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Orbital Lymphoma of Mucosa-Associated Lymphoid Tissue in a Patient With Rheumatoid Arthritis

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Abstract: A 75-year-old male with rheumatoid arthritis complained of proptosis of the left eye. On examination, 6-mm left proptosis was seen. A hyperemic and swollen optic disc was visible in the left fundus. Laboratory test results showed positive rheumatoid factor. Computed tomography and magnetic resonance imaging showed a mass lesion in the left orbit. Histopathologic study of the excised specimen revealed centrocyte-like cells and lymphoplasmocytoid cells. Immunohistochemical study showed positive staining of CD 20 (B cells) and lambda light chain (immunoglobulin). The combination of rheumatoid arthritis and lymphoma of mucosa-associated lymphoid tissue in the orbit, as demonstrated in our patient, is rare. **Jpn J Ophthalmol 1998;42:223–226** © 1998 Japanese Ophthalmological Society

Key Words: Orbital lymphoma, rheumatoid arthritis.

Introduction

The association of malignant lymphoma and autoimmune disease including rheumatoid arthritis and Sjögren syndrome is well known.[1–8] Malignant lymphomas of mucosa-associated lymphoid tissue (MALT), characterized by low-grade B-cell lymphoma containing centrocyte-like cells and lymphoplasmocytoid cells, have been found in the gut, lung, salivary gland, thyroid, orbit and conjunctiva.[9–14] Mucosa-associated lymphoid tissue lymphomas are clinically indolent and often remain localized.[12] To our knowledge, however, patients with rheumatoid arthritis and MALT lymphoma in the orbit have rarely been reported. We recently examined a patient with such a rare association.

Case Report

A 75-year-old man was first seen in our clinic complaining of painless proptosis in the left eye on August 8, 1995. The patient's family history was noncontributory. At age 55 years he was diagnosed as having rheumatoid arthritis, but he received no treatment.

On examination, his visual acuity was 1.0 OU. His intraocular pressure was 19 mm Hg OU in both eyes. Hertel exophthalmometry revealed 6-mm left proptosis. The eye movement was not disturbed. The corneas and anterior chambers appeared clear bilaterally. Cortical opacities were noted in both lenses. The right fundus appeared normal ophthalmoscopically. A hyperemic and swollen optic disk was found in the left fundus. Schirmer test results revealed normal lacrimation bilaterally. Goldmann visual field testing showed an enlarged blind spot in the left eye. Ultrasonography revealed a retrobulbar abnormal shadow in the left orbit.

Results of laboratory tests disclosed positive rheumatoid factor (1+) and antinuclear antibody (cutoff index, 1.2; normal, <0.7). Other results, including blood cell counts, blood chemistry, blood pressure, Treponema pallidum hemagglutinin, chest x-ray, and urinalysis, were negative or within normal range. Finger deformity was found in both hands. Computed tomography showed a retrobulbar lesion in the left orbit. On magnetic resonance imaging (Figure 1), the orbital lesion appeared isointense on T_1-weighted image. The lesion was enhanced after administration of gadolinium. No other uptake than the left orbit was found on Gallium scintigraphy.

On August 14, 1995, the patient underwent an incisional biopsy of the lesion in the left orbit. During

Received: June 18, 1997
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Jpn J Ophthalmol 42, 223–226 (1998)

Published by Elsevier Science Inc.

0021-5155/98/$19.00
PII S0021-5155(97)00135-4

Jpn J Ophthalmol
Vol 42: 223–226, 1998

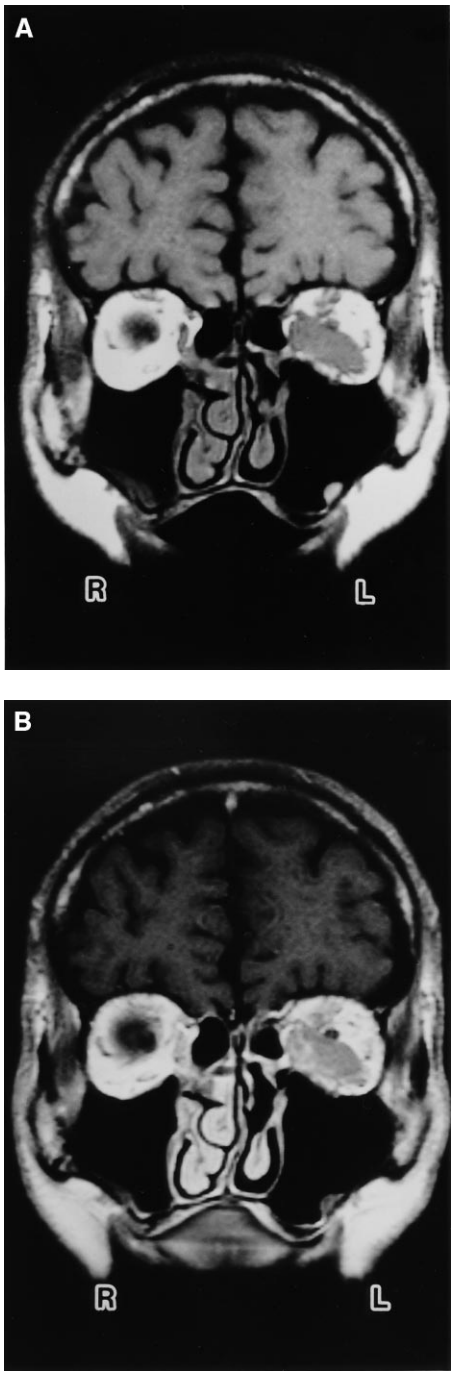

Figure 1. Magnetic resonance imaging. (**A**) The retrobulbar lesion appears isointense in the vertically sectioned T_1-weighted image. (**B**) The lesion in the left orbit is enhanced after administration of gadolinium.

the surgery, no encapsulation of the lesion was found. Histopathologic study of the specimen showed centrocyte-like cells and lymphoplasmocytoid cells (Figure 2). Immunohistochemical study of the specimen revealed positive staining of CD 20 (Dako, Carpinteria, CA, USA) (specific for pan B cells) (Figure 3) and lambda light chain (Dako) (specific for immunoglobulin lambda light chain) but negative staining of UCHL-1 (P Beverley) (specific for T cells) and kappa light chain (Dako) (specific for immunoglobulin kappa light chain). The histopathologic and immunohistochemical findings were compatible with a MALT-type B-cell lymphoma.

The patient was treated with a total of 30 Gy of radiation to the left orbit from October to December of 1995. After radiotherapy, the lesion in the left orbit decreased markedly, and the left disc edema disappeared. No recurrence of the orbital mass lesion was noted during the follow-up period of 20 months.

Discussion

Our patient had positive rheumatoid factor, finger deformity, left proptosis, an orbital mass lesion on magnetic resonance imaging, an ophthalmoscopically visible disc edema, histopathologically proved centrocyte-like cells, and immunohistochemically verified B-cell lymphoma. It is possible that the disc edema resulted from compression by the large mass lesion, because the edema disappeared after radiotherapy.

The association of autoimmune disease and malignant lymphoma, in particular, a combination of rheumatoid arthritis and B-cell malignant lymphoma, is well known.[1–8] Isomaki et al[6] reported that of 46,101 patients with rheumatoid arthritis in Finland, 130 developed lymphoproliferative disorders. The authors showed that the relative risk for lymphoma in rheumatoid arthritis patients was 2.7 compared with the general population.[6] Prior et al[7] reported that of 489 patients with rheumatoid arthritis in England, six developed lymphoproliferative disorders. They showed that the relative risk for lymphoma in rheumatoid arthritis patients was 15 compared with the general population.[7] To our knowledge, however, patients with an association of rheumatoid arthritis and malignant lymphoma in the orbit have rarely been reported. Some patients with an association between autoimmune disease and MALT lymphoma have been described. Hyjek and Isaacson[15] reported a close association between Hashimoto's thyroiditis and MALT lymphoma. Hyjek et al[16] further reported an association of B-cell lymphoma and myoepithelial sialadenitis. Strelkauskas et al[17] suggested that a regulatory subset of lymphocytes is missing in patients with juvenile rheumatoid arthritis. Aozasa[18] speculated that the presence of long-standing inflammation may have been involved in a case of MALT

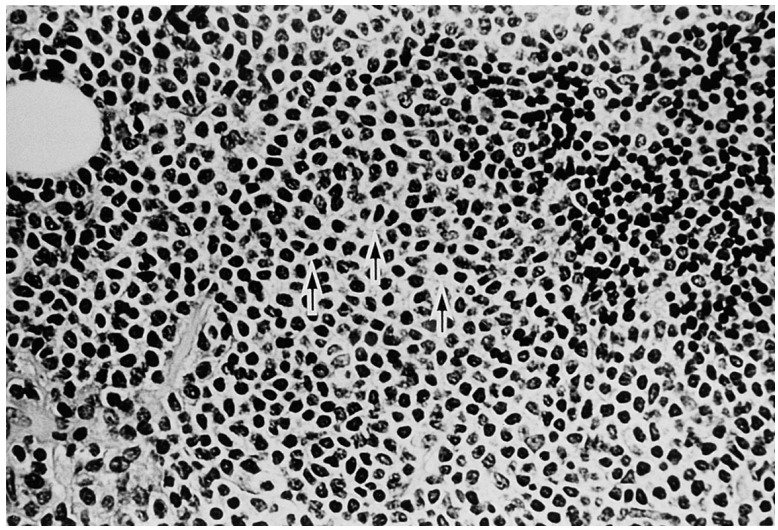

Figure 2. Histologic study of the specimen shows centrocyte-like cells (arrows) and lymphoplasmocytoid cells with hyperchromatic nuclei (hematoxylin-eosin, ×200).

lymphoma. Our patient had long-standing untreated rheumatoid arthritis. The rheumatoid arthritis might have played a role, in part, in the development of MALT lymphoma in our patient, although the exact pathogenesis is unclear. A combination of rheumatoid arthritis and MALT lymphoma in the orbit, as demonstrated in our patient, is rare.

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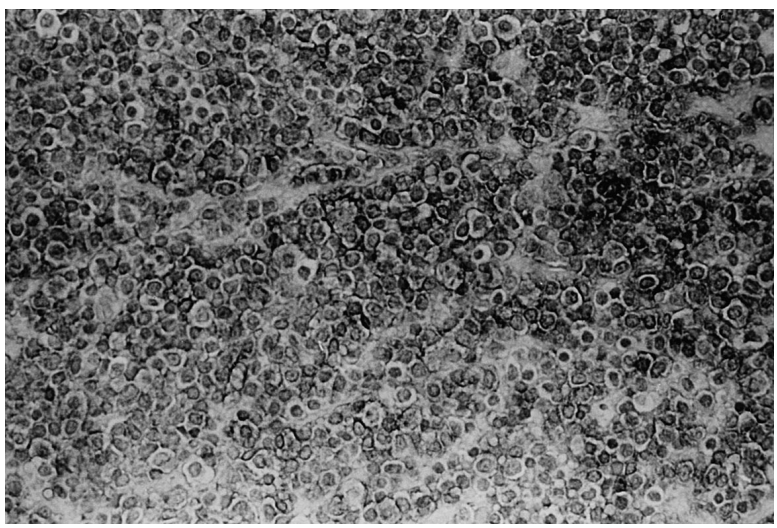

Figure 3. Immunohistochemical examination of the specimen reveals strongly positive staining of cell membranes of neoplastic centrocyte-like cells with CD 20 (×200).

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