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Radical Surgery for Primary Thyroid Lymphoma in Elderly Patients: A Short Report

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ABSTRACT

Primary thyroid lymphoma (PTL) is a rare neoplasm that requires early and accurate diagnosis, as its approach and management is different from other thyroid malignancies. Primary thyroid lymphoma (PTL) usually presents with rapidly growing neck mass with pressure symptoms. We report two elderly females with PTL who developed a painless enlarging thyroid mass. They had no medical history or family history of thyroid disease. Based on clinical symptoms and radiologic findings, both of them underwent surgical resection of the tumors. Ultrasonography and CT scan showed no specific findings, and like Hashimoto's thyroiditis, only a large diffuse thyroid with nodular tissue was found. Postoperative histologic examination revealed high-grade non-Hodgkin lymphoma. Overall, these cases emphasize the importance of considering Primary thyroid lymphoma (PTL) when dealing with large thyroid tumors. Although the treatment of choice for Primary thyroid lymphoma (PTL) is chemoradiotherapy, surgery could be performed when the diagnosis is inconclusive or has compressive symptoms.

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Introduction

Primary thyroid lymphoma (PTL) is a rare group of thyroid malignancies that affect the thyroid gland with/without involving local neck lymph nodes (1). PTL represents 5% of malignant tumors in the thyroid gland and is more common in females, with a peak incidence in the seventh decade of life (2). Besides, Hashimoto's thyroiditis is an important risk factor as it occurs simultaneously in 70% to 90% of patients with PTL (2). Most reported cases are B-cell non-Hodgkin lymphoma (NHL), the most common histotypes being mucosa-associated lymphoid tissue (MALT) lymphoma and diffuse large B-cell lymphoma (DLBCL), whereas Hodgkin lymphoma are less common (3).

Although PTL is uncommon, it should be diagnosed early because its management is different from the other thyroid neoplasms and it is curable if treated properly (3). Here, we discuss two cases of elderly females with PTL who were successfully treated with surgical resection and the diagnosis was confirmed by histology and immunohistochemistry (IHC).

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Case Reports

Case 1:

A 65-year-old female presented to the surgery department with dyspnea caused by rapidly expanding neck swelling in the last five weeks. She had no history of pain, hoarseness, dysphagia, cold or heat intolerance, or weight loss. She also had no medical history or family history of thyroid disease. Physical examination showed diffuse non-tender swelling that involved the entire thyroid gland. There was no cervical or another lymphadenopathy. Complete blood count, thyroid hormone levels, and thyroid autoantibodies were normal.

Thyroid ultrasonography showed enlargement of both thyroid lobes and a low echoic lesion. Fine needle aspiration (FNA) was performed, which reported thyroid cytology as a benign nodule and adenomatoid goiter. Figure 1 illustrates the neck computerized tomographic (CT) scan. Therefore, based on the patient's clinical and radiological information, we considered the possibility of a retrosternal goiter.

The patient underwent total thyroidectomy with isthmusectomy. Histological examination revealed high-grade NHL.

Case 2:

A 60-year-old female was hospitalized with painless neck mass from 2 months ago, without accompanying compressive mass symptoms such as dyspnea or dysphagia. Her past medical history and family history were unremarkable. On examination, the enlargement of the entire thyroid gland, mainly the right lobe, was palpable.

Ultrasound examination revealed swelling of both lobes with heterogeneous hypoechoic nodularity and diffused hyperechoic areas. FNA cytology examination was not performed due to patient disagreement. The patient's CT scan is shown in Figure 2. Based on the obtained evidence, she underwent surgery with a preoperative diagnosis of anaplastic thyroid carcinoma.

In surgery, a transverse incision superior to the sternal notch was made. It was a firm, lobular mass that extended from the hypopharynx below the hyoid bone to the anterior mediastinum behind the sternum. After releasing the thyroid from the surrounding tissue, due to arteries and tracheal involvement, it was not possible to have a total thyroidectomy without a residue. Therefore, subtotal thyroidectomy and tumor debulking were conducted. The tumor on the right side involved the strap muscles, as well as the jugular vein, which had no blood flow, which ligated.

Macroscopic pathological examination showed six irregular thyroid lesions with soft to elastic consistency, the largest of which was 7.5 × 6 × 2.5 cm and the rest were 7, 5, 4, 3 and 3 cm in length. Microscopically, sections of neoplastic proliferation of atypical lymphoid cells with extensive infiltration to adjacent tissue and necrosis were found. The final histopathologic diagnosis was high-grade NHL.

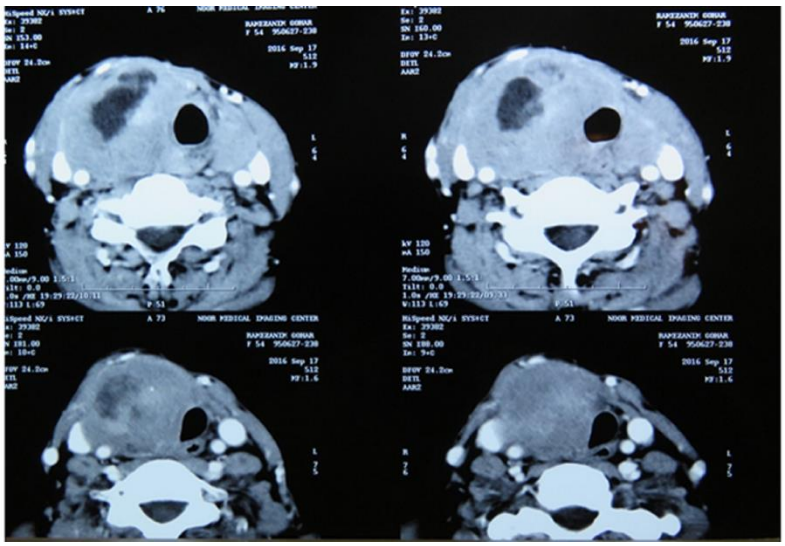

Figure 1. Illustrates the neck computerized tomographic (CT) scan.

Discussion

PTL is a rare malignancy that has been found in approximately 2% of extranodal lymphomas (2). It is often derived from B lymphocytes and the female-to-male ratio being 3:1 (2). It is crucial to differentiate PTL from secondary thyroid lymphoma. PTL is limited to the thyroid gland, usually occurs in the context of autoimmune thyroiditis and occurs in the older age group, on average 60 years. On the other hand, in secondary thyroid lymphomas, most patients have disseminated disease that the thyroid is secondarily affected by lymphoma and it often occurs in the middle-aged group, on average 42 years (4). In our cases, only the thyroid was involved and there was no evidence of disseminated disease in our evaluation. Also, the age of patients was more in line with PTL.

The exact etiology of PTL is still unknown, but studies have shown that about 40% to 85% of patients had a history of autoimmune thyroid disease (5,6). It has been hypothesized that Hashimoto's thyroiditis or chronic lymphocytic thyroiditis, activates B-cells, which secrete autoantibodies and eventually lead to the malignant proliferation of thyroid lymphoid tissues (7). In a meta-analysis of 1346 PTLs, 78.9% of patients had evidence of Hashimoto's thyroiditis (8). However, our cases had no history of Hashimoto's thyroiditis and also anti-TPO levels were negative.

As our patients, the most common clinical manifestation of PTL is a rapidly enlarging mass in the neck that can cause other symptoms such as dysphagia, dyspnea, and voice change (2). These symptoms overlap with numerous benign and malignant thyroid lesions and require a radiological examination for an accurate diagnosis.

Ultrasonography and CT scan do not show specific findings, and like Hashimoto's thyroiditis, only a large diffuse thyroid with nodular tissue is found (7). FNA cytology is another common method used to diagnose thyroid masses, but there are different results regarding its role in diagnosing PTL. Some studies have reported FNA cytology accuracy as 25% - 90% (9, 10), while some researchers considered that to be limited in preoperative diagnosis [6]. FNA cytology was performed in our patients before surgery, but none of them was definitively diagnosed with lymphoma. As a result, the preoperative diagnosis of PTL seems complicated, and postoperative histopathology and immunohistochemistry (IHC) are considered the gold standard for PTL diagnosis.

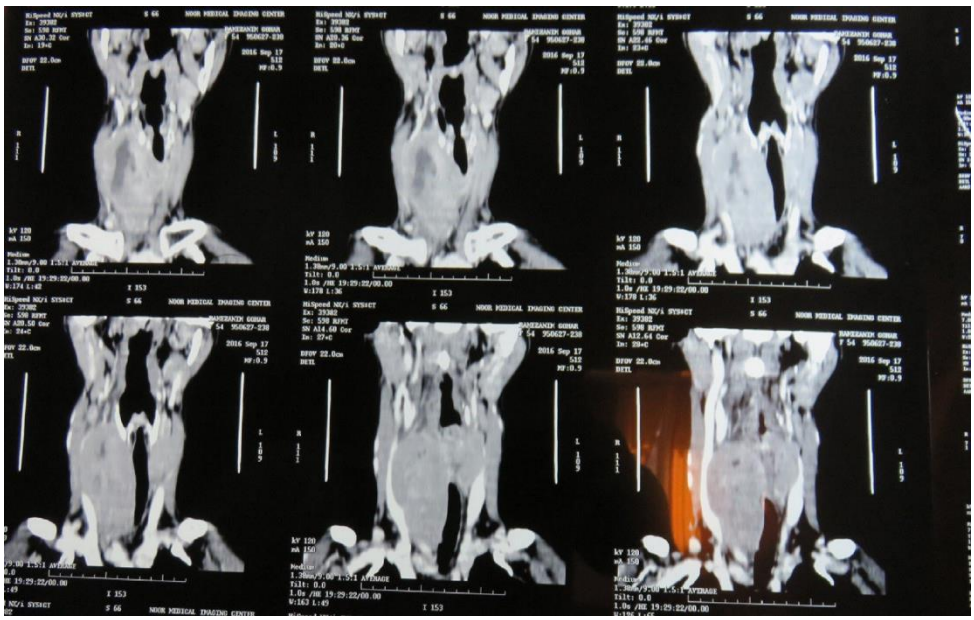

Figure 2. CT scan is shown with a preoperative diagnosis of anaplastic thyroid carcinoma.

Histologically, the majority of PTLs are B-cell NHL, whereas T-cell NHL and Hodgkin lymphoma are rare. The most common subtype is DLBCL, which accounts for about 70% of patients (4). Approximately 60% of patients in this subtype are diagnosed with disseminated disease, indicating a more aggressive clinical course (4). Another common subtype is MALT lymphoma, a relatively indolent course, and accounts for 6-27% of thyroid lymphomas (4).

There are both surgical and non-surgical methods for PTL management. In patients that the mass has caused airway and digestive obstruction, the diagnosis is uncertain, or the lymphoma is localized in the thyroid, surgery may be necessary. However, chemo radiotherapy is the gold standard treatment in PTL (11). In our patients, the preoperative findings did not specify the exact type of disease, and the mass had also caused airway obstruction symptoms, so both of our patients underwent surgery.

PTLs are rare malignancies that should be considered in patients with a large thyroid mass, especially in females with Hashimoto's thyroiditis background. Although the diagnostic approach to PTL is more complicated than other thyroid malignancies, early diagnosis and appropriate treatment are essential because it affects the prognosis. FNA cytology is one of the first steps in the thyroid mass evaluation but may not always be diagnostic, while histopathological evaluation of the mass completed by the IHC is the gold standard of diagnosis. Chemoradiotherapy is the treatment of choice, but surgery is necessary in patients such as our cases who have airway obstruction or to ascertain the diagnosis.

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