

Magnetic Resonance Imaging Features of Extraocular Muscle Lymphoma in Five Cases

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Lymphoma is the most common primary orbital malignant neoplasm in adults, accounting for 8–15% of extranodal non-Hodgkin's lymphomas and 2% of all lymphomas.^[1] Most of these tumors involve the conjunctiva, orbit, lacrimal gland, or eyelid. It is uncommon for lymphoma to originate from the extraocular muscles. The prevalence of intramuscular lymphoma is between 0.1% and 8.7% of all extranodal lymphomas. Fewer than 60 cases have been reported in the last 30 years,^[2] mostly as case reports with scant or no radiology information.^[2]

The study was approved by the Institutional Review Board of Beijing Tongren Hospital. We reviewed 98 cases involving patients who underwent surgical or needle aspiration biopsy treated in Beijing Tongren Hospital between September 2007 and April 2014. The inclusion criteria were as follows: (1) patients who were treatment naïve before hospitalization; (2) patients who underwent magnetic resonance imaging (MRI) examination before treatment; and (3) patients with lesions originating from the extraocular muscles. Five patients were finally enrolled in the study. They had all undergone orbital MRI, dynamic contrast-enhanced (DCE) imaging, and additional delayed (contrast-enhanced) acquisitions. Their MRI and pathology were primarily analyzed.

For DCE data analysis, based on parameters, such as time, maximal enhancement intensity (SI_{max}), and final enhancement intensity (SI_{final}),^[3] the time-intensity curve (TIC) was assigned into three patterns: Type I, a persistent pattern with a continuous curved line in the entire dynamic period; Type II, a plateau pattern with a prominent increase in slope with $SI_{final} = 90\text{--}100\%$ of SI_{max} ; and Type III, a washout pattern with a rapid increase in slope with the $SI_{final} < 90\%$ of SI_{max} .

Five patients were male with a median age of 50.6 ± 11.8 years (range, 33–64 years). Four cases were unilateral (three on the left side and one on the right side) and one case was bilateral. The disease affected six muscles in five patients, including the inferior oblique muscle in two patients, and the levator palpebrae superioris muscle and the superior ocular muscle group in one patient each. In addition, both superior ocular muscle group and inferior rectus were affected in one patient. All tumors presented enlargements in the tendons and belly of the muscle and had well-defined margins. Four enlarged muscles were fusiform, and two were irregularly shaped [Figure 1a–1c]. All cases displayed homogeneous isointensity to gray matter on T1-weighted image (WI) and T2-WI. After injection of the gadopentetate dimeglumine, mildly and moderately homogeneous enhancement appeared in one and four cases, respectively [Figure 1d]. In the DCE-MRI phase, three patients had Type III TIC patterns and two had Type II [Figure 1e]. The optic nerve sheaths, posterior fascia, orbital nerve canal, and contralateral intraconal compartment were involved in one patient. In another patient, the homolateral orbital apex and posterior sheaths of the optic nerve were involved. The orbit was displaced superiorly, inferiorly, or anteriorly in three of the patients. All patients were pathologically identified

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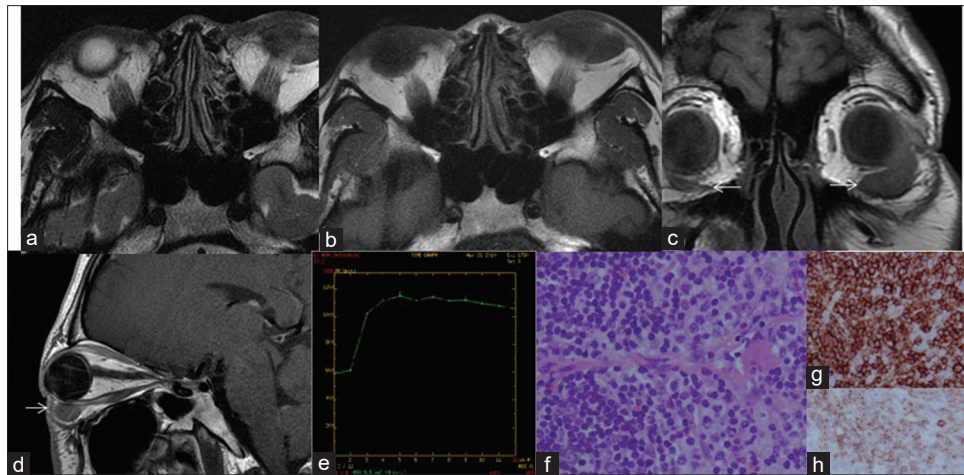


Figure 1: Magnetic resonance imaging features and pathology results. (a and b) Axial T1-WI and T2-WI showing the enlargement of the left inferior oblique muscle with homogeneous isointensity. (c) Coronal T1-WI revealing that both the right (arrow) and the left inferior oblique muscles are enlarged. (d and e) After injection of contrast, the lesions undergo slight isointense homogeneous enhancement (arrow on d) and were Type II in DCE-MRI. (f) Hematoxylin and eosin stained section showing some hyperplasia in the lymphoid tissue with consistent form (original magnification, $\times 400$), there are hundreds of uniform hyperplasia cells with mild atypia and monocytic B-cell differentiation. (g and h) The immunohistochemistry indicates that the tissue is positive for both CD20 and CD79, respectively (original magnification, $\times 400$). WI: Weighted image; DCE-MRI: Dynamic contrast-enhanced magnetic resonance imaging.

as having mucosa associated lymphoid tissue lymphoma [Figure 1f–1h].

In previously reported cases, the origin of extraocular muscle lymphoma has varied. Fukuhara *et al.*^[4] and Surov *et al.*^[5] described one and three intramuscular lymphomas, respectively; they were located in the rectus (one in the superior rectus muscle, two in the lateral rectus muscle, and one in the medial rectus muscle). After reviewing local data and the previous literature, Watkins *et al.*^[2] also concluded that the lesions were more frequently involved with the rectus muscles (73%) than the oblique or the levator palpebrae superioris muscle (11%). Moreover, the majority of cases consisted of single muscle involvement.^[2] Orbital lymphoma tumors have been reported to be most frequently located in the superior rectus, followed by the inferior rectus, lateral rectus, and medial rectus. However, in our study, lesions were found in the inferior oblique muscle (2/5 patients), levator palpebrae superioris muscle (1/5 patient), superior ocular muscle group (1/5 patient), and superior ocular muscle group and inferior rectus (1/5 patients). These differences may be the result of statistical errors caused by the small patient population.

The extraocular muscle lymphomas involved thickening of both muscle tendon and belly. The majority of muscle enlargements was fusiform on axial MRI, similar to the results of previous case studies.^[2] In addition, the lesions tended to be unilateral with no side bias. Only 9% of the reported cases have involved both eyes.^[2] MRI features of extraocular muscle lymphomas were nonspecific, with most of them having homogeneous isointensity on T1-WI and T2-WI, making it difficult to differentiate them from other lesions. In this situation, DCE may provide helpful information. Yuan *et al.*^[3] found that orbital lymphomas usually presented as Type II or Type III in DCE-MRI, corresponding to higher cell density in the lymphoma. In

the present study, two and three cases of extraocular muscle lymphoma presented as Type II and Type III, respectively.

In conclusion, extraocular muscle lymphoma frequently presents as a painless swelling, often with a single muscle affected. On MRI, the lesions demonstrate isointensity on T1-WI and T2-WI with Type II or Type III enhancement on TIC.

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Conflicts of interest

There are no conflicts of interest.

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