

Case Study

Multiple Thyroid Ectopia With A Normally Located Thyroid: Can It Be A Hereditary Disorder?

ABSTRACT

Heterotopic thyroid tissue - also called as ectopic thyroid - is a rare developmental anomaly. Above all, multiple ectopic thyroid glands including an intrapulmonary one are extremely rare.

A forty seven years old female patient has been admitted to our clinic with a 12 mm pulmonary nodule detected in the lower lob anterobasal segment of her right lung while she was under preoperative investigation for recurrent multinodular goiter (MNG). We have learned that she had undergone thyroidectomy 31 years ago and MNG has occurred in the residual gland. Her sister had a history of operations both for orthotopic MNG and multiple ectopic thyroid; and also her mother had MNG but has not been investigated in terms of an ectopic gland. In the operative exploration, we have found the mentioned mass at the inferior part of middle lobe and performed a wedge resection. Pathologic examination of the nodule revealed a 1 cm gray-yellow colored well circumscribed thyroid tissue with the characteristics of colloid goiter. It was positive for Thyroid transcription factor -1 and Thyroglobulin in immunohistochemical staining. Interestingly, a sublingual ectopic thyroid tissue was detected by Tc99 scintigraphy at the postoperative period.

Such a case sharing a similar medical history with her sibling in terms of thyroid pathology brings to mind that the condition might have a hereditary aspect.

Key words; Ectopic thyroid, Multiple, intraparenchymal, lung, hereditary

Introduction

Heterotopic thyroid gland - also known as ectopic thyroid- is a developmental anomaly which can be described as the existence of thyroidal tissue beside its expected place. The condition is caused by the abnormalities in migration of thyroid cells during the intrauterine development phases

As it is known, the thyroid tissue migrates from endodermal protrusion of the first and second pharyngeal arch at 3rd weeks to 7th weeks of gestation. It descends anteriorly along the midline from foramen caecum. A thyroglossal duct forms along the descending path of thyroid and undergoes atrophy during the embryonic stages. An abnormal migration of the tissue causes the gland to be misplaced, leading to ectopia

Case

A forty seven years old female patient has been admitted to our clinic for the investigation of a lung mass. She had a history of subtotal thyroidectomy 31 years ago for MNG; and years after, she had experienced recurrent MNG within the residual tissue (Figure 1). During the preoperative examinations for second thyroidectomy, chest X- ray and thorax CT were carried out and a 12 mm solitary nodul had been detected in lower lob antero basal segment of her right lung in CT images (Figure 2). In her family; her sister had a history of operations for ectopic thyroid glands which were detected in the neck and intratracheal region. Also her mother had goiter but has not been examined in terms of any ectopia. The preoperative thyroid function tests were as: T3;2,60 pg/mL(1,71-3,71),T4; 1,13 ng/dL(0,7-1,48), TSH;1,4054 uIU/mL(0,35-4,94); under levothyroxine therapy. Her respiratory function tests were as FEV1: 2,63 %85,9, Fvc: 3,34 Lt %93,9, FEV1/FVC: 78.80.

A diagnostic explorative surgery was performed by muscle sparing thoracotomy. The mass had been found out at the inferior part of middle lobe and wedge resection was carried out. Pathologic examination of the nodule revealed a 1 cm gray-yellow colored well circumscribed thyroid tissue with the characteristics of colloid goiter. It was positive for Thyroid transcription factor -1 and thyroglobulin, in immunohistochemical staining. We did not face any postoperative complications and the drain was removed at the second day. At the postoperative period, while being searched for a possibility of any other ectopic focus; a sublingual ectopic thyroid tissue was detected by whole body Tc99 scintigraphy scanning (Figure 3). She has been in the follow- up for 12 months and yet has no problem in terms of thyroid pathologies.

Discussion

In the literature; multiple ectopic thyroid glands are not commonly reported. In English series, 45 cases with dual ectopic glands had been notified. Another study based on the thyroid scintigraphy results announced the incidence of dual ectopy as 0.05 %. The most common ones can be pronounced as lingual (33.73%), sublingual (27.71%) and subhyoid (22.89%) (1). Furthermore, dual ectopic thyroid tissue with a normally located thyroid gland have been reported in only two cases (2,3) and none of these were in the lung.

Our case is consonant to dual ectopic thyroid tissue with a normally located thyroid gland, as we have detected a sublingual tissue in addition to the pulmonary one.

Three or more ectopic foci are quite rare. By the reason of the fact that we do not have enough information about the prior subtotal thyroidectomy operations, it is impossible for us to classify our case in such a group; but also, the available data let us declare that it is a multiple one.

Ectopic thyroid gland is generally observed throughout the radiological monitoring and CT scans. Radiologic examinations for the diagnosis include CT, magnetic resonance imaging, Tc-99m, I-131, and I-123. It has been reported that ectopic tissue might be seen in the mediastinum, lungs, and heart; manifesting usually with dry cough, dyspnea, and hemoptysis. Less commonly, patients may present with dysphagia or the superior vena cava syndrome. Intrathoracic thyroid may also be revealed incidentally by a chest radiography (4–6) or during an autopsy (7). Ectopic intrapulmonary thyroid can mimic pulmonary metastasis, like in our case (8). Malignant transformation is relatively uncommon; it is reported in about 15% of all patients (9).

As it is usually asymptomatic; an ectopic tissue can also cause symptoms due to its location, size, and the presence of malignancy. Indeed, our case had no symptoms; ectopic gland was detected incidentally by computerised tomography (CT) during the preoperative examinations.

In addition, it is reported that orthotopic thyroid coexisted in all cases of intrapulmonary thyroid [6,10]. Interestingly; in our case, another ectopic gland has been determined at tongue basement by whole body technetium scintigraphy at eighth postoperative month.

On the other hand, her sister had a history operations both for orthotopic MNG and ectopic thyroid. Moreover, we have learned that she had undergone servical trachea resection for intratracheal ectopic thyroid. The family history of more than three ectopic foci made us think that the condition might have a familial aspect.

Also, a recently identified familial occurrence of congenital hypothyroidism (CH) associated with thyroid dysgenesis suggests the involvement of hereditary factors (11).

Evaluation of asymptomatic first- degree relatives of children with CH has revealed a high frequency of different thyroid developmental abnormalities (11). This suggests that the thyroid phenotype may be variable despite a common genetic alteration.

Also by the genetic researches on these cases; it has been detected that 19 genes involved in embryonic development and in the Wnt pathway are exclusively associated with thyroid ectopy. Our case might have such a genetic disorder (12).

There is no consensus on the optimal therapeutic strategy for ectopic thyroid tissue because of the rarity of this clinical entity. Surgical treatment is generally planned according to the patient's age, size of the tissue, presence of local symptoms, functional thyroid status and complications; and can be performed considering the presence of any symptoms or malignancy. Depending on the position of the tissue; sternotomy or thoracotomy can be carried out. Complete surgical resection may be preferred if there is a suspicion of malignant transformation.

The follow-up in terms of mass enlargement or complications is suggested as the first option in management of asymptomatic cases with euthyroid status (13).

Also, thoracoscopic treatment might be considered as a surgical option (14). Like in our case, surgery can be performed in order to determine the presence of any malignancy; and/or to diagnose and treat the ectopic thyroid.

Considering the existence of multiple ectopic thyroid gland in both siblings; we may suggest that a genetic predisposition might have a role in the development of this condition. It can be beneficial to scan the whole body in all the cases and scrutinize their relatives.

References

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Figure 1; A; Pulmonary nodule seen on thorax computed tomography B; Giant multinodular goiter caused tracheal deviation on neck computed tomography

Figure 2; A; Lesion consisting of well-circumscribed thyroid follicles in the pulmonary parenchyma (HE, x100) B; Thyroglobulin positivity of the lesion (Thyroglobulin, X100), C; TTF-1 positivity of the lesion (TTF-1, x100)

Figure 3; A sublingual ectopic thyroid tissue detected by Tc99 scintigraphy

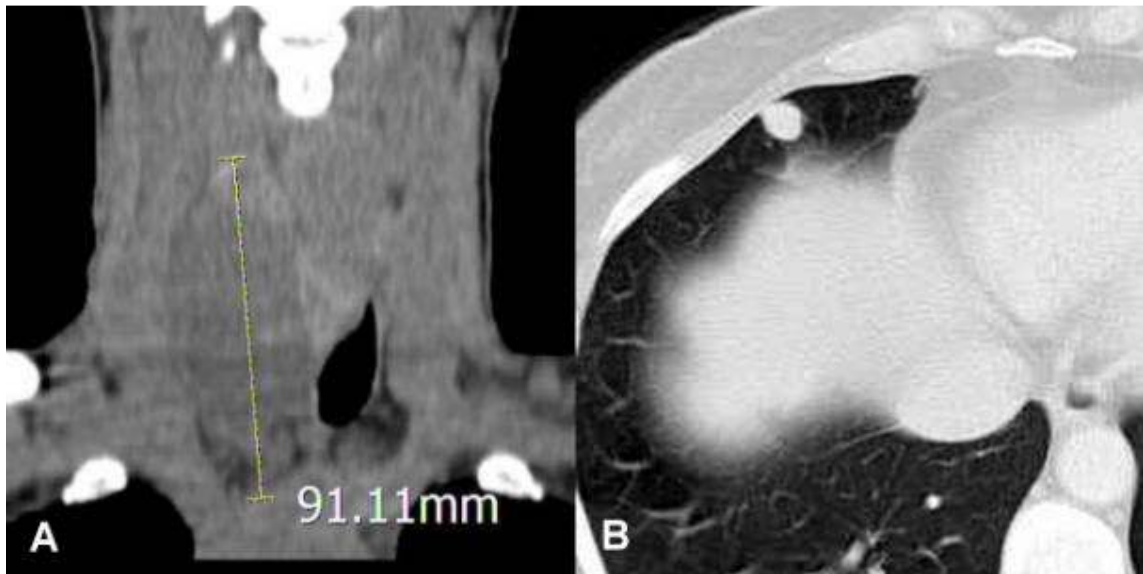


Figure 1

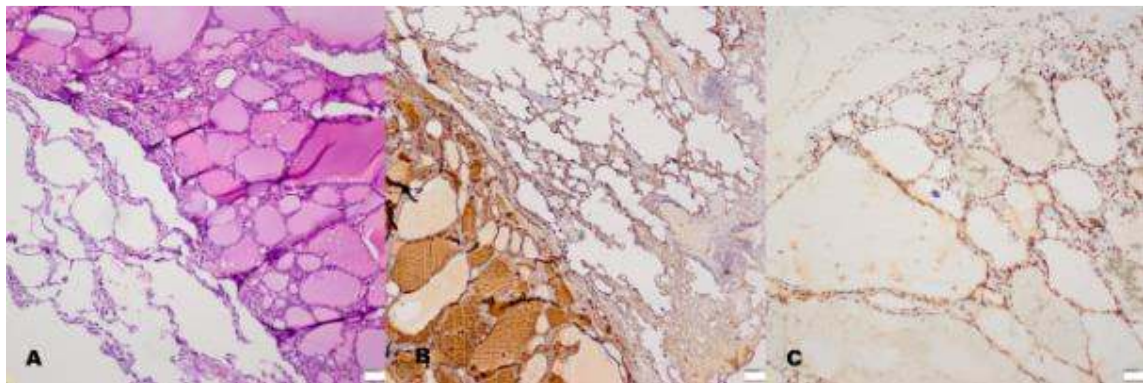


Figure 2

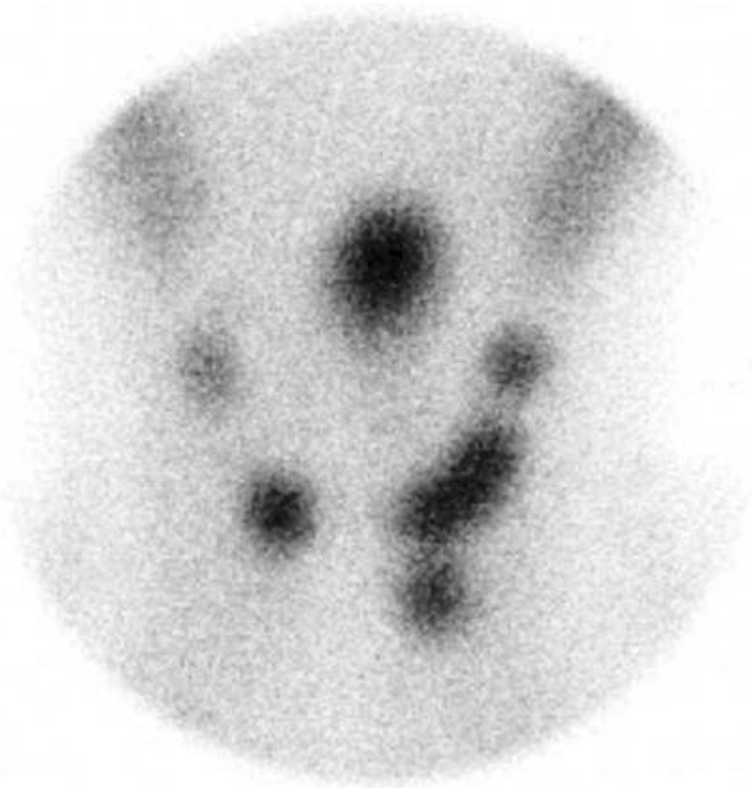


Figure 3