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Co-occurrence of papillary thyroid cancer and MALT lymphoma of the thyroid with severe airway obstruction: A case report and review of the literature

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ABSTRACT

INTRODUCTION: Co-occurrence of papillary thyroid carcinoma (PTC) and mucosa-associated lymphoid tissue (MALT) lymphoma resulting in severe airway obstruction is very rare.
PRESENTATION OF CASE: A 58-year-old woman visited our department because of enlargement of a neck mass. Computed tomography (CT) and ultrasonography showed 2 discrete hypoechoic nodules. Fine-needle aspiration biopsy revealed thyroid lymphoma in the left lobe and PTC in the right lobe. After 1 week, she returned to the emergency room at our hospital with shortness of breath and difficultly in swallowing. CT revealed enlargement of the left lobe, which was severely compressing the trachea. We performed emergency total thyroidectomy with lymphadenectomy. The postoperative course was uneventful, and the patient was discharged without any symptoms.
DISCUSSION: The most common treatment for PTC is surgery; however, the treatment for thyroid lymphoma remains controversial. We propose that surgery be performed in the cases of symptoms such as shortness of breath and difficulty in swallowing.
CONCLUSION: We performed emergency total thyroidectomy to relieve obstruction of the trachea and to remove the two malignant tumors. We suggest total thyroidectomy for a case of co-occurrence of two malignant tumors, causing severe airway obstruction.

1. Introduction

Papillary thyroid carcinoma (PTC) is the most common thyroid cancer and accounts for nearly 80% of all cases of thyroid cancer. It occurs most predominantly in women (female to male ratio, 3:1), usually presents in the third or fourth decade of life, and is associated with an excellent prognosis.[1,2] Mucosa-associated lymphoid tissue (MALT) lymphoma of the thyroid gland is rare, accounting for between 0.6% and 5% of all cases of thyroid cancer.[1,3] Diagnoses of PTC and MALT lymphomas are based on histologic features. MALT lymphomas are strongly associated with Hashimoto thyroiditis (HT), whereas there does not seem to be a strong association with PTC. The simultaneous occurrence of both malignancies in the same patient is rare. Further, cases with symptoms of shortness of breath and difficulty in swallowing due to severe compression of the trachea by the mass have not been reported previously.

2. Presentation of case

A 58-year-old woman was referred to our department because of progressive enlargement of the thyroid gland. She had a history of ovarian cancer which has not showed recurrence after surgery. Enlargement of the thyroid gland had been noticed when the patient was a teenager, but this had been treated conservatively. In March 2013, the patient noticed an progressive enlargement of the neck mass and was referred to our department. She did not report a change in speech, shortness of breath, or weight loss at that time. Upon physical examination, her thyroid gland was enlarged, with the left lobe being larger than the right. There were no palpable cervical lymph nodes. Other findings of the physical examination were unremarkable.

Laboratory examination indicated normal thyroid function, although the serum thyroglobulin antibody level was slightly elevated. Levels of serum markers for leukemia, such as beta-2 microglobulin and sIL-2R, were also normal. Thyroid ultrasonography revealed a heterogeneous thyroid gland. In particular, 2 discrete hypoechoic nodules were detected: one in the left lobe, measuring 47.3 mm × 36.4 mm × 51.2 mm, and the other in the right lobe, measuring 7.4 mm × 7.8 mm × 9.9 mm, with speckled calcification. Both nodules were subsequently examined by

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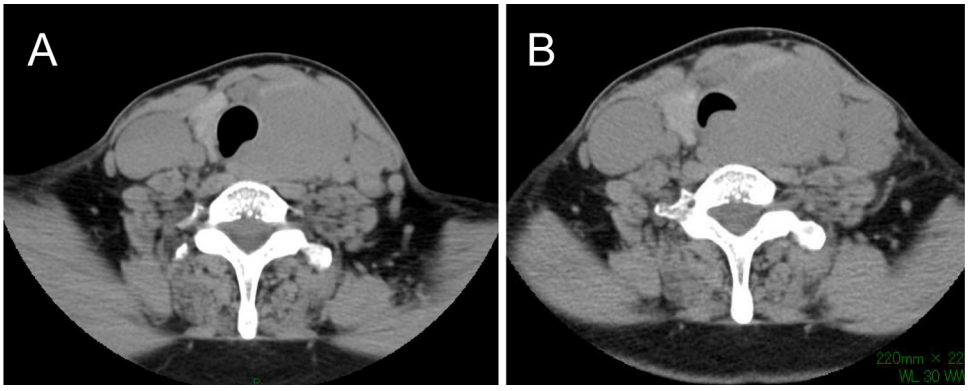

Fig. 1. Enhanced computed tomography (CT) revealed progressive enlargement of the left lobe, which had caused severe compression of the trachea compared to the CT scan 1 week before.

fine-needle aspiration (FNA) biopsy. The left nodule showed a predominance of lymphoid cells, associated with rare scattering of atypical cells. These lymphoid cells comprised a relatively uniform sample of intermediate-sized plasmacytoid lymphocytes/plasma cells, which suggested the possibility of a low-grade lymphoproliferative disorder. However, because of insufficient material for additional tests such as flow cytometry and immunohistochemistry, a definitive diagnosis could not be made. The aspirate from the right nodule was distinctively different from that taken from the left nodule. The sample extracted from the right nodule contained epithelial cells arranged in syncytial groups and microfollicles. Nuclei were enlarged, oval, and overlapping. Diagnostic nuclear features of PTC were observed, such as pale, powdery chromatin; nuclear grooves; small nucleoli; and intranuclear inclusions. The patient was scheduled to undergo further investigation, but after 1 week, she returned to the emergency room with shortness of breath and difficulty in swallowing. Upon physical examination, her thyroid had significantly increased in size since her last visit. Enhanced computed tomography (CT) revealed progressive enlargement of the left lobe, which had caused severe compression of the trachea (Fig. 1). Because of rapid enlargement of the thyroid, which resulted in severe trachea compression, surgical resection was inevitable. Chemotherapy and radiation therapy were not viable options in this case, given the requirement of an immediate response.

We performed emergency total thyroidectomy and lymph node adenectomy under general anesthesia. The right recurrent nerve was preserved, but the left recurrent nerve had to be resected because of severe adhesion to the left lobe of the thyroid. There was no invasion into the esophagus or trachea, and tumor resection was possible. Histological examination of the right lobe showed a predominant follicular pattern in the PTC (Fig. 2A). The tumor measured 10 mm × 8.0 mm, and no extrathyroidal extension or lymph node involvement was noted. Histopathological examination in the left lobe indicated extranodal marginal zone B-cell lymphoma of MALT type with extreme plasmacytic differentiation (Fig. 2B). Scattered reactive germinal centers were also observed. Immunohistochemical staining was performed, and the plasmacytic cells showed dominant expression of CD20+ (B cell marker) rather than of CD3+ (T cell marker) (Fig. 2C) and of the kappa light chain rather than that of the lambda light chain. The test for BCL-2 gene mutation was negative.

The postoperative course was good, and the patient did not show any further symptoms of shortness of breath or difficulty in swallowing. The function of the right vocal cord remained intact and required no intervention by postoperative laryngoscopy. She was discharged 10 days after surgery. One month later, positron emission tomography-CT did not reveal any abnormal accumulation and showed no evidence of recurrence of the lymphoma. The patient continues to be monitored, without the administration of chemotherapy or radiation.

3. Discussion

PTC is the most common type of thyroid cancer and is associated with a 20-year survival rate of more than 90%.[1] This type of thyroid cancer has distinct cytological features: sheets, papillary architecture, and/or microfollicles with characteristic nuclear enlargement; pale powdery chromatin; grooves; small but distinct nucleoli; and intranuclear cytoplasmic inclusions.[4] In this case, hypoechogenicity and the presence of microcalcification on ultrasonography supported the suspicion of PTC. Surgery is the mainstay of treatment and often results in high cure rates with an excellent prognosis.

Thyroid lymphoma accounts for between 0.6% and 5% of malignant tumors found in the thyroid and MALT lymphomas accounts for 10% of all thyroid lymphomas.[1,4,5] MALT lymphomas tend to have a more indolent course and are associated with better prognosis: the 5-year disease-specific survival rate is 90–96%.[1,4,5] Diagnosis is based on the presence of a combination of morphologic features and immunophenotyping, determined by flow cytometry and immunohistochemistry. The diagnostic features of MALT lymphoma include lymphoepithelial lesions, reactivation of germinal centers, and frequent plasmacytic differentiation.[6] However, the cytological diagnosis of MALT lymphoma is challenging because of an overall heterogeneous appearance and the difficultly in differentiating this type of lymphoma from HT. Furthermore, in many cases, MALT lymphoma presents in the thyroid gland in conjunction with HT, increasing the number of false-negative results due to a sampling error.[7]

FNA is being used more frequently for the diagnosis of tumors, especially in the thyroid. However, results on its effectiveness are conflicting, with a reported sensitivity ranging from 55% to 86%.[1,8,9] A major source of error is misdiagnosis as HT. Matsuzaka et al. reported that 65 out of 83 patients (78.3%) had been correctly diagnosed.[10] The authors recommend open biopsy in all cases to establish the final diagnosis. In our case, the tumor in the right lobe was clearly PTC. Malignant lymphoma was suspected in the left lobe, but a confirmatory diagnosis was not possible. We therefore concur that open biopsy would be required for further investigation and to determine the appropriate treatment.

Currently, the optimal treatment and follow-up duration for patients with thyroid lymphoma remain controversial. Usually, patients are treated with radiotherapy for localized disease and

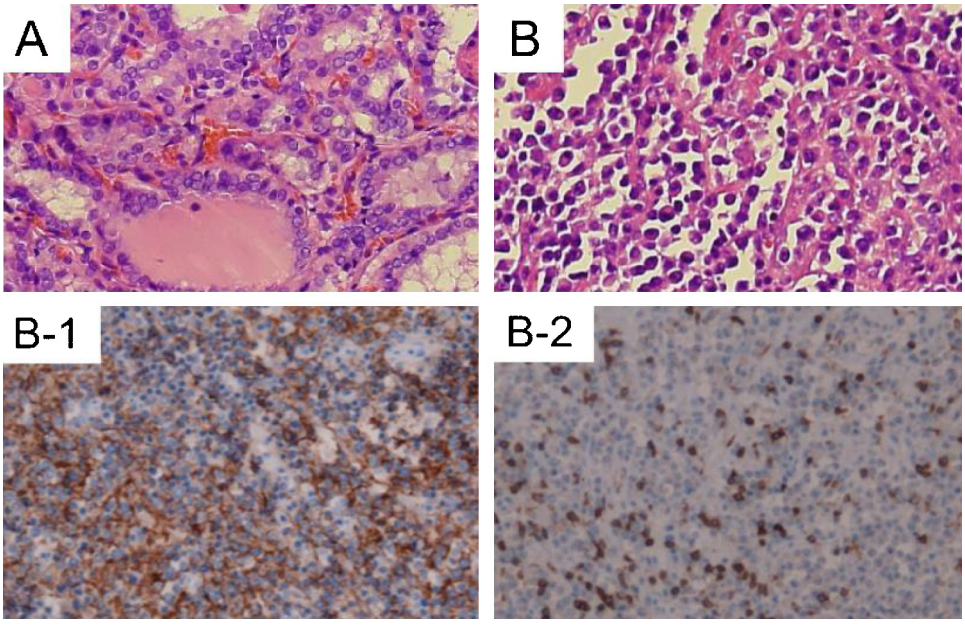

Fig. 2. Histology of the right thyroid tumor (A) showed a predominant follicular pattern, and the left thyroid tumor (B) showed accumulation of the lymphnode cells. Immunohistochemical staining showed dominant expression of CD20+ (C-1) rather than of CD3+ (C-2) suggesting MALT lymphoma of the thyroid.

chemotherapy for disseminated disease, whereas recurrence is treated with either radiotherapy or a combination of radiotherapy and chemotherapy.[6] There is also no clear differentiation in the treatment for diffuse B-cell lymphoma or mixed type and MALT in thyroid lymphomas, although their clinical courses are clearly different.[6] In a retrospective study of 108 patients with all subtypes of thyroid lymphoma, Derringer et al. reported no correlation between treatment type (surgery alone, surgery and radiation, surgery and chemotherapy, or surgery and multimodality therapy) and survival outcome.[10]

In cases of co-occurrence of PTC and MALT lymphoma, the most important consideration is whether surgery or radiation and chemotherapy should be performed followed by, if surgical treatment is selected, whether total thyroidectomy or hemithyroidectomy should be performed. In the case presented herein, the patient exhibited rapid enlargement of the thyroid gland and symptoms of dyspnea and difficulty in swallowing. For conservation of thyroid gland function, stent implantation and radiation and chemotherapy would normally be the choice of treatment, but in this case, progression of the tumor was so rapid that the time to production of the effect was crucial. To relieve compression on the trachea, hemithyroidectomy is recommended, but in our case, PTC was present in the right lobe. Therefore, total thyroidectomy was the only possible course of action in our case.

Two other groups, Cheng et al.[1] and Melo et al.[3] reported co-occurrence of PTC and MALT lymphoma, but neither described the symptom of airway obstruction. In both cases, total thyroidectomy was performed and had no recurrence for six years and two years respectively. No evidence of recurrence of either PTC or MALT lymphoma has been noted in the present case, but careful follow-up is essential because there are no reports for long-term prognosis of co-occurrence of the two malignant tumors.

4. Conclusion

We report a case of co-occurrence of PTC and MALT lymphoma in the thyroid gland, causing severe airway obstruction. We performed emergency total thyroidectomy to relieve obstruction of the trachea and to remove the two malignant tumors. We suggest total thyroidectomy for a case of co-occurrence of two malignant tumors, causing severe airway obstruction, but there is little published information in the selection of treatment and long-term survival.

Conflict of interest

None of the authors have any conflicts of interest to disclose.

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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 of this journal on request.

Author contributions

Tatsuya Tarui MD is the chief author, and other authors (Norihiko Ishikawa MD, PhD, Shinichi Kadoya MD, PhD, and Go Watanabe MD, PhD) are contributors.

Key learning points

- We suggest emergency total thyroidectomy should be performed without delay for a case of co-occurrence of two malignant tumors, causing severe airway obstruction.

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