosis is based on the morphology. However, the immunohistochemistry and gene rearrangement support the diagnosis and aid to distinguish different types of lymphoma.[11] In our patient, these studies confirmed the type of lymphoma to be extranodal marginal zone B cell lymphoma. This type is considered the most common orbital and adnexal lymphomas followed by follicular lymphoma. Other types include mantle cell lymphoma, diffuse large B cell lymphoma and chronic lymphocytic leukemia rarely affect the ocular adnexa. NK/T-cell lymphoma and Hodgkin lymphoma are also rare.[12]

In conclusion, isolated SO lymphoma is an extremely rare presentation. Malignancy should be considered, and diagnosis confirmation by biopsy is warranted even in the presence of other provisional diagnoses.

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Conflict of interest

The authors declared that there is no conflict of interest.

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