

CT, Magnetic Resonance, and ^{18}F -Fluorodeoxyglucose Positron Emission Tomography/CT Imaging Features of Mucosa-Associated Lymphoid Tissue Lymphoma Involving Medial Rectus Muscle: A Case Report¹

내직근을 침범한 점막 연관성 림프조직 림프종의 CT, MRI 및 ^{18}F -Fluorodeoxyglucose Positron Emission Tomography/CT 영상소견: 증례 보고¹

Sang Kwon Lee, MD¹, Mi Sun Choe, MD²

Departments of ¹Radiology, ²Pathology, Dongsan Medical Center, Keimyung University School of Medicine, Daegu, Korea

We report a case of mucosa-associated lymphoid tissue (MALT) lymphoma involving the medial rectus muscle in a 47-year-old man along with CT, MRI, ^{18}F -fluorodeoxyglucose positron emission tomography/CT (^{18}F -FDG PET/CT), and pathologic features. The lesion was manifested as a fusiform enlargement isolated to the right medial rectus muscle with involvement of its tendinous insertion. The lesion was isoattenuating to the brain on non-enhanced CT images, showing as isointense to gray matter on fast spin echo T1- and T2-weighted images with fat saturation, and showed homogeneous enhancement on contrast-enhanced CT and MR images. The maximum standardized uptake value on ^{18}F -FDG PET/CT was 4.9 g/mL. The results of histological and immunohistochemical examinations of the specimen obtained by biopsy of the right medial rectus muscle were consistent with MALT lymphoma. It should be noted that the extraocular muscle (EOM) is a rare location for the involvement of MALT lymphoma, and MALT lymphoma of the EOM may mimic thyroid orbitopathy.

Index terms

Lymphoma, B-Cell, Marginal Zone
Eye Diseases
Tomography, X-Ray Computed
Magnetic Resonance Imaging
Positron-Emission Tomography and Computed Tomography

INTRODUCTION

The extraocular muscle (EOM) is the most frequent site for the involvement of thyroid orbitopathy, followed in order by inferior, medial, superior, lateral rectus, and oblique muscles. The EOM is the rarest site for the involvement of extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma) in the orbit. Several cases of MALT

lymphoma involving the EOM have appeared in the English literature, and most of these are in the form of case reports (1-5). The imaging findings of MALT lymphoma involving the EOM are limited to those of a single imaging modality only (1-3). As such, imaging features of this rare entity at CT, MRI, and ^{18}F -fluorodeoxyglucose positron emission tomography/CT (^{18}F -FDG PET/CT) altogether in a single case have not appeared in the literature.

Received March 1, 2013; Accepted April 11, 2013

Corresponding author: Sang Kwon Lee, MD
Department of Radiology, Dongsan Medical Center,
Keimyung University School of Medicine,
56 Dalseong-ro, Jung-gu, Daegu 700-712, Korea.
Tel. 82-53-250-7735 Fax. 82-53-250-7766
E-mail: sklee@dsmc.or.kr

This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (<http://creativecommons.org/licenses/by-nc/3.0>) which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

We report here a case of MALT lymphoma involving the medial rectus muscle in a 47-year-old man along with imaging features at CT, MRI, and ^{18}F -FDG PET/CT.

CASE REPORT

A 47-year-old man presented with a chief complaint of right ocular discomfort lasting for 1 year. He had not experienced ocular pain or diplopia. His past history was unremarkable. The ophthalmologic examination disclosed proptosis of the right eye, but the motion of the EOM was preserved. The laboratory tests, including blood cell count, biochemistry, and thyroid hormones, were normal. CT, performed by using a SOMATOM Definition Flash scanner (Siemens Healthcare, Forchheim, Germany) with a slice thickness of 3.0 mm revealed a fusiform enlargement isolated to the right medial rectus muscle, appearing as isoattenuating compared with the brain on non-enhanced CT images, and showed homogeneous, moderate enhancement on contrast-enhanced CT images, with involvement of the tendinous insertion (Fig. 1A, B). Orbital MRI was performed for better characterization of the lesion and for better assessment of the extent by using a 3.0-T unit (Signa Excite; GE Medical System, Milwaukee, WI, USA). The enlarged medial rectus muscle was isointense to the gray matter on fast spin echo (FSE) T1-weighted images (T1WI) [repetition time (TR)/echo time (TE) = 650.0/13.6; echo train length (ETL) = 3; field of view (FOV) = 160 × 160; matrix number/number of excitation = 320 × 192/2; slice thickness = 3.0 mm] with fat saturation (Fig. 1C) and FSE T2-weighted images (T2WI) (TR/TE = 5000.0/111.9; ETL = 20; FOV = 160 × 160; matrix number/number of excitation = 384 × 224/2; slice thickness = 3.0 mm) with fat saturation (Fig. 1D), and showed homogenous intense enhancement on gadolinium (Gd)-enhanced FSE T1WI with fat saturation (Fig. 1E). ^{18}F -FDG PET/CT, performed by using Discovery STE (GE Healthcare, Milwaukee, WI, USA), demonstrated increased FDG uptake [maximum standardized uptake value (SUV_{max}) = 4.9 g/mL] in the right medial rectus muscle with fusiform enlargement (Fig. 1F). No abnormally increased FDG uptake was noted in other parts of the body. In view of clinical and imaging findings, our tentative diagnosis was lymphoma and euthyroid Graves' ophthalmopathy. The patient underwent biopsy of the right medial rectus muscle. The histological and immunohisto-

chemical findings of the biopsy specimen were interpreted as MALT lymphoma (Fig. 1G, H). No findings of bone marrow involvement were documented in the bone marrow biopsy. Fractionated external beam irradiation of a total dose of 3600 cGy was applied to the right orbit. Near complete resolution of the MALT lymphoma of the right medial rectus muscle was achieved at follow-up MRI 230 days after the initial study (Fig. 1I). The patient underwent endoscopic biopsy for a raised, erosive mucosal lesion of the stomach 250 days after initial diagnosis of orbital MALT lymphoma, which revealed gastric MALT lymphoma. He underwent additional curative fractionated external beam radiation therapy at a dose of 3060 cGy targeted to the stomach. Follow-up endoscopic examination with biopsy, performed 133 days after initial diagnosis of gastric MALT lymphoma, demonstrated complete remission. No evidence of recurrent disease was observed in the orbit, stomach, or elsewhere until 13 months after complete remission.

DISCUSSION

Thyroid orbitopathy is the most common cause of abnormal enlargement of the EOM. Enlargement of the EOM associated with thyroid orbitopathy is bilateral and symmetric in 70% of cases. The inferior rectus muscle is most frequently affected, and the muscle enlargement is characteristically fusiform with sparing of tendinous insertion. The most common nonthyroidal causes of EOM enlargement are inflammatory and vascular disorders, and neoplastic diseases (6). MALT lymphoma, an extranodal, monoclonal, small B-cell proliferation, is an indolent disease that usually presents as a localized extranodal tumor. MALT lymphoma arises from various mucosal and nonmucosal tissues. It most commonly occurs in the stomach, but can involve the orbital/ocular adnexa, salivary glands, and lungs (7). Orbital MALT lymphoma most frequently involves the conjunctiva (51%), while the EOM is the rarest site of involvement (5%) (8). Several cases of MALT lymphoma involving the EOM have been presented in the English literature in the form of case reports or clinical studies (1-5). Of them, a few cases have presented imaging features (1-3). However, none have described imaging findings in detail. Rossman et al. (1) reported a case of an enlarged medial rectus muscle caused by MALT lymphoma that was misdiagnosed as thyroid orbitopathy for over 3 years, even

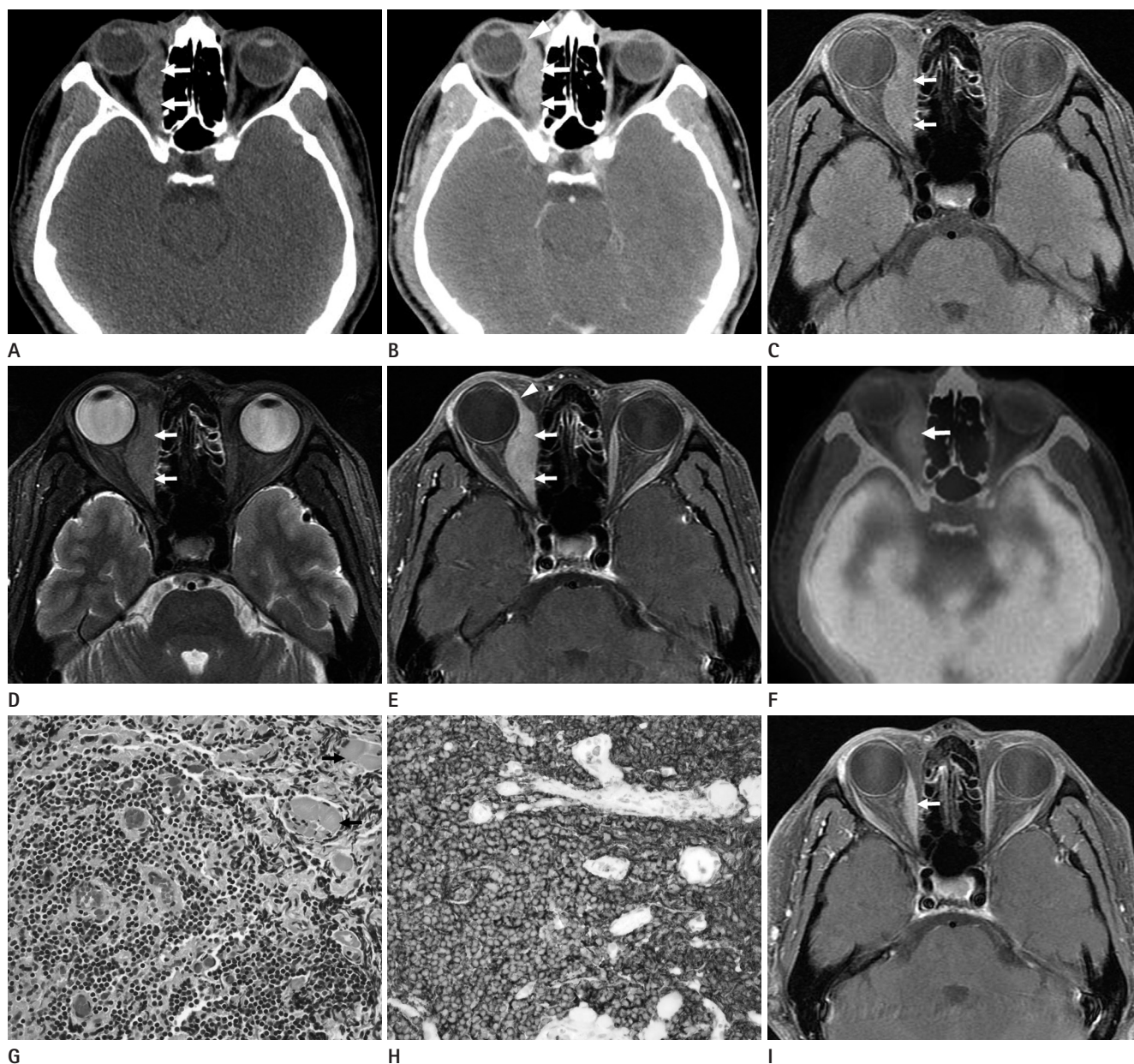


Fig. 1. CT, MRI, ^{18}F -FDG PET/CT, and pathologic features of MALT lymphoma involving the medial rectus muscle in a 47-year-old man.

A. A non-enhanced CT image shows fusiform enlargement isolated to the right medial rectus muscle (arrows), which is isoattenuating compared with brain.

B. A contrast-enhanced CT image reveals homogeneous, moderate enhancement of the enlarged muscle (arrows) with involvement of its tendinous insertion (arrowhead).

C, D. Fast spin echo T1- (**C**) and T2-weighted MR images (**D**) with fat saturation show fusiform enlargement of the right medial rectus muscle (arrows), which is isointense to the gray matter.

E. A gadolinium-enhanced T1-weighted MR image with fat saturation demonstrates homogeneous intense enhancement of the enlarged muscle (arrows) with involvement of its tendinous insertion (arrowhead).

F. A ^{18}F -FDG PET/CT image reveals moderate FDG uptake ($\text{SUV}_{\text{max}} = 4.9 \text{ g/mL}$) within the enlarged right medial rectus muscle (arrow).

G. A photomicrograph of histological examination of the specimen obtained by biopsy of the right medial rectus muscle demonstrates diffuse infiltration of relatively small monomorphic atypical lymphoid cells which infiltrate into the skeletal muscle (arrows) (H&E stain, $\times 400$).

H. A photomicrograph of immunohistochemical staining demonstrates diffuse strong positivity for CD20 (original magnification, $\times 400$).

I. A follow-up gadolinium-enhanced MR image 230 days after initial MR study reveals near complete resolution of MALT lymphoma of the right medial rectus muscle (arrow).

Note.—MALT = mucosa-associated lymphoid tissue, SUV_{max} = maximum standardized uptake value, ^{18}F -FDG PET = ^{18}F -fluorodeoxyglucose positron emission tomography

though the patient was euthyroid. CT of the orbits simply showed enlargement of the medial rectus muscle with lateral displacement of the globe. Byard et al. (2) reported a case of MALT lymphoma isolated to a single extraocular muscle, presenting as an approximately 30×15 -mm-sized mass arising from the region of the left superior rectus muscle with no signs of bone erosion. Benetatos et al. (3) reported a case of MALT lymphoma in which Gd-enhanced MR imaging revealed tumefaction of the inferior rectus muscle with subsequent decrease of tumor extent on follow-up study 6 months after rituximab treatment.

Our case represents the first case in which imaging findings at CT, MRI, and ^{18}F -FDG PET/CT were presented together in a single case of this rare entity. In our case, the medial rectus muscle which involved the MALT lymphoma was manifested as a fusiform enlargement of the muscle on CT and MRI, which resembled findings of thyroid orbitopathy. However, the differentiation from thyroid orbitopathy was established by involvement of the tendinous insertion in MALT lymphoma of our case. ^{18}F -FDG PET/CT findings of MALT lymphoma involving the EOM have not appeared in the English literature. The SUV_{max} of MALT lymphoma involving the medial rectus in our case was 4.9 g/mL, which was similar to that of MALT lymphoma elsewhere (9). We hypothesized that the low FDG uptake ($\text{SUV}_{\text{max}} = 4.9$ g/mL) of MALT lymphoma involving the medial rectus muscle in our case might be related to the indolent course of this entity.

Although MALT lymphomas respond well to surgical removal, radiotherapy is the treatment of choice for localized disease in the orbit/ocular adnexa. A dose of 30-35 Gy has been reported to be sufficient to provide local control and cure of the disease localized to the orbit/ocular adnexa and is associated with excellent survival (10).

In summary, it should be noted that the EOM is a rare location for the involvement of MALT lymphoma, and fusiform enlargement of EOM in MALT lymphoma may mimic that of thyroid orbitopathy on CT or MRI. The differentiation can be established by the fact that the tendinous insertion of EOM is involved in MALT lymphoma, which is not the case in the thyroid orbitopathy. In cases with solitary muscle enlargement without features

of thyroid orbitopathy, muscle biopsy should be considered to exclude MALT lymphoma.

REFERENCES

1. Rossman D, Michel R, Codere F. A case of an enlarged medial rectus muscle. *Int Ophthalmol* 2009;29:319-321
2. Byard SD, Chowdhury HR, Lee RM, Hyer J, Hart-George AL. Unilateral isolated extraocular muscle lymphoma. *BMJ Case Rep* 2012;2012:bcr0220125750
3. Benetatos L, Alymara V, Asproudis I, Bourantas KL. Rituximab as first line treatment for MALT lymphoma of extraocular muscles. *Ann Hematol* 2006;85:625-626
4. Abalo-Lojo JM, Baleato-Gonzalez S, Abdulkader I, Gonzalez F. Extraocular muscle involvement in MALT lymphomas. *Orbit* 2011;30:186-188
5. Izambart C, Robert PY, Petellat F, Petit B, Gastaud P, Lagier J, et al. Extraocular muscle involvement in marginal zone B-cell lymphomas of the orbit. *Orbit* 2008;27:345-349
6. Lacey B, Chang W, Rootman J. Nonthyroid causes of extraocular muscle disease. *Surv Ophthalmol* 1999;44:187-213
7. Raderer M, Streubel B, Woehrer S, Puespoek A, Jaeger U, Formanek M, et al. High relapse rate in patients with MALT lymphoma warrants lifelong follow-up. *Clin Cancer Res* 2005;11:3349-3352
8. Lee JL, Kim MK, Lee KH, Hyun MS, Chung HS, Kim DS, et al. Extranodal marginal zone B-cell lymphomas of mucosa-associated lymphoid tissue-type of the orbit and ocular adnexa. *Ann Hematol* 2005;84:13-18
9. Zhao K, Luo YZ, Zhou SH, Dai BL, Luo XM, Yan SX, et al. ^{18}F -fluorodeoxyglucose positron emission tomography/computed tomography findings in mucosa-associated lymphoid tissue lymphoma of the larynx: a case report and literature review. *J Int Med Res* 2012;40:1192-1206
10. Pfeffer MR, Rabin T, Tsvang L, Goffman J, Rosen N, Symon Z. Orbital lymphoma: is it necessary to treat the entire orbit? *Int J Radiat Oncol Biol Phys* 2004;60:527-530

내직근을 침범한 점막 연관성 림프조직 림프종의 CT, MRI 및 ^{18}F -Fluorodeoxyglucose Positron Emission Tomography/CT 영상소견: 증례 보고¹

이상권¹ · 최미선²

저자들은 47세 남자의 내직근을 침범한 점막 연관성 림프조직(mucosa-associated lymphoid tissue: MALT) 림프종 1예를 CT, MRI, ^{18}F -fluorodeoxyglucose positron emission tomography/CT (^{18}F -FDG PET/CT) 및 병리학적 소견과 함께 보고하고자 한다. 병변은 힘줄부착부위의 침범을 동반한 우측 내직근 단독의 방추형 종대로 발현하였으며, 비조영증강 CT 영상에서 뇌와 비슷한 감쇠를, 지방포화 고속스핀에코 T1- 및 T2-강조영상에서 회질과 등신호강도를 보였으며, 조영증강 CT 및 MR 영상에서 균질한 조영증강을 보였다. ^{18}F -FDG PET/CT에서 maximum standardized uptake value는 4.9 g/mL였다. 우측 내직근의 생검에 의한 검체의 조직학적 및 면역조직화학적 검사의 결과는 MALT 림프종과 일치하였다. 외안근은 MALT 림프종에 의해 드물게 침범될 수 있으며, 외안근의 MALT 림프종은 갑상선 안병증을 닮을 수 있다.

계명대학교 의과대학 동산의료원 ¹영상의학과, ²병리과