CASE REPORT

Korean J Intern Med 2013;28:361-364
http://dx.doi.org/10.3904/kjim.2013.28.3.361

KJIM

A case of mediastinal ectopic thyroid presenting with a paratracheal mass

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Received: July 11, 2011
Revised : September 6, 2011
Accepted: February 14, 2012

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Mediastinal ectopic thyroid is a very rare condition, with few reported cases in the literature and no reported cases in Korea. This report describes an asymptomatic 65-year-old man with a right paratracheal mass compressing the superior vena. Additionally, the epidemiology, clinical manifestation, diagnosis, and management of mediastinal ectopic thyroids are discussed. A mediastinal ectopic thyroid should be considered in the differential diagnosis of all mediastinal masses. Surgical excision is recommended for both the diagnosis and treatment of this condition, because of its potential for malignancy and compression of mediastinal structures. This case demonstrates the clinical importance of mediastinal etopic thyroid.

Keywords: Thyroid dysgenesis; Mediastinum

INTRODUCTION

An ectopic thyroid gland is defined as thyroid tissue that is not located anterolaterally to the second to fourth tracheal cartilages. Anatomically, an ectopic thyroid can be lingual (at the base of the tongue), sublingual (below the tongue), prelaryngeal (in front of the larynx), or can be found at other rare sites. Mediastinal ectopic thyroids are very rare, accounting for less than 1% of all cases [1], but rare mediastinal ectopic thyroid is also important to consider in the differential diagnosis of mediastinal masses. Here, we review a rare case of mediastinal ectopic thyroid presenting with a paratracheal mass compressing the superior vena cava but without symptoms, a condition that has not been reported previously in Korea.

CASE REPORT

A 65-year-old male presented with a right paratracheal mass, incidentally detected on a computed tomography (CT) scan performed during a health examination after a traffic accident (Fig. 1). No evidence or

pISSN 1226-3303
eISSN 2005-6648
http://www.kjim.org

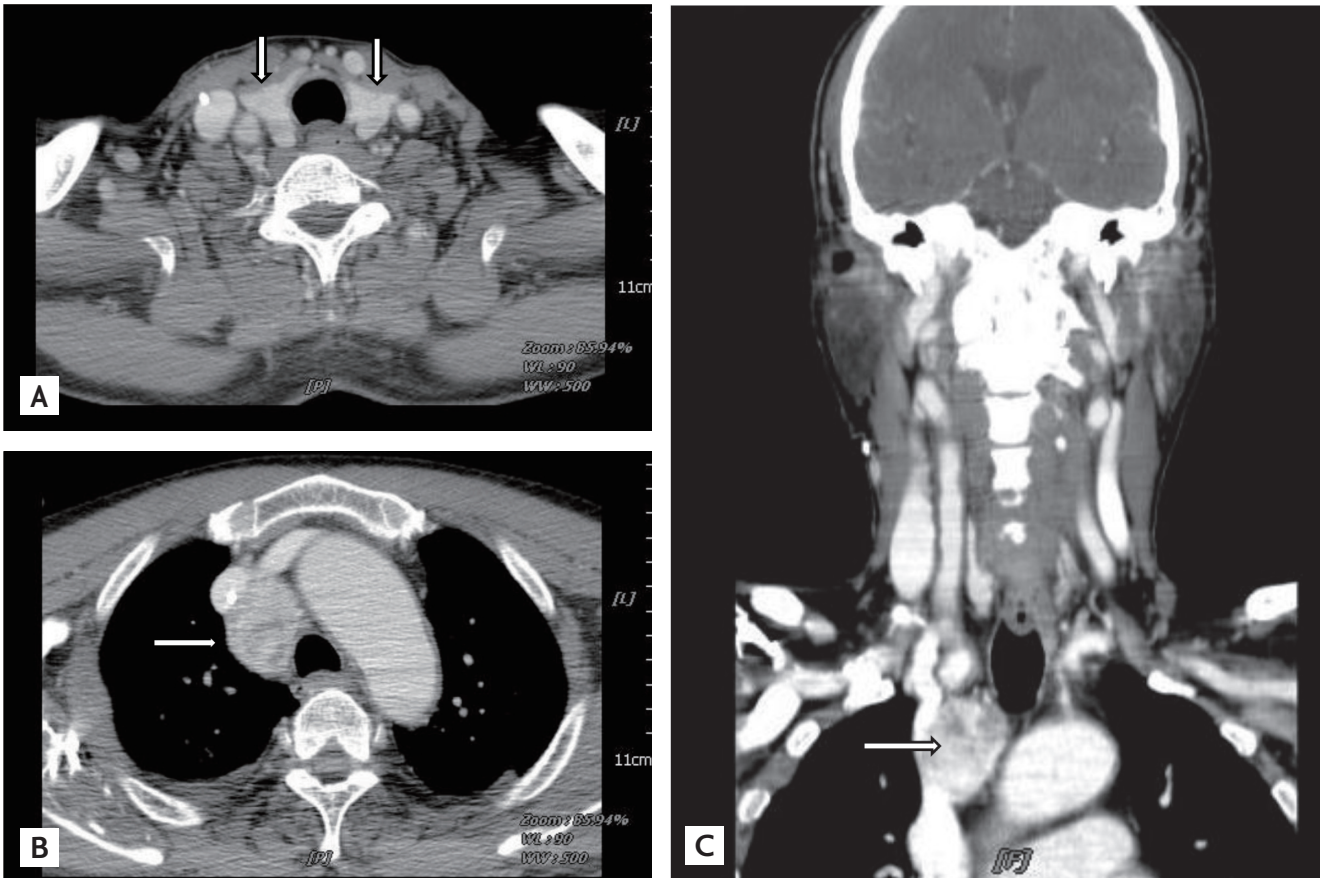

Figure 1. (A) Normally located thyroid gland (arrows). (B) Heterogenously enhanced supramediastinal mass (arrow). (C) Coronal imaging showing the right paratracheal mass compressing the superior vena cava (arrow).

symptoms of thyroid dysfunction were found. The patient had had hypertension for several years. He had no history of exposure to radiation, and there was no family history of thyroid disease. His thyroid was not palpable, and there was no evidence of cervical lymphadenopathy. The results of his laboratory tests were all within normal limits and included a thyroid stimulating hormone level of 0.96 mU/L (normal range, 0.40 to 4.00), T3 of 1,090 pmol/L (normal range, 600 to 1,950), and free T4 of 11.2 pmol/L (normal range, 7.8 to 19.4). The titers of serum thyroid autoantibodies were also within normal ranges; antithyroid peroxidase antibody was < 25 U/mL (normal range, < 100) and antithyroglobulin antibody was < 25 U/mL (normal range, < 100).

A CT scan of the neck (Fig. 1) revealed a 4.5 × 2.9 cm heterogeneously enhanced mass in the right paratracheal area. The mass was located at the intersection of the caudal margin of the left brachiocephalic vein and the trachea, and it compressed the superior vena cava. However, there were no symptoms related to the compression. The thyroid gland was located in the normal position and showed slightly heterogeneous enhancement and no cervical adenopathy. There were no focal nodules in either lobe.

Endobronchial ultrasound (EBUS) (Fig. 2) and EBUS-transbronchial needle aspiration were performed to obtain a tissue sample of the paratracheal mass. During EBUS, the large mass was observed as a right upper paratracheal lesion (2R lesion). The radiologic diagnosis of the mass by EBUS was metastatic adenopathy, Castleman's disease, or tuberculous lymphadenitis. Biopsies of the mass showed normal thyroid follicles (Fig. 3).

Excision of the ectopic thyroid tissues was recommended to the patient. Recently, he had undergone a

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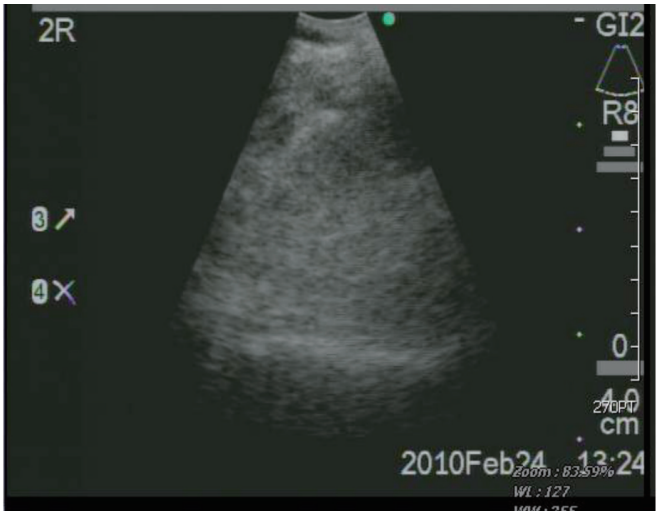

Figure 2. Endobronchial ultrasound of the right paratracheal mass (2R lesion).

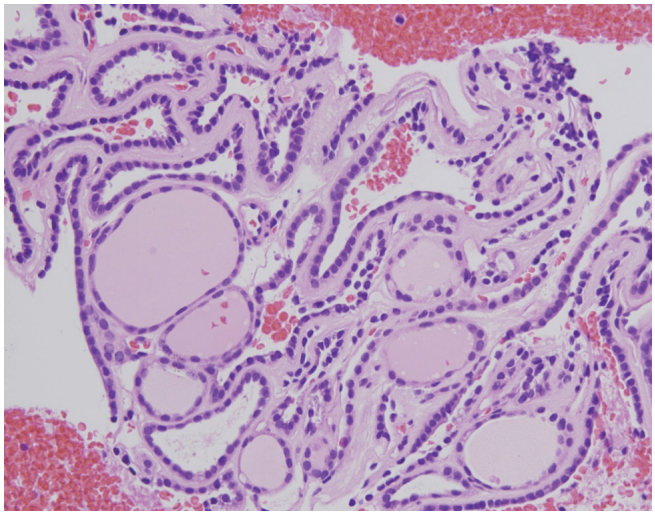

Figure 3. Photomicrograph of the ectopic thyroid tissue showing some fragmented, benign-looking thyroid follicles (H&E, × 40).

coil embolization for an aneurysm in his left posterior communicating artery; he is now waiting for the ectopic thyroid tissue to be surgically excised after his general condition has stabilized.

DISCUSSION

Ectopic thyroid tissue is the result of abnormal gland migration from the foramen caecum to its normal pretracheal position. Lingual thyroid tissue accounts for 90% of these abnormalities, and sublingual ectopic tissues are much less frequent. Intratracheal ectopic thyroid tissues have also been reported. Dual ectopic thyroid has been described, with thyroid gland tissue also present in the normal location. The wall of a thyroglossal duct cyst is a common site for ectopic thyroid tissue, and the presence of a solid mass along a thyroglossal duct cyst should raise a suspicion of ectopic thyroid tissue [2]. Furthermore, other rare sites, such as the mediastinum, gall bladder, porta hepatis, and duodenum have also been described. Biallelic mutations in FOXE1 have been shown to result in thyroid ectopy in mice; however, to date, no mutations in known genes have been associated with human ectopic thyroid tissues [3].

Ectopic thyroid tissue in the thorax with no connection to the cervical thyroid gland is very rare [1]. We could find only a small number of cases of mediastinal ectopic thyroid tissues in the literature, and there were no reported cases in Korea. Most Korean cases reported have been lingual or sublingual (infrahyoid) ectopic thyroids, although several cases of dual ectopic thyroids, one lateral ectopic thyroid, and two cases of intratracheal thyroids have been reported [4-6]. To our knowledge, this is the first case of mediastinal ectopic thyroid in Korea.

The mediastinum is a unique anatomic area containing several structures and pluripotent cells that allow for the development of a range of tumors. Mediastinal tumors include primary thymic carcinomas, neuroendocrine carcinomas, germ-cell tumors, and lymphomas, as well as neurogenic, endocrine, and mesenchymal tumors. Endocrine tumors include ectopic thyroids, intramediastinal goiters, and parathyroid tumors. Intramediastinal goiters are not uncommon and usually represent direct extensions of large eutopic glands. More rarely, primary thyroid tumors (adenomas or carcinomas) may occur in the mediastinum without cervical disease.

Ectopic thyroid tissue should be considered in the diagnosis of mediastinal masses. The neck should be examined for a normally located thyroid gland. CT or magnetic resonance imaging is mandatory for evaluating the site and size of the lesion. Thyroid scintigraphy with ^{131}I or technetium-99m is highly sensitive and specific for detecting normal and ectopic thyroid tissues. EBUS has been shown to be of increasing

importance in the diagnosis of mediastinal masses. Histological findings are the most important for accurate diagnosis. Most mediastinal ectopic thyroid cases showed normal thyroid follicles, and one case revealed a papillary thyroid cancer [7]. Based on the location, CT-guided fine needle aspiration, EBUS-transbronchial needle aspiration or surgical excision is chosen to obtain tissues.

Because of their invasiveness, mediastinal thyroid carcinomas usually present with dyspnea, wheezing, and chest pain, whereas benign lesions are usually discovered incidentally. Mediastinal goiter can remain asymptomatic until the structures located in the thoracic inlet are compressed. The most common symptoms are dyspnea, dysphagia, cough, and hoarseness, and, occasionally, some patients present with superior vena cava syndrome. The chief complaints in reported mediastinal ectopic thyroid cases are painful or pulsating retrosternal mass, dyspnea, and cough. Our case was found incidentally and was asymptomatic, although the mass compressed the superior vena cava. If this mass had remained undiscovered, the patient might have suffered superior vena cava syndrome.

In mediastinal ectopic thyroid cases, both euthyroidism and hypothyroidism are found, regardless of the presence of a normal thyroid gland. In previous Korean studies, hypothyroidism has been found in up to two-thirds of patients with ectopic thyroid [4], and these patients did not have a normal thyroid gland. However, in one case of hypothyroidism there was a normally located thyroid [8]; thus, a thyroid function test is necessary, irrespective of the existence of a normal thyroid. In the present case, the thyroid gland was located in the normal position, and the thyroid function test showed euthyroidism. The titers of serum thyroid autoantibodies were also within normal ranges.

Surgery for mediastinal goiters should always be considered, even in elderly patients, because of the high risk of tracheal compression and the low morbidity of the surgery [9]. Although there is no real consensus regarding the proper management of mediastinal ectopic thyroids, surgical excision must be considered because they can be malignant, and can have mass effects on the surrounding structures.

In summary, ectopic thyroid is a rare condition, and its location in the mediastinum is even rarer. Although entirely intrathoracic ectopic thyroids are rare, they must be considered in the differential diagnosis of all mediastinal masses. Because they have the potential to become malignant and to compress mediastinal structures, surgical excision of mediastinal ectopic thyroids is recommended for both diagnosis and treatment.

Conflict of interest

No potential conflict of interest relevant to this article is reported.

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