Carcinoma Showing Thymus-like Differentiation (CASTLE) with Non-Recurrent Laryngeal Nerve: A Case Report

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Carcinoma showing thymus-like element (CASTLE) is a very rare malignant neoplasm in the lower portion of the thyroid gland or soft tissue of the neck. Recurrent laryngeal nerve (RLN) is the most frequent site of CASTLE. Non-RLN is also a rare anomaly. Both CASTLE and non-RLN are risk factors for vocal cord paralysis. In this report, the authors describe a 73-year-old patient diagnosed with CASTLE and non-RLN. During total thyroidectomy, one RLN was sacrificed inevitably because of tumor invasion, while the other non-RLN was successfully saved, which was expected based on preoperative computed tomography (CT). If the diagnosis is uncertain, CT should be checked to prevent unexpected risks.

Key Words: Carcinoma, Carcinoma showing thymus-like element (CASTLE), Thyroid, Thymus, Laryngeal nerve

Introduction

Carcinoma showing thymus-like element (CASTLE) is a very rare malignant neoplasm. About 50 cases have been reported in the English-language literature since it was first described by Miyauchi et al. 1) as an intrathyroidal epithelial thymoma in 1985. This tumor characteristically slowly progresses from the lower portion of the thyroid gland or soft tissue of neck. 2) This tumor is recognized as independent clinicopathologic entity and is included in the World Health Organization’s classification of endocrine tumors. 3) The diagnosis of CASTLE is difficult not only because of its rarity, but also its histologic appearance is similar to thymic carcinoma of the lymphoepithelioma or squamous cell variety. 3)

The non-recurrent laryngeal nerve (non-RLN) originates from the anomalous subclavian artery, which occurs in less than 1% of patients. 4) Non-RLN has a short track arising from the vagus nerve directly, while normal RLN has a long track circumscribing the subclavian artery or aortic arch on the right or left side. The aberrant path of the non-RLN makes the risk of nerve injury during thyroidectomy higher than normal. This is the first reported case having these two rarities occurring in the same patient. The authors discuss the high points of evaluation and management as well as review the literature.

Case Report

A 73-year-old male visited the Otorhinolaryngology Department at Hallym University Sacred Heart Hospital...
in April 2007 for gradually worsening hoarseness, which appeared 1 year prior. He did not report any dyspnea, dysphagia and odynophagia. Physical examination revealed a 2 cm, solid, fixed neck mass at the left lower portion of the thyroid gland without tenderness, while no neck node was palpated. The left vocal cord was completely paralyzed, as was detected on laryngoscopic examination. He did not have any significant past medical history or family medical history, including cancer. Thyroid function and calcitonin tests were within normal limits. Ultrasonography showed a 2.2×1.9×2.8 cm hypoechoic solid mass without any significant neck nodes (Fig. 1). Fine needle aspiration biopsy (FNAB) was performed and follicular carcinoma was suspected. However, pathologic diagnosis with FNAB was uncertain. The computed tomography (CT) revealed a 2.2 cm low-density mass in the lower portion of the left thyroid gland (Fig. 2A, B). The aberrant right subclavian artery, which ran against the vertebral column behind the esophagus, was also detected on CT, which suggested non-RLN (Fig. 2C).

Total thyroidectomy and central neck dissection were attempted. Because the left RNL was encased within the tumor, it was resected and re-anastomosed. Frozen biopsy was performed and carcinoma was suspected. However, the type of carcinoma could not be determined. The non-RLN, which was suspected based on preoperative CT was found during right thyroidectomy, which was successfully saved.

Histologic examination of the biopsy showed variably-sized and irregularly-shaped lobules, which were divided with fibrous septa. The septa were infiltrated by lymphocytes and plasma cells (H&E stain, ×40) (Fig. 3A). Tumor cells had large vesicular nuclei and prominent nucleoli (H&E stain, ×400) (Fig. 3B). Tumor cells were negative for thyroid transcription factor-1 (TTF-1) and positive for CD5 and CD117 on immunohistochemistry (Fig. 3C–E).

This tumor was diagnosed as CASTLE with these immunohistochemistry findings. The patient underwent radiation therapy of 6120 cGy over 7 weeks because of tumor invasion into perithyroid soft tissue, while no central lymph nodes were involved. He did not show any evidence of recurrence after 72 months of follow-up. The voice of patients was not improved as it has been completely paralyzed before the surgery.
He did not complain any dyspnea or aspiration because we saved non-RLN.

Discussion

Tumor invasion into extended sites are detected in 60% of CASTLE and the RLN is the most frequent site (50%), followed by the trachea (38%). Not only tumor invasion of the RLN itself, but also resection of the extended site to achieve oncologically sound surgery could lead to vocal cord paralysis. This suggests that CASTLE has a high risk of vocal cord paralysis. Non-RLN is another risk factor for vocal cord paralysis during thyroidectomy. The risk of non-RLN injury is
CASTLE with Non–Recurrent Laryngeal Nerve

10–times higher than normal RLN if it is not detected preoperatively.6) We checked the CT preoperatively in this case and found the aberrant retroesophageal subclavian artery, which indicated the possibility of non–RLN. As a result, we could save the non–RLN, which is the only functional RLN in this patient.

Ultimobranchial body remnants such as thyroid solid cell nests and/or from ectopic thymic tissue are regarded as the origin of CASTLE.7) The non–RLN always accompanies the congenital anomaly of the subclavian artery. We could not find any study that explains the embryologic comparability of CASTLE and the non–RLN. CASTLE and the non–RNL are thought to develop independently.

CASTLE typically occurs in adults in the fifth decade of life with a slight female predominance (M: F=1:1.3).5,8) Surgery is an accepted principle for this tumor and radiation therapy seems to be important to prevent locoregional recurrence, while the role of chemotherapy is still controversial.9) CASTLE shows a favorable prognosis,9) However, older patients with CASTLE usually have worse prognosis than younger patients.10) Despite his older age, this patient has done well so far. However, CASTLE may recur after a long disease–free interval, even decades after initial therapy.9) Therefore, long–term follow–up is necessary for this patient.

CASTLE usually presents as a lobulated hypodensity mass with marginal enhancement on CT and a hypoechoic solid mass without a calcification on ultrasonography. However, it is very difficult to suspect CASTLE with these findings and it is also very hard to diagnose CASTLE with FNAB because of its rarity. Only one case was diagnosed correctly by FNAB.3) Thus, clinicopathologic correlation and immunohistostaining is the most important for the diagnosis of CASTLE. Sensitivity and specificity of CD5 in CASTLE were 82% and 100%, respectively.5) Negativity in TTF–1 and positivity in CD5 and CD117 immunostaining are characteristics of CASTLE,10) which led us to diagnose this case as CASTLE.

In conclusion, both CASTLE and non–RLN are risk factors for vocal cord paralysis. Even if one RLN was sacrificed inevitably because of tumor invasion, the other non–RLN could be successfully saved by the aid of preoperative CT. If confirmatory diagnosis is not made, CT should be checked to prevent unpredicted risks.

References