A Case of Mediastinal Ectopic Thyroid Cancer With a Normal Thyroid Gland

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ABSTRACT

Ectopic thyroid cancer is extremely rare disease, only a few case reports are published about it. Ectopic thyroid cancer usually arises from ectopic thyroid tissue. The predominant location of ectopic thyroid tissue is the lingual thyroid, whereas a mediastinal thyroid is uncommon. Ectopic thyroid cancer below the neck is extremely rare; therefore, the diagnosis of ectopic thyroid cancer below the neck is based on pathologic results. We describe here a case of asymptomatic mediastinal ectopic thyroid cancer with a normal thyroid gland, underwent mediastinal mass excision and no relapse for 3 years.

Keywords: Thyroid dysgenesis; Thyroid cancer; Mediastinum

INTRODUCTION

The ectopic thyroid gland can reside anywhere along its embryological path. Ectopic thyroid tissue is rare, with a reported incidence of 1 in 300,000 (1-3). It can cause hypothyroidism and a palpable mass; however, it can also be asymptomatic (1).

Ectopic thyroid cancer is an extremely rare condition that arises from ectopic thyroid tissue. The predominant location of ectopic thyroid tissue is the lingual thyroid, whereas a mediastinal thyroid is uncommon (4). Ectopic thyroid cancer below the neck is extremely rare; therefore, the diagnosis of ectopic thyroid cancer below the neck is based on pathologic results.

Here, we described a rare case of mediastinal asymptomatic ectopic thyroid cancer with a normal thyroid gland. Written informed consent was obtained from the patient for the publication of this case report.

CASE REPORT

A 30-year-old woman presented with abnormal chest radiography findings during a health checkup. The patient had no other symptoms or signs on physical examination. Her medical and family histories were unremarkable.
Chest radiography revealed a mass-like lesion in the right paratracheal region. Computed tomography (CT) scan showed an intensely enhancing mass of approximately 4.0×4.5×6.9 cm in size between the right brachiocephalic vein and innominate artery with a centrally hypo-enhancing portion in the right superior mediastinum. There were no remarkable lung or lymph node lesions (Fig. 1).

Positron emission tomography (PET)-CT showed a well-defined mass measuring approximately 4.7 cm in size with F-fluorodeoxyglucose (FDG) uptake in the right paratracheal area, with no other hypermetabolic foci to suggest distant metastasis. Magnetic resonance imaging (MRI) showed a T2 high and T1 intensely high-intensity lesion with hyper-enhancement in the right middle mediastinum, displacing the adjacent structure without invasion into the bronchus.

Cytology via endobronchial ultrasound-guided transbronchial needle aspiration (EBUS-TBNA) revealed a suspicious neuroendocrine tumor.

Surgery was performed under the suspicion of a neuroendocrine tumor of unknown origin. The mediastinal mass was excised using 3-port video-assisted thoracic surgery. Using one camera port and 2 working ports on the right thorax, the large cystic mass that was positioned between the right brachiocephalic vein and azygos vein was successfully excised without postoperative complications.

The final pathologic report revealed a 6.5 cm sized solid/trabecular subtype papillary thyroid carcinoma, and lympho-vascular invasion free carcinoma in the resection margin was not identified. Necrosis and mitosis were not observed. The tumor cells showed nuclear features of typical papillary thyroid carcinoma with strong and diffuse immunoreactivity for TTF-1, PAX8, and thyroglobulin (Fig. 2).

After the diagnosis of thyroid cancer, the patient was referred to the thyroid cancer center, and her thyroid gland was evaluated.

Thyroid function tests (TFTs) results were normal; T3, 77.8 ng/dL (71–161 ng/dL); free T4, 1.0 ng/dL (0.8–1.7 ng/dL); thyroid stimulating hormone, 4.01 µIU/mL (0.86–4.69 µIU/mL).
Thyroid ultrasonography (US) revealed no suspicious lesions or lymph nodes in the thyroid, and the neck field was visualized (Fig. 3).

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**Fig. 2.** Histopathologic features of the mediastinal mass. (A) The mass is encapsulated solid ovoid mass. Cut surface shows tumor capsule (arrow) and gray-tan colored mass. (B) The tumor shows mainly solid/trabecular and focal follicular patterns (hematoxylin-eosin stain, ×100). (C) The tumor cells show nuclear features of typical papillary thyroid carcinoma, including nuclear clearing, irregular nuclear membrane, and nuclear groove (arrow) (hematoxylin-eosin stain, ×400). (D) The tumor cells show strong and diffuse immunoreactivity for thyroglobulin (×200).

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**Fig. 3.** Thyroid ultrasonography findings show no abnormal suspicious lesion on both thyroid lobes.
At that time, this case was discussed at the tumor board meeting, and it was decided that the patient would be followed-up with careful examination because there were no suspicious lesions in the thyroid gland.

The patient was carefully followed up with TFTs, neck US and chest CT every year, and the patient continued to do well without any recurrence of the disease after 3 years.

DISCUSSION

An ectopic thyroid gland is caused by developmental abnormalities at an early stage of embryogenesis. In the early stages, the median thyroid anlage migrates from the floor of the primitive foregut to its final tracheal position. During this process, remnants or dislocation of the thyroid tissues can cause ectopic thyroid. In most cases, ectopic thyroid tissues are found along the path of thyroid descent, corresponding to the course of the thyroglossal duct, in the midline, or laterally in the neck. In a few cases, however, it is found in the mediastinum or in the subdiaphragmatic organs: ovaries (struma ovarii), adrenals, gallbladder, pancreas, duodenum, and mesentery (5-7).

Primary ectopic thyroid cancer is a rare condition, and it has been reported to arise from the thyroid tissue in the thyroglossal cysts, lateral aberrant thyroid tissue, lingual thyroid, mediastinal, and struma ovarii (8). True malignant transformation of ectopic thyroid tissue is extremely rare. Almost all malignant transformations are diagnosed after surgical excision. In this case, we report a malignant ectopic thyroid cancer in the mediastinum with a normal orthotopic thyroid. The first diagnosis of the mediastinal mass was a neuroendocrine tumor with atypical cells by EBUS-TBNA. In another case of mediastinal ectopic thyroid cancer (9), only scanty follicles containing colloid material were detected by fine-needle aspiration biopsy for preoperative evaluation. Due to the limitations of fine needle aspiration biopsy, ectopic thyroid mass should be suspected as a malignancy, and mass excision should be considered.

Most malignancies in ectopic locations include papillary carcinomas, mixed follicular and papillary carcinomas or Hurthle cell tumors, and anaplastic carcinomas (8,10). They can show aggressive behavior and give rise to distant metastases; therefore, it is often challenging to differentiate these lesions from a metastatic spread from a carcinoma developed in an orthotopic thyroid gland (9). In this case, there was no evidence of malignancy in the orthotopic thyroid or lateral neck lymph nodes. The last pathological report of a mediastinal mass was a solid subtype papillary thyroid carcinoma. The solid subtype of papillary thyroid carcinoma is associated with a slightly higher frequency of distant metastases and less favorable prognosis than conventional papillary carcinoma (11). After surgery, there were no signs of relapse in the orthotopic thyroid on neck US conducted regularly every year for 3 years without any medical therapy.

Ectopic thyroid tissue can cause hypothyroidism, a palpable mass, and dyspnea, depending on the location of the tissue; however, it can also be asymptomatic (1). Usually, mediastinal tumors give rise to compressive symptoms; however, this case showed a normal thyroid function with no other symptoms. With compressive symptoms, patients with mediastinal tumors are suggested to undergo surgical excision with thoracotomy, sternotomy, or thoracoscopic excision.
In this case, there were no suspicious findings in the thyroid gland on US, and there was no relapse of thyroid cancer without medical therapy. Several case reports have reported similar findings (9,12,13). Further studies are needed to evaluate the association between ectopic thyroid cancer and malignant changes in the thyroid glands. Furthermore, research on the necessity of additional medical therapies after the excision of ectopic thyroid cancer is needed.

In conclusion, although ectopic thyroid tissue can be found anywhere along its embryological path, thyroid cancer arising from ectopic thyroid tissue is extremely rare. Diagnosis of ectopic thyroid cancer may not be easy since it can be diagnosed after surgery only; hence, surgeons should be actively aware and consider such possibility.

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