Thyroid Hemiagenesis Associated with Papillary Thyroid Carcinoma and Central Compartment Lymph Node Metastasis

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－ ABSTRACT －

Thyroid hemiagenesis is a rare form of thyroid congenital abnormally, in which one thyroid lobe fails to develop. Presence of carcinoma in a patient with thyroid hemiagenesis is an extremely rare. Case reports and a review of the world literature concerning thyroid hemiagenesis with papillary thyroid cancer are presented. A 47-year-old female patient without previous thyroid surgery history presented with an incidentally discovered right thyroid nodule. The left thyroid was not seen by ultrasonography and computed tomography. Fine-needle aspiration of right thyroid nodule showed that positive finding for papillary thyroid carcinoma. A total thyroidectomy with central compartment neck dissection was performed. The pathologic finding was confirmed to be well-differentiated papillary thyroid microcarcinoma and metastatic right paratracheal lymph nodes. To our knowledge, this case represents the first reported thyroid hemiagenesis associated with papillary thyroid microcarcinoma and central compartment lymph node metastasis. **(J Clinical Otolaryngol 2013;24:311-313)**

KEY WORDS : Thyroid hemiagenesis · Papillary thyroid carcinoma · Lymph node metastasis.

Introduction

Thyroid hemiagenesis is a rare form of thyroid congenital abnormally, in which one thyroid lobe fails to develop. Although the exact pathogenesis of thyroid morphogenesis and descent is unknown, some genetic abnormalities (TSH-R and PAX8 genes) identify in few cases.[1,2)] Absence of the left lobe is more common than absence of the right lobe.[3,4)] The prevalence of thyroid hemiagenesis in the normal population is from 0.025% to 0.05%.3,4 Among the pathologic conditions commonly occurring in the residual lobe are thyroid adenoma, multinodular goiter, Grave's disease, and chronic thyroiditis. Presence of carcinoma in a patient with thyroid hemiagenesis is very rare. Here we reported a case presented with papillary thyroid cancer and central compartment lymph nodes metastasis associated with congenital thyroid hemiagenesis and we also review the relevant literature.

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Case Report

A 47-year-old female patient without previous thyroid surgery history presented with an incidentally discovered right thyroid nodule. Ultrasonography revealed an ill defined solid nodule at mid-portion of right thyroid gland, measuring 0.6 × 0.5 cm in size (Fig. 1). But the left thyroid was not seen by ultrasonography and computed tomography (Fig. 2). Fine-needle aspiration of right thyroid nodule showed that positive finding for papillary thyroid carcinoma.

A total thyroidectomy with central compartment neck dissection was performed with preservation of

the right parathyroid glands and recurrent laryngeal nerve. Left thyroid gland was not seen in the operation field. The pathologic finding was confirmed to be well-differentiated papillary thyroid microcarcinoma and metastatic right paratracheal lymph nodes.

Discussion

Thyroid hemiagenesis is a rare anomaly resulting from the failed embryologic development of a lobe of the thyroid. The prevalence in women is greater than in man (75% vs. 25%) in several previous studies based on observations in patients affected by thyroid disease.[5-7] However, the higher estimated prevalence of thyroid hemiagenesis in women is possible consequence of the fact that thyroid diseases are more frequent in women than in man. When the estimated ratio is recalculated based on equal numbers of females and males, the female to male ration is 1.3 : 1.[8] There is no sex preponderance in the study about the prevalence of thyroid hemiagenesis in normal population.[4]

There has been considerable disagreement regarding the compensation hypertrophy and thyroid dysfunction of the intact thyroid lobe. Maiorana et al.[4] reported that the compensatory hypertrophy of the residual thyroid lobe occurred in most, but not all cases, and was due to thyroid tissue overstimulation by TSH. They would recommend systemic follow-up of all identified cases of thyroid hemiagenesis because of the high risk of goiter and hypothyroidism. However, Gursoy et al.[3] reported that no compensatory increase in intact thyroid lobe volume is noted and there was no association with other thyroid malformation or dysfunction.

Associated diseases in the remaining thyroid lobe include benign adenoma, multinodular goiter, hypothyroidism, Grave's disease, and chronic thyroiditis.[8-10] However, Co-occurrence of thyroid hemiagenesis and papillary thyroid carcinoma is extremely rare.[6,11,13] There is no consensus regarding the optimal extent of surgical resection of papillary thyroid carcinoma with thyroid hemiagenesis due to rarity. Although one thyroid lobe fails embryologic development in thyroid hemiagenesis, all four parathyroid glands present in normal position.[6]

Conclusion

Thyroid hemiagenesis is almost always discovered

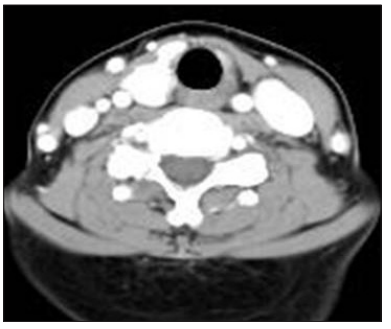

Fig. 2. Computed tomography showing hemiagenesis of the left thyroid gland.

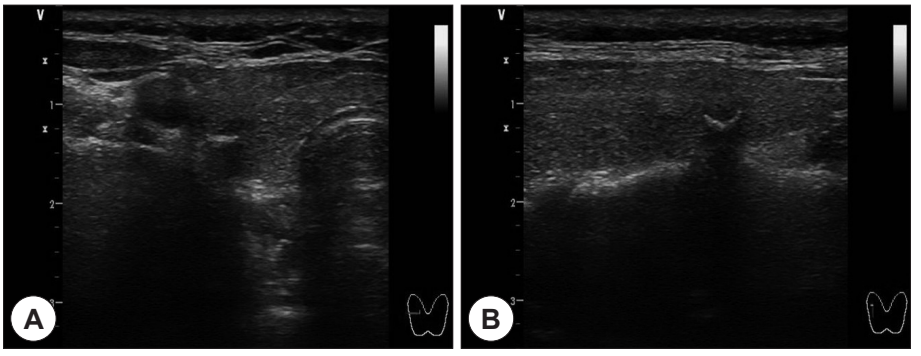

Fig. 1. Ultrasonographic image. Transverse (A) and longitudinal (B) images of right thyroid gland show an ill-defined, tall-than-wide, coarse calcification, irregular shape, and hypoechoic nodule.

incidentally while searching for a contralateral pathological abnormality. If nodular thyroidal disease is associated with residual thyroid gland, fine needle aspiration cytology should be performed in order to rule out primary malignancy. To our knowledge, this case represents the first reported thyroid hemiagenesis associated with papillary thyroid microcarcinoma and central compartment lymph node metastasis.

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