

# Extranodal marginal zone B-cell lymphoma involving superior rectus muscle: A clinicopathological case report

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**Abstract.** A 66-year-old female had suffered from proptosis in the left eye (OS) and double vision for 1 month due to abnormality of the superior rectus muscle. Visual acuity was noted as 20/20 in both eyes (OU). Eye movement showed limited OS supraduction. Magnetic resonance imaging revealed an indistinct mass in the orbit involving the superior rectus muscle. A biopsy specimen of the orbital tumor led to the histological diagnosis of extranodal marginal zone B-cell lymphoma. Radiotherapy with a total dosage of 30 Gy was administered, which subsequently resolved the tumor. However, the supraduction limitation of ocular movement remained unchanged. Supraduction limitation is due to muscular contraction disorder of the superior rectus muscle, caused by direct lymphoma cell invasion.

## Case report

Extranodal marginal zone B-cell lymphoma (EMZL) of mucosa-associated lymphoid tissue lymphoma arises from the marginal zone of mucosa-associated lymphoid tissue. EMZL is common among ocular adnexal malignant tumors; however, orbital lymphomas involving striated muscles are rare. This report examines an unusual case of orbital EMZL involving the superior rectus muscle.

A 66-year-old female suffering from proptosis of the left eye (OS) and double vision for 1 month was referred to our hospital due to an abnormality of the superior rectus muscle OS. Visual acuity was found to be 20/20 in both eyes with a normal intraocular pressure. Extraocular examination demonstrated upper eyelid swelling without ocular pain. The fundus was normal. Hess screen analysis revealed supraduction OS (Fig. 1). The laboratory values, including a blood cell count, biochemistry, and thyroid hormones, were found to be normal.

No systemic abnormality was detected with the exception of the orbit. Initial magnetic resonance imaging (MRI) of the orbit revealed an indistinct mass in the superior orbit close to, or within, the superior rectus muscle (Fig. 2). Differential diagnoses of the mass lesion in the extraocular muscle were orbital tumor, Graves' disease, and orbital myositis. Biopsy of the orbital mass was performed.

A histological examination showed diffuse atypical lymphoid cell infiltration, mixed with plasma cells with Russell bodies. Lymphoid cells were present within fragmented striated muscles (Fig. 3A). Immunohistochemically, the atypical lymphoid cells were positive for CD20 and CD79a, markers for B-cells (Fig. 3B and C), and negative for CD3 and CD5, markers for T-cells. The immunohistochemical examination of immunoglobulin showed deviation to  $\kappa$  chains in the infiltrating lymphoid cells (Fig. 3D).

The orbital tumor was diagnosed as EMZL involving the superior rectus muscle. Radiotherapy with a total dosage of 30 Gy was administered. The radiotherapy reduced the volume of the tumor (Fig. 4, red arrow), and the superior rectus muscle was clearly identified (Fig. 4, white arrow). The supraduction limitation in OS movement remained unchanged, although tumor recurrence was not observed one and a half years after radiotherapy.

## Discussion

Since the border between the tumor and superior rectus muscle was not differentiated in the initial MRI of the orbit, the origin of the tumor remains to be determined. Histological examination showed atypical lymphoid cell infiltration within the fragmented striated muscle. Muscle fibers in the remaining striated muscle were clearly noted in the specimen, indicating that the muscle was not degenerative. Following radiotherapy, superior rectus muscle with a reduced tumor was visualized. Therefore, the clinicopathological findings indicate that EMZL did not arise in the extraocular muscle, but arose in the orbital soft tissue adjacent to the muscle, followed by invasion to the superior rectus muscle.

In this case, only superior muscle was involved in tumor cell invasion, presenting with supraduction limitation, but not inferior adduction limitation. These results suggest that supraduction limitation occurred due to muscular contraction disorder of the superior rectus and not due to restriction by an enlarged antagonist muscle as observed in Graves' disease.

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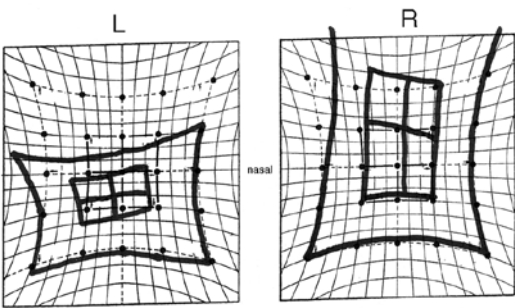


Figure 1. Hess screen analysis reveals difficulties with elevation of the left eye. L, left; R, right.

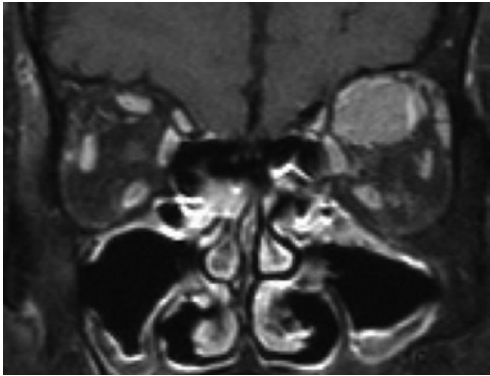


Figure 2. Magnetic resonance imaging shows a mass lesion at the level of the superior rectus muscle in the left orbit.

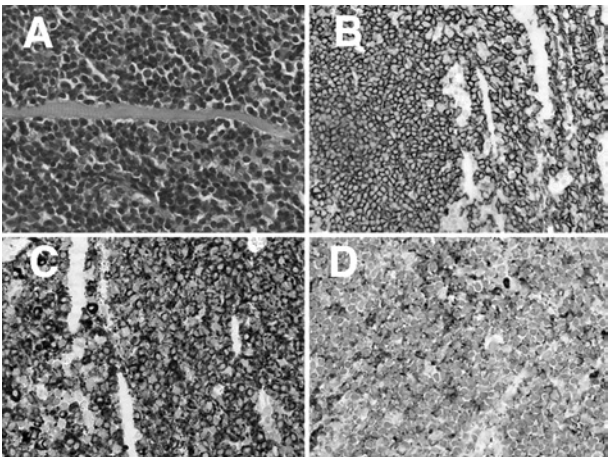


Figure 3. Histopathology of the biopsied orbital mass. (A) Typical lymphoid cells were diffusely infiltrated and mixed with fragmented striated muscle (hematoxylin-eosin). (B) Cytoplasmic immunoreactivity for CD20, a marker for B-cells, is observed in tumor cells. (C) Cytoplasmic immunoreactivity for CD79a, a marker for B-cells, is observed in tumor cells. (D) Immunohistochemical staining of immunoglobulin shows deviation to the  $\kappa$  chain in the infiltrating lymphoid cells.

Together with the histological findings, it was noted that the extraocular muscle contraction disorder is caused by direct lymphoma cell invasion. Despite resolution of the tumor after irradiation, supraduction limitation was not improved, suggesting that irradiation led not only to tumor cell death, but also subsequent irreversible muscle fibrosis.

Table I. Clinicopathological characteristics in orbital lymphomas involving the superior rectus muscle in the literature.

Case	Gender	Age (years)	Eye	Orbital site involved	Histology	Extraorbital involvement	Therapy	Eye movement disorder after therapy	Refs.
1	F	62	R	Superior rectus-levator muscle complex	Diffuse large B-cell lymphoma	No	C	Mildly limited	2
2	M	82	L	Superior and medial rectus muscle	Diffuse large B-cell lymphoma	Yes (stage III)	C and R	Improved	3
3	M	44	Bilateral	All extraocular muscles	Peripheral T-cell lymphoma	Yes (stage III)	C	Mildly limited	4
4	M	40	R	Superior rectus-levator muscle complex	Follicular lymphoma	No	R	Improved	5
5	M	57	R	Superior rectus-levator muscle complex	Diffuse T-cell lymphoma	No	R	Improved	5
6	F	61	R	Superior rectus-levator muscle complex	Poorly differentiated lymphocytic lymphoma	Yes	R	Improved	5
7	M	60	R	Superior rectus muscle	Lymphoplasmacytoid lymphoma	No	R	Improved	5
8	F	62	R	Superior rectus muscle	Mature T-cell lymphoma	No	R	Improved	5
9	F	67	L	Superior rectus muscle	Diffuse mature lymphoma	No	R	Improved	5
10	F	66	L	Superior rectus muscle	Extranodal marginal zone B-cell lymphoma	No	R	Not improved	Present case

M, male; F, female; R, right; L, left; C, chemotherapy; R, radiotherapy.

Table II. Clinicopathological characteristics with extraocular muscle involvement in extranodal marginal zone B-cell lymphoma of the orbit.

Case	Gender	Age (years)	Eye	Orbital site involved	Extraorbital involvement	Therapy	Eye movement disorder after therapy	Refs.
1	F	54	R	Inferior oblique muscle	No	R	-	6
2	M	52	L	Inferior rectus muscle	No	Rituximab	-	7
3	F	57	R	Inferior rectus muscle	No	R	-	8
4	M	75	R	Medial rectus muscle	Yes (stage IV)	C	-	8
5	M	80	L	Lateral rectus muscle	No	Chlorambucil	-	8
6	F	54	R	Inferior oblique muscle	No	R	-	8
7	M	34	R	Levator muscle	No	R	-	8
8	F	66	L	Superior rectus muscle	No	R	Not improved	Present case

M, male; F, female; R, right; L, left; C, chemotherapy; R, radiotherapy.

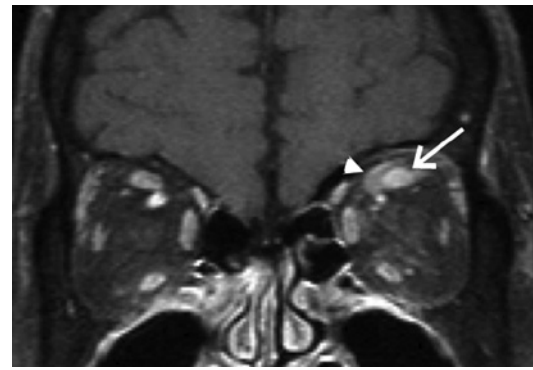


Figure 4. Magnetic resonance imaging following radiotherapy shows a decreased orbital tumor volume (arrowhead) and the superior rectus muscle (arrow).

It was previously reported that the most common site of ocular EMZL was the conjunctiva (51%), while extraocular muscles were the most rare localization (only 5%) (1). A review the literature revealed only 9 cases of orbital lymphoma invading the superior rectus muscle, in which, histologically, EMZL was not included (2-5) (Table I). In contrast, 7 cases of extraocular muscle involvement in EMZL have been reported, but no case exists involving the superior rectus muscle and ocular movement disorder with the exception of this case (6-8) (Table II). Two cases of diffuse large B-cell lymphoma and peripheral T-cell lymphoma exhibited the mild limitation of eye movement following treatment (Table I). Irreversible eye movement disorder involving EMZL following either chemo- or radiotherapy has not been reported as in this case. Therefore, EMZL arising in the orbit shows extraocular muscle involvement and leads to impairment of the visual function.

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