Thymic mucosa-associated lymphoid tissue lymphoma involving lymph nodes

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INTRODUCTION: Thymic mucosa-associated lymphoid tissue (MALT) lymphoma involving lymph nodes is quite rare with only 13 previous cases reported in the literature.

PRESENTATION OF CASE: The 33-years-old female was referred to our department for the investigation of abnormalities on computed tomographic (CT) scans. CT scans showed a 9-cm × 3-cm mass composed of a mixture of soft tissue and fat at the anterior mediastinum with lymphadenopathy in the neck, axillary and mediastinal regions. She was underwent complete surgical resection of the mass with regional lymph node dissection through a median sternotomy. Histological examination of the surgical specimens confirmed the diagnosis of MALT lymphoma arising in the thymus with nodal metastasis. She achieved complete remission after postoperative rituximab combined chemotherapy.

DISCUSSION: Thymic MALT lymphoma occurs most frequently in Asian female aged 40–60 years and commonly appears anterior mediastinal masses on CT scans. The excised tissue is necessary to confirm the accurate histological diagnosis. The disease usually remains localized for a long time, making local surgical resection highly effective. However, when the lymph nodes are involved, effective treatment approaches of the disease is still undefined.

CONCLUSION: We report a case of thymic MALT lymphoma involving lymph nodes, in which the patient was successfully treated with primary site resection with regional lymph node dissection followed by rituximab combined chemotherapy. Surgery provided not only a useful approach for collecting tissue for an accurate histological diagnosis, but also an effective local treatment, even in the case of advanced-stage thymic MALT lymphoma.

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1. Introduction

Mucosa-associated lymphoid tissue (MALT) lymphoma, or extranodal marginal zone lymphoma of MALT, is a low-grade B-cell non-Hodgkin's lymphoma.1–7 MALT lymphoma occurs most frequently in the Waldeyer ring, gastrointestinal tract, skin, lungs, thyroid, salivary glands, and orbit; thymic MALT lymphoma is quite rare. In fact, approximately 50 cases of thymic MALT lymphoma have been reported in the literature.1–7 These reports demonstrate that thymic MALT lymphoma usually grows locally and responds well to surgery, and the involvement of lymph nodes is unusual, with only 13 prior cases reported in the literature.5,6 Because of the rarity of advanced-stage thymic MALT lymphoma, a standard treatment for the disease has not been fully defined yet.

Here, we report a case of thymic MALT lymphoma involving the lymph nodes, which was successfully managed with complete surgical resection with regional lymph node dissection followed by rituximab combined chemotherapy.

2. Presentation of case

A 33-year-old female with no history of smoking was referred to our department for the investigation of an anterior mediastinal mass, detected incidentally on a postoperative follow-up computed tomographic (CT) scan for thyroid cancer. She underwent total thyroidectomy with bilateral modified radical neck dissection for papillary thyroid carcinoma 3 years ago: Stage I (T1N0M0). There was no evidence of recurrent thyroid cancer.

On admission, she was asymptomatic. Her body temperature was 36.5 °C, blood pressure was 112/73 mmHg, and pulse was 85 beats per min. A surgical scar was noted on her neck. There were no abnormalities of the chest or abdomen and no neurological abnormalities. Laboratory data showed slightly elevated serum lactate dehydrogenase (239 IU/L). Her thyroid levels were normal since she had been receiving thyroid hormone replacement therapy. Serum immunoglobulin levels were found to be within the normal range and autoantibodies were not detected.

CT scans showed a 9-cm × 3-cm mass composed of a mixture of soft tissue and fat at the anterior mediastinum (Fig. 1). The central part of the mass was irregularly enhanced by contrast medium. Lymphadenopathy was found in the neck, axillary and mediastinal regions, but not in abdominal or inguinal regions. Surgical excision of the tumor through a median sternotomy was performed because

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MALT lymphoma arising in the thymus was mostly suspected on the basis of image findings.

During surgery, an elastic soft mass in the anterior mediastinum was easily removed from the surrounding tissue. Intra-operative pathologic examination of the specimen obtained from left lobe thymectomy showed infiltration of lymphoid cells around Hassall’s corpuscle, suggesting thymic MALT lymphoma. Therefore, complete surgical resection with regional lymph node dissection was performed.

The resected tumor, which was white-gray in color, measured 10.5 cm × 7 cm × 2 cm and weighed 82 g. Histological examination showed that the normal architecture of the thymus was diffusely effaced by a dense infiltration of CD20-positive lymphoid cells (Figs. 2 and 3). No local invasion into surrounding tissue was found. Lymphoepithelial lesions were formed by centrocyte-like cells infiltrating the Hassall’s corpuscles. Lymphoma cells were found in the resected lymph nodes.

The mediastinal tumor was diagnosed as a MALT-type B-cell lymphoma. Lymphadenopathy in the neck and axillary regions was suspected to have relevance to nodal metastasis of MALT lymphoma. Bone marrow aspiration revealed no infiltration by lymphoid cells and no chromosomal abnormalities. The patient’s postoperative course was uneventful, and she was discharged on the 10th postoperative day. She received 8 courses of R-CHOP therapy (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisolone) and achieved complete remission without severe adverse reactions 6 months after operation. She has been well for 5 years without relapse.

3. Discussion

Thymic MALT lymphomas often take an indolent course and remain localized for prolonged periods, making local surgical excision highly effective. However, an optimal strategy in the management of thymic MALT lymphoma with nodal metastasis is still controversial. In the present case, thymic MALT lymphoma was diagnosed by surgical specimens, and the patient responded well to surgical treatment and postoperative R-CHOP therapy. Surgery facilitated the establishment of an accurate histological diagnosis and provided an effective local treatment for the primary tumor, even in advanced-stage thymic MALT lymphoma.

Thymic MALT lymphoma has unique characteristics compared with MALT lymphoma in other organs. For example, thymic MALT lymphomas occur most frequently in Asian female aged 40–60 years and usually appear on CT scans as anterior mediastinal masses with multiple solid cystic regions. In some cases, pulmonary amyloid nodules associated with polycyonal hyperglobulinemia are found and immunological disorders, particularly Sjögren syndrome, are thought to be associated with thymic MALT lymphoma tumorigenesis. Moreover, prolonged antigenic stimulation by autoantigens or chronic inflammation in patients with thymic lymphoid hyperplasia may allow the development of MALT lymphoma. Although API2-MALT1 fusion resulting from t(11;18)(q21;q21) is present in up to 50% of MALT lymphomas in general, this chromosomal translocation is not detected in thymic MALT lymphoma.

Thymic MALT lymphoma is usually diagnosed by histological examination of surgical specimens. In adults, primary mediastinal lymphomas, which represent approximately 5–7% of mediastinal tumors operated on in Japan, are generally diagnosed as Hodgkin’s lymphoma, primary mediastinal large B-cell lymphoma, or thymic MALT lymphoma using histology and immunohistochemistry. Thymic MALT lymphoma is difficult to diagnose from small tissue fragments obtained by percutaneous needle biopsy. Therefore, surgical resection is a suitable alternative approach to obtaining enough tissue for accurate histological diagnosis.

Postoperative recurrence after complete surgical resection is considered to be rare since thymic MALT lymphoma generally presents as a nonmetastasized completely encapsulated tumor. Only 3 cases of postoperative recurrence of thymic MALT lymphoma have been reported to date; one case of bone marrow
metastasis\(^2\) and 2 cases of distant lymph node metastasis.\(^6\) For these reasons, Sunohara et al.\(^5\) recommended that total thymectomy through a median sternotomy should be an effective procedure for removal of the primary lesion. In addition, Yamada et al.\(^13\) recently reported a minimally invasive tumor dissection procedure using video-assisted thoracoscopic surgery. However, these techniques require careful attention due to lack of lymph node dissection. Lymphoma cells often involved the regional lymph nodes.\(^2\) Patients without recurrence have generally received postoperative radiation therapy, indicating that remnants in the regional lymph nodes may be managed with that treatment.\(^2\) Therefore, total thymectomy or tumor dissection may result in failure to remove involved regional lymph nodes completely, potentially leading to postoperative recurrence. Thus, we recommended en bloc resection of thymic MALT lymphoma and mediastinal lymph nodes for local treatment.

A commonly accepted treatment strategy for thymic MALT lymphoma involving lymph nodes has not been established.\(^5\) In the current case, we used primary site resection followed by rituximab combined chemotherapy to achieve complete remission of advanced-stage thymic MALT lymphoma. To the best of our knowledge, this is the first report of successful treatment of rituximab combined chemotherapy for the disease. Rituximab, a chimeric monoclonal antibody targeting CD20, is a highly effective agent in B-cell non-Hodgkin’s lymphoma, and several clinical trials are currently evaluating the activity of the combination of rituximab and chemotherapy in MALT lymphoma.\(^14,15\) Our case suggests that rituximab combined chemotherapy would be effective with low toxicity in thymic MALT lymphoma.

4. Conclusion

Here, we reported a rare case of thymic MALT lymphoma involving lymph nodes that was successfully managed by complete surgical resection with regional lymph node dissection followed by postoperative rituximab combined chemotherapy. In this case, surgery facilitated histological diagnosis and local treatment of the primary tumor.

Conflict of interest statement

None.

Funding

None.

Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief on this journal on request.

Author contribution

All authors contributed.

References