

# Thyroid Hemiagenesis Associated with Papillary Carcinoma

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This paper reports a rare case of a left sided thyroid hemiagenesis associated with papillary carcinoma. Thyroid hemiagenesis is a rare congenital anomaly resulted from failure of development on one of the thyroid lobes. Among several thyroid developmental anomalies, hemiagenesis is considered to be the rarest anomaly. In world literature to date, only about 310 cases have been reported. We present a case of thyroid hemiagenesis of the left lobe and isthmus in a 51-year-old female, along with radiological and operative findings. Our report can give the idea that the thyroid hemiagenesis may not influence the development of parathyroid glands.

**Key Words:** Thyroid gland, Hemiagenesis, Carcinoma

## Introduction

Thyroid hemiagenesis is a rare congenital anomaly resulted from failure of development on one of the thyroid lobes. Among several thyroid developmental anomalies, hemiagenesis is considered to be the rarest anomaly.<sup>1)</sup> The exact incidence of thyroid hemiagenesis is not clear as the patients are often asymptomatic and most cases may remain undiscovered unless they become symptomatic. The estimated prevalence of thyroid hemiagenesis is about 0.05% to 0.2%.<sup>2,3)</sup> and in world literature to date, only about 310 cases have been reported.<sup>4)</sup> Malignancy associated with thyroid hemiagenesis is extremely rare.<sup>5)</sup> The clinical features, radiological and operative findings of this unusual case are presented with a review of literature.

## Case Report

A 51-year-old female was referred for treatment of thyroid mass located on the right lobe. On history taking, the patient had no previous history of neck surgery or radiation of the neck. Clinical examination revealed palpable multiple nodules on the right thyroid and the trachea was on central position. No abnormality was noted in the left side of the neck and no other mass was palpable in the central compartment of the neck. The rest of the examination, including fiberoptic nasopharyngoscopy and laryngoscopy was normal. No previous operative scar was found.

Previously the patient ran through evaluation for incidentally detected thyroid mass during cervical spine CT scan at local hospital. The CT scan showed absence of left lobe and isthmus of the thyroid. It also revealed several thyroid nodules on the remaining lobe. Neck ultrasonography demonstrated five thyroid

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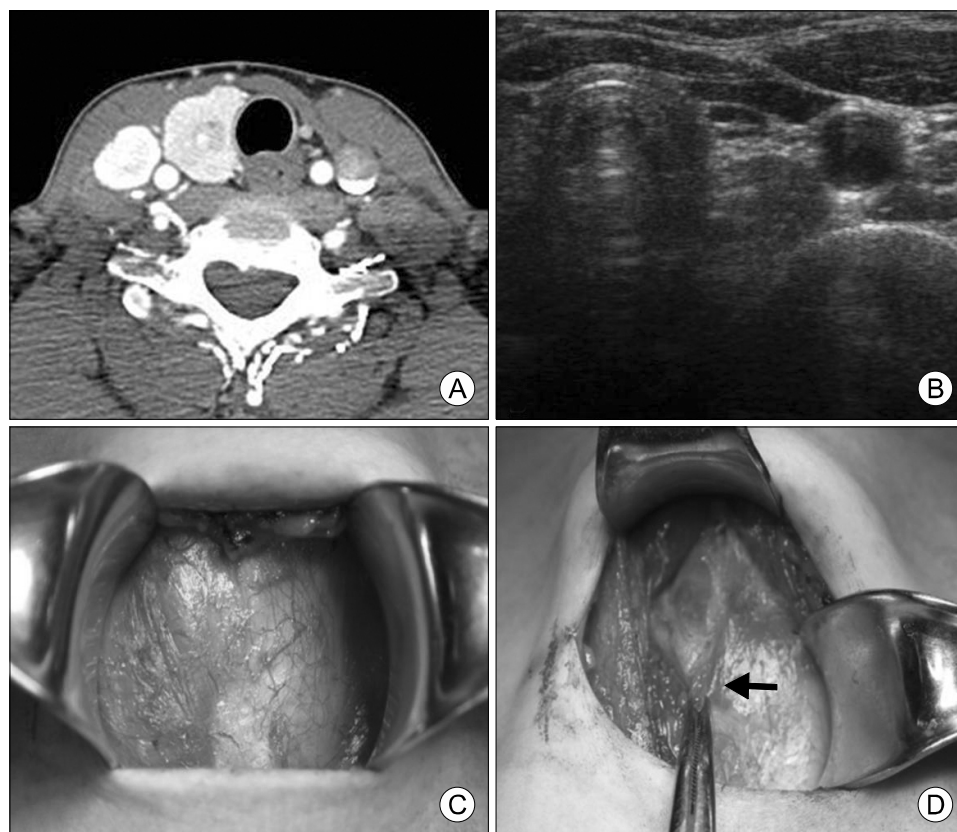
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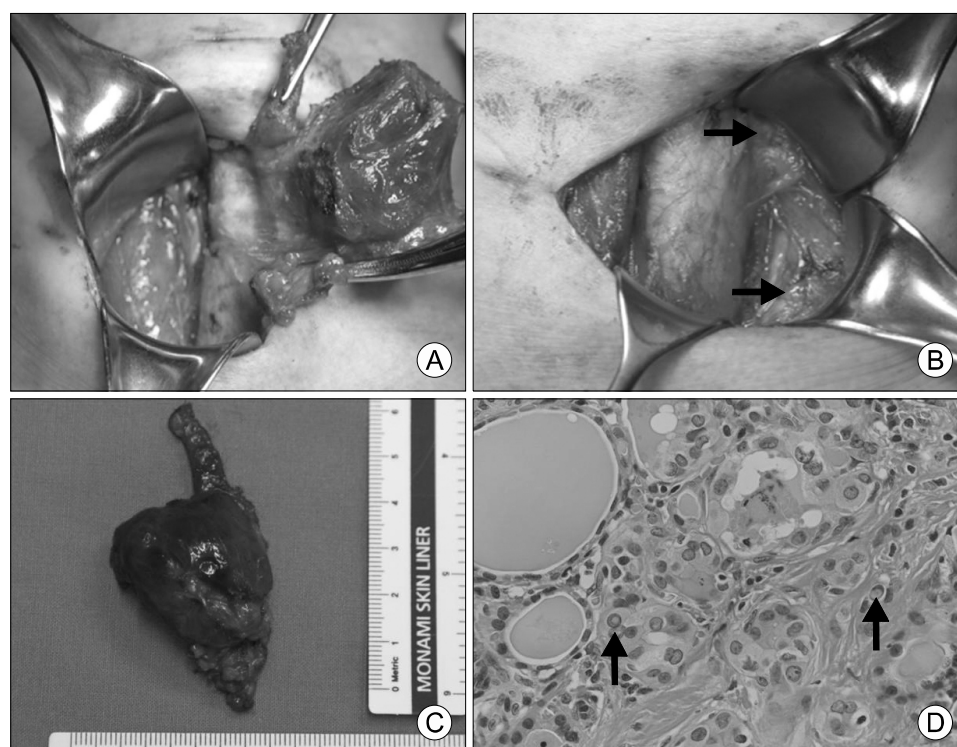
nodules including a 4 mm sized nodule, which was suspicious for malignancy (Fig. 1). Neither left lobe nor isthmus of the thyroid was found. The fine-needle as-

piration of the right thyroid lobe favored this to be papillary carcinoma (Fig. 1).

Other diagnostic evaluation was done, including



**Fig. 1.** (A) A CT scan of neck showing left hemiagenesis and multiple nodules on the right thyroid. (B) A neck ultrasound showing left thyroid hemiagenesis. (C) Intraoperative finding revealing left thyroid hemiagenesis with isthmus agenesis (C) and existence of the pyramidal lobe (arrow) (D).



**Fig. 2.** (A) An operative image taken just before the right thyroid lobectomy. (B) An image showing left parathyroid glands in normal position (arrows). (C) A picture of the specimen after the operation, Note that left lobe and isthmus of the thyroid is absent. (D) Pathologic finding showed enlarged tumor cells with intranuclear pseudo inclusions (arrows). Original magnification  $\times 400$ .

thyroid function test, biochemical analysis and CT scan of the neck. All the results for blood investigations and biochemical analysis were within normal limit. The CT scan revealed multiple thyroid nodules in right lobe without visualization of left lobe and isthmus of the thyroid. There was no evidence of cervical lymphadenopathy.

Based on these findings, a diagnosis of thyroid hemiagenesis of the left lobe and thyroid cancer on the right lobe was made. The patient underwent surgery with standard horizontal skin incision. Intraoperative finding was right lobe with multiple nodules. A confirmation of complete absence of the left lobe and isthmus was possible (Fig. 2). A total thyroidectomy and central neck dissection was done. Inspection of left side of the neck was done. Left upper and lower parathyroid glands were identified in normal position (Fig. 2).

In the pathology report, the final diagnosis of papillary microcarcinoma with tumor size of 0.3 cm was reported. Parathyroid gland and metastatic lymph node were not found in the specimen.

## Discussion

The embryological development of the thyroid gland begins during the fourth week of fetal gestation, with a median endodermal diverticulum on the floor of the pharynx at the foramen cecum. Normally, This primordium of the thyroid gland elongates and forms a bilobed diverticulum, which descends to a final position anterior to the trachea in the seventh week of gestation.<sup>1)</sup> Suggested mechanism of hemiagenesis is failure of bilobe formation of the primordium. The development of the thyroid gland is orchestrated by several series of transcription factors such as TTF1, TTF2 and pax-8.<sup>6)</sup> Dysfunction in these genes plus other environmental factors may contribute to the thyroid hemiagenesis.

The embryological development of parathyroid gland is another story. Parathyroid glands develop from the pharyngeal pouches. The superior parathyroid glands are developed from the 4th pharyngeal pouch and the inferior parathyroid glands are developed from the 3rd pharyngeal pouch, respectively. They begin to differ-

entiate in five to six week of gestation, which is followed by descent. Superior parathyroid and inferior parathyroid migrate with thyroid gland and thymus respectively, then disconnected to their normal position.

The prevalence of thyroid hemiagenesis is estimated to be about 0.05 to 0.2%.<sup>2,3)</sup> The involvement can occur in either lobe with or without isthmus hemiagenesis. The left lobe involvement contribute to 80% of hemiagenesis.<sup>7)</sup> Thyroid hemiagenesis accompanied by isthmus agenesis occurs in 50% of the cases.<sup>8)</sup> The absence of the left lobe is more frequent than the right lobe, with left:right ratio of 3:1. Thyroid hemiagenesis is estimated to be predominant in females with a ratio of 3:1. However, this is may be the result of bigger populations investigated in females than males. Moreover, recent study in normal children showed there was no difference between the genders, in the prevalence of thyroid hemiagenesis.<sup>2)</sup>

Thyroid hemiagenesis itself does not cause clinical symptoms. This leads to result that the hemiagenesis is usually detected with other accompanied thyroid diseases. Those thyroid diseases include Graves' disease, Hashimoto's thyroiditis, subacute thyroiditis, nodular goiter, hyperfunctioning adenoma, primary carcinoma as our case, and metastatic carcinoma.

Diagnosis of thyroid hemiagenesis is usually done by imaging methods, including ultrasonography, CT scan, MRI, and thyroid scanning. In our case the diagnosis was done incidentally after taking CT scan of neck. Clinical exam may give a clue to the diagnosis as the tracheal rings are easy to palpate on the affected side and border of sternocleidomastoid muscle may be shifted to midline. However, careful exam is necessary as it may mimic thyroid solitary nodule. The prevalence of thyroid hemiagenesis may increase, since office based neck ultrasound exam is widely performed around the world today.

In our case, the patient had left sided thyroid hemiagenesis with isthmus agenesis, which was accompanied with papillary carcinoma. The patient underwent right thyroid lobectomy (might call it total thyroidectomy in this case), which revealed all four parathyroid glands in normal position. However, There were no reports that included operative findings

showing parathyroid glands in thyroid hemiagenesis. Our report can give the idea that the thyroid hemiagenesis may not influence the development of parathyroid glands. This may be the result of the difference in developmental lineage thyroid and parathyroids take.

The surgeons considering thyroidectomy should be aware of this anatomical variation that the thyroid lobectomy, in the patient, can result in total loss of thyroid function. Further study is needed to reveal the exact developmental mechanism of thyroid hemiagenesis and the relationship between coexisting diseases.

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