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Primary MALT lymphoma of lacrimal gland in association with Sjögren syndrome: assessment with MR imaging

Case Report

Sjögren Sendromuyla Birlikte Görülen Lakrimal Gland Lenfoması: MR ile Değerlendirme

Olgu Sunumu

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ABSTRACT

We describe the case of a Sjögren syndrome presented with bilateral lacrimal gland enlargement. The histopathological examination revealed MALT lymphoma on one side and lymphoepitelial disease on the other side. The discrimination of these two entities was not possible with magnetic resonance imaging (MRI). However the involvement was clearly shown. Patients with Sjögren syndrome are at increased risk of lymphoma development. MRI clearly depicts the orbital involvement and should be the first diagnostic tool for the diagnosis of orbital imaging in these patients.

Key words: *MALT lymphoma, MRI*

ÖZET

Bu olgu sunumunda Sjögren sendromlu olgudaki bilateral lakrimal gland büyümesini tanımladık. Histopatolojik değerlendirmede bir tarafta MALT lenfoma, diğerinde lenfoepitelyal hastalık tutulumu belirlendi. Bu iki antite manyetik rezonans görüntüleme (MRG) ile ayırt edilemedi, ancak tutulum net bir şekilde belliydi. Sjögren sendromlu olgular lenfoma gelişimi açısından artmış bir riske sahiptir. MRG orbital tutulumu açık bir şekilde belirler ve orbital görüntülemede ilk tercih edilecek yöntem olmalıdır.

Anahtar Kelimeler: *MALT lenfoma, MRG*

INTRODUCTION

Sjögren syndrome is a chronic inflammatory disorder of probable autoimmune nature characterized by infiltration of the exocrine glands, particularly the salivary and lacrimal glands, by lymphocytes and plasma cells. The classic signs of the Sjögren syndrome, therefore, includes enlargement of the parotid glands with mucosal dryness manifest by xerostomia and xerophthalmia (1). Malignant or pseudomalignant lymphoproliferation may be a prominent part of the illness, especially in primary Sjögren's syndrome (1,2). In this paper, we present magnetic resonance imaging findings of bilateral lacrimal gland enlargement in a patient with Sjögren syndrome.

CASE REPORT

A 43-year-old female patient who had a history of primary Sjögren syndrome for 5 years, was presented with the enlargement of bilateral lacrimal glands apparent on the left. Magnetic resonance imaging (MRI) was performed on a 1.5 T scanner with a head coil. Imaging protocol included axial T1-weighted spin-echo (SE) , fat saturated T2-weighted SE and contrast-enhanced fat saturated T1-weighted SE images. Both glands showed isointensity to extraocular muscles on T1-weighted images and slight hyperintensity to vitreous on T2-weighted images (Fig. 1B and C). On postcontrast fat-saturated T1-weighted images, diffuse homogeneous enhancement was seen in both glands (Fig.1D). Following MR examination, an excisional biopsy of bilateral lacrimal glands was performed. Histopathologic examination showed lymphoepithelial disease on the right and mucosa-associated lymphoid tissue (MALT) lymphoma on the left lacrimal glands.

Figures: *Bilateral lacrimal gland enlargement due to lymphoepithelial disease on the right and mucosa-associated lymphoid tissue (MALT) lymphoma on the left.*

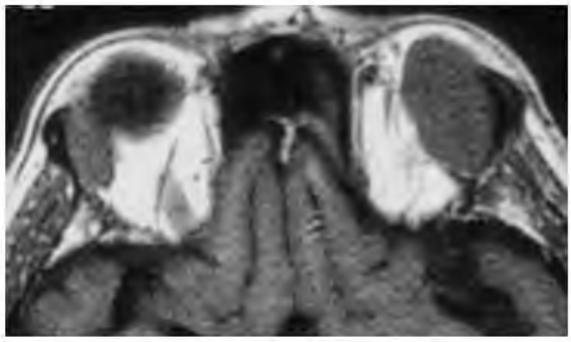

Figure 1-A: *Axial T1-weighted MR image shows both glands to be isointense to muscles.*

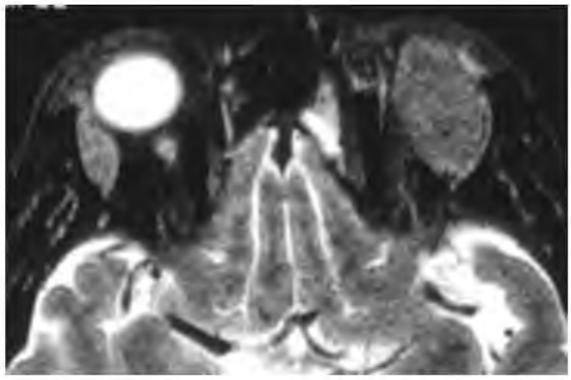

Figure 1-B: *Axial fat saturated T2-weighted MR mage shows both glands to be hypointense to vitreous.*

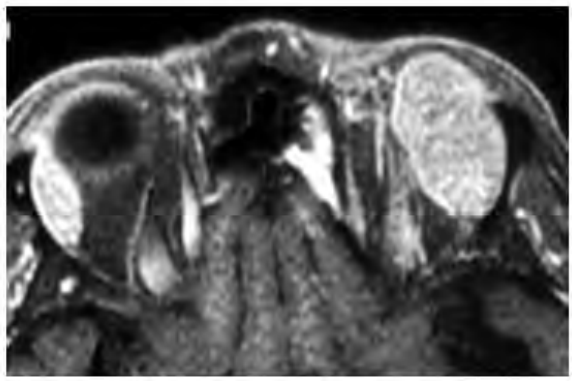

Figure 1-C: *Axial fat saturated contrast-enhanced T1-weighted MR image shows diffuse homogeneous enhancement in both glands.*

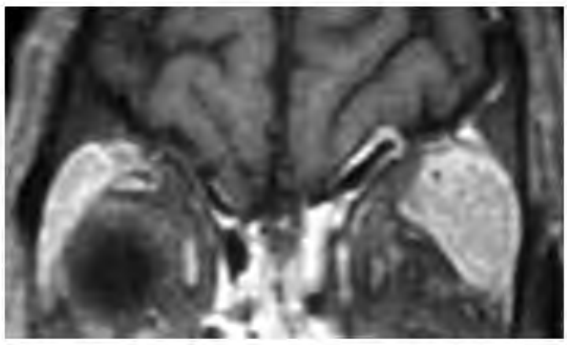

Figure 1-D: *Coronal fat saturated contrast-enhanced T1-weighted MR image shows diffuse homogeneous enhancement in both glands.*

DISCUSSION

Sjogren's syndrome is an important autoimmune disease arising in the head and neck region. The disease affects predominantly middle-aged women, in the peri- or post-menopausal period, in a female-to-male ratio of 9:1. However, it can be seen in both sexes and all ages. The prevalence is still unknown. Nevertheless, it is considered to be quite common since in addition to the primary syndrome, 30% of patients with rheumatoid arthritis, systemic lupus erythematosus and systemic sclerosis suffer secondary Sjögren's syndrome (1,2).

Patients have an increased arrival risk of up to 6% per year for developing B-cell lymphomas, including mucosa-associated lymphoid tissue (MALT) lymphomas (3,4). In the study that was carried out by Tonami H et al (5) they have found 27 lymphomas among 463 patinets with Sjogren syndrome. Six of these lymphomas were MALT lymphomas and two of them were localized in the lacrimal glands (5).

The evolution from lymphoepithelial disease characteristic of Sjögren syndrome to pseudolymphoma and malignant lymphoma is probably a multistep process (6). Lymphoepithelial disease is generally composed of a mixture of polyclonal B- and T-cells. In contrast, malignant lymphoma represents neoplastic, monoclonal B-cell proliferations. So-called pseudolymphoma has been considered an intermediate stage in the transition from benign to malignant lymphoproliferation (7). To date, various histologic subtypes of malignant lymphoma for these patients with Sjögren syndrome have been described, including follicular center lymphoma, diffuse large B-cell lymphoma, immunoblastic lymphoma and especially MALT lymphoma (7). In our study, the histologic subtype of the case with malignant lymphoma was MALT lymphoma. MALT lymphoma, characterized by low-grade B-cell lymphoma containing centrocyte-like cells and lymphoplasmocytoid cells, has been found in the mucosa and ducts of a variety of organs, including the gastrointestinal tract and lung along with some endocrine and exocrine glands (8).

They are increasingly recognized as a distinct clinical-pathologic entity among the non-Hodgkin's lymphomas (9). MALT lymphomas of the lacrimal gland, like their counterparts in other sites of the body, have a tendency to remain localized for prolonged times, have a less aggressive course and respond well to therapy (10). It must be kept in mind that low-grade lymphomas including MALT lymphomas can transform into high-grade large cell lymphomas. Voulgarelis et al. reported that although the majority of lymphomas in patients with Sjögren syndrome were marginal zone B-cell lymphomas of low-grade, they evolve toward a less-differentiated cell type and that high-grade lymphoma is significantly associated with worse survival (7,8). Because the prognosis of malignant lymphoma depends mainly on early diagnosis, a reliable noninvasive diagnostic procedure is necessary to differentiate benign lymphoproliferative disorders and malignant lymphoma. As the progression from benign lymphoproliferative disorders to malignant lymphoma is concerned,

suspicious lacrimal gland lesions should be biopsied following clinical enlargement (11).

The differential diagnosis of lacrimal gland enlargements include many pathologies. The most important point of evaluation is the detection of whether enlargement is unilateral or bilateral. Unilateral enlargement is mostly caused by tumors which make up about half of all the masses seen in lacrimal glands. Benign pleomorphic adenomas and mixed tumors are common. Malignant tumors are either primary or metastatic. Dacryoadenitis and pseudotumor are both inflammatory causes of lacrimal enlargement. Dermoid cysts may also cause unilateral enlargement. The causes for bilateral enlargement of the lacrimals are most often systemic disorders, though any cause of unilateral enlargement can also cause bilateral enlargement of the lacrimals. The causes are sarcoidosis, Mikulicz's syndrome, Sjögren syndrome, Graves disease, myxedema, amyloidosis, blood dyscrasias, pseudotumor and lymphoid hyperplasia (12,13).

CONCLUSION

In conclusion, MR imaging is useful in the detection of the lacrimal gland enlargement in patients with Sjögren syndrome, although no specific imaging features are ascribed to these lesions. For the precise diagnosis histopathological examination is necessary.

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