INTRODUCTION

Extranodal lymphoma of mucosa-associated lymphoid tissue (MALT) was first described by Isaacson and Wright in 1983 (1) and was recently reclassified as “extranodal marginal zone B-cell lymphoma (MZBL)” according to the REAL (2) and WHO classifications (3). Primary MZBLs of MALT-type have been described in a variety of sites, including gastrointestinal tract (1), thyroid gland (4), lung (5), salivary gland (6), and ocular adnexa (7) associated with clinical setting of autoimmune diseases or chronic inflammation, usually Sjogren’s syndrome, Hashimoto’s thyroiditis, and Helicobacter pylori infection. Thymic MZBL of MALT-type is an extremely rare disease with only 22 cases having been reported in the literature to date and the largest series has included 15 cases (8-19). In this paper, I report a rare case of primary thymic MZBL of MALT-type arising in the thymus in a patient with Sjogren’s syndrome and rheumatoid arthritis.

CASE REPORT

A 43-yr-old Korean woman presented with a long history of Sjogren’s syndrome and rheumatoid arthritis. She had complained of dry eyes, dry mouth, and arthralgia for about 16 yr and also complained of both parotid swelling that had developed 3 months before. The symmetrical swelling of the wrist, metacarpophalangeal, and proximal interphalangeal joints was accompanied by morning stiffness and pain. On admission, antinuclear antibody (ANA) revealed a speckled pattern and showed positive reaction for SS-A/Ro antibody and negative for the SS-B/La antibody. Rheumatoid factor was positive (147.3 IU/mL). A lobulated anterior mediastinal mass was detected by computerized tomography (Fig. 1) and the thymus was resected through median sternotomy. There were no symptoms of myasthenia gravis. The resected mass was confined within the thymus and did not show invasion to the surrounding structures. The specimen consisted of a solid, smooth, lobulated, encapsulated mass that measured 7 cm in maximum diameter and weighed 79 g. The cut surface was pale pink to tan, and showed homogenous appearance with some small cysts (Fig. 2). The tissue was fixed in 10% neutral buffered formalin and embedded in paraffin. Sections were stained with hematoxylin and eosin and periodic acid-Schiff. Histologically, the normal architecture of the thymus was diffusely effaced by a dense infiltration of lymphoid cells. Reac-
tive lymphoid follicles with germinal centers could be identified within the tumor (Fig. 3A). The microscopic cysts filled with eosinophilic proteinaceous fluid were lined by attenuated epithelium infiltrated by centrocyte-like (CCL) cells (Fig. 3B). The lymphoid infiltrate comprised monotonous CCL cells with monocytoid features (Fig. 3C, D). There were considerable numbers of plasma cells and small lymphocytes. A few large transformed cells were also recognized (Fig. 3C, D).
Extranodal Marginal Zone B-cell Lymphoma of MALT-type

In type B thymomas, the lymphoid cells usually consist of lymphocytes which could be confused with more common thymoma. Poor prognosis. The mixture of epithelial elements and small lymphocytes, CCL cells, a few blastoid cells, and plasma cells showing mostly B-cell immunophenotype. In summary, I described herein is a thymic low-grade extranodal MZBL of MALT-type in a patient with both Sjögren’s syndrome and rheumatoid arthritis. It is possible that the lymphoma arose on the underlying basis of long standing autoimmune diseases. Pathologists should be aware that MZBL of MALT-type may occasionally involve thymus. In a small biopsy, the presence of mixture of small lymphoid cells and epithelial cells may closely mimic a thymoma. Morphologic features of CCL cells, plasmacytoid cells, lymphoepithelial lesions as well as immunophenotype or molecular studies are helpful for a definitive diagnosis.

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