

RIGHT THYROID HEMIAGENESIS ASSOCIATED WITH PAPILLARY THYROID CANCER AND AN ECTOPIC PRELARYNGEAL THYROID: A CASE REPORT

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Abstract: Failure of embryologic development of a lobe of the thyroid gland is a rare anomaly. We report a 47-year-old female with right thyroid hemiagenesis, papillary thyroid cancer, and an ectopic prelaryngeal thyroid rest. The preoperative diagnosis was made as a result of evaluating a progressively enlarging left thyroid mass by means of ultrasonography, technetium-99m thyroid scintigraphy, and fine-needle aspiration cytology. During the operation, an isthmus and an enlarged left lobe with a cancer mass 2 cm in diameter were found, and a solitary piece of thyroid tissue about 0.7 cm in length was present just below the hyoid bone. We conclude that recognition of this rare anomaly by means of both scintigraphy and ultrasonography is valuable for a complete thyroidectomy, and for avoiding unnecessary contralateral dissection and damage.

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Key words:
thyroid hemiagenesis
thyroid cancer
ectopic thyroid
scintigraphy
ultrasonography

The congenital absence of one thyroid lobe is a rare anomaly. About 130 cases of thyroid hemiagenesis have been reported with an incidence of about 0.01 to 0.07% [1–16]. The presence of carcinoma in a patient with hemiagenesis is quite rare, and very few cases have been reported. We report a case with thyroid hemiagenesis, papillary thyroid cancer, and an ectopic prelaryngeal thyroid rest.

ectopic prelaryngeal thyroid, the absence of a right lobe (Fig. 2), and a cold lesion in the left lobe. The serum concentration of thyroxine was 7.1 µg/dL (normal, 4.5–12.5 µg/dL), of triiodothyronine was 119 ng/dL (normal, 86–187 ng/dL), of

Case Report

A 47-year-old woman was seen for a painless mass in the anterior cervical region that had gradually enlarged in size over a period of 2 months. There was no history of radiation therapy or operation, nor family history of thyroid disorders. Physical examination revealed an elastic mass 2 cm in diameter on the left thyroid gland. Ultrasonography showed an intrathyroid mass in the left thyroid gland and no thyroid tissue on the right side (Fig. 1). Fine-needle aspiration biopsy revealed a suspected papillary thyroid carcinoma. Technetium-99m imaging revealed a functional left lobe with an isthmus shaped like a hockey stick, a separate functional



Fig. 1. Neck transverse ultrasonogram demonstrating the absence of the right lobe of the thyroid gland and the presence of a hypoechoic mass (arrow) 2 cm in diameter in the left lobe of the thyroid gland.

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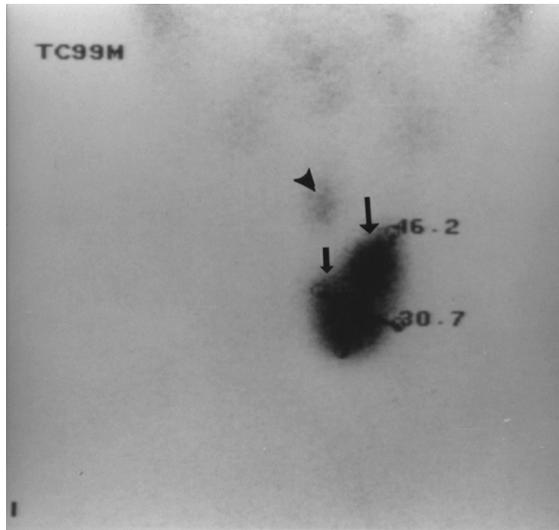


Fig. 2. Thyroid scan showing the absence of the right lobe with good uptake in the left lobe (long arrow) and isthmus of the thyroid (short arrow), which is shaped like a hockey stick, and a functional ectopic prelaryngeal thyroid rest (arrowhead).

thyroid stimulating hormone was 2.8 $\mu\text{U}/\text{mL}$ (normal, 0.5–5.6 $\mu\text{U}/\text{mL}$), and of thyroglobulin was 2.2 ng/mL (normal, 1.6–55.2 ng/mL). At operation, the right lobe was absent, while the isthmus and an intact left lobe with a mass 2 cm in diameter were found. A totally separate thyroid rest about 0.7 cm in length was present just below the hyoid bone (Fig. 3). The left parathyroid glands and left recurrent nerve were in their normal positions. A left lobectomy, isthmectomy, and excision of the pyramid lobe were performed. The pathologic

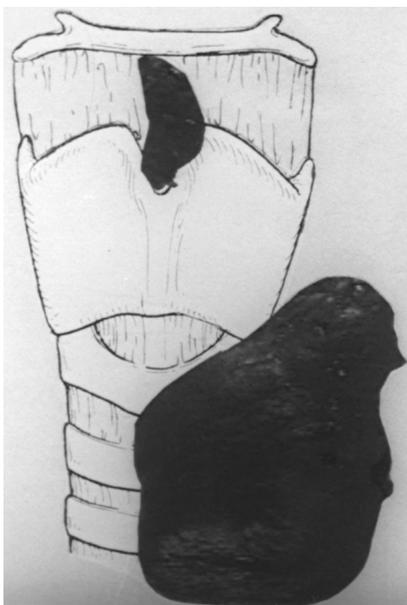


Fig. 3. Left thyroid lobe and ectopic prelaryngeal thyroid after removal.

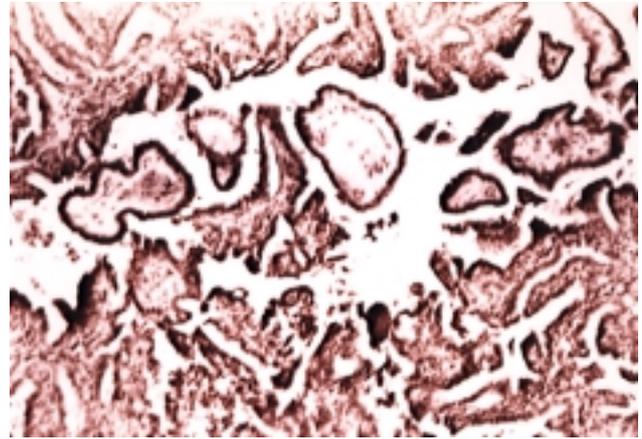


Fig. 4. Histologic examination of the thyroid mass showing papillary adenocarcinoma. (Hematoxylin and eosin, x 100)

diagnosis was papillary cancer (Fig. 4). Postoperatively, radioiodine 131 for cancer work-up showed no residual thyroid uptake. The postoperative course was uneventful, and the patient received thyroxine 150 $\mu\text{g}/\text{day}$ for replacement therapy and regular follow-up.

Discussion

The potential thyroid gland can initially be recognized during the fourth week of fetal life on the endoderm of the floor of the pharynx at the foramen cecum as a midline thickening from which the thyroglossal duct develops. With development of the neck and descent of the heart, this becomes elongated, eventually forming the new bilobular diverticulum that will become the thyroid gland proper. During its caudal migration, the bilobular diverticulum remains connected to the floor of the mouth by the thyroglossal duct, which usually disappears subsequently. The normal adult thyroid gland is composed of two lateral lobes connected by an isthmus [17]. Anomalies of embryonic development of the two lobes result in a large variety of shapes and sizes. Very rarely, the entire thyroid gland, one or both lobes, or the isthmus may not develop. The cause of one-lobe agenesis has been postulated to be a failure of the thyroid anlage to become bilobular, followed by unilateral migration [8]. Normally, the epithelium of the thyroglossal duct disappears. Occasionally, the duct may form thyroglossal cysts or fistulas, and the epithelium may persist or differentiate into thyroid tissue at any level. Midline ectopic thyroid rests are the result of failure of, or incomplete, descent of the thyroglossal duct and of abnormal development of its epithelium.

Complete failure of descent of the thyroglossal duct results in a lingual thyroid, located at the base of the tongue. Accentuated descent causes an intrathoracic thyroid gland. Other midline ectopic thyroid rests of the thyroglossal duct may be found below or above the hyoid bone. The incidence of ectopic thyroid gland is approximately 1 per 100,000 persons according to Mayo Clinic records [18], and one in three to four thousand people with thyroid disease will exhibit ectopic tissue [19]. The present case showed both lobar agenesis and a midline ectopic thyroid.

The true incidence of thyroid hemiagenesis is difficult to determine, since the diagnosis is usually made in an abnormal population being evaluated for some other thyroid pathology. Poate and Wyndham reported five patients with thyroid hemiagenesis out of 45,369 cases at Mayo Clinic [20]. Similarly, Heimann and Mortensen reported five thyroid hemiagenesis cases among 700 cases [21], Harada et al seven among 12,456 cases [1], while Williams et al reported 10 from 29,004 autopsies. Melnick and Stemkowski reviewed the literature and found that the left lobe was absent in 80% of cases, and the right lobe was absent in 20% of cases (a left-to-right hemiagenesis ratio of 4:1) [4]. They found that the isthmus was absent in 50% of patients where the isthmus was specifically mentioned. Females accounted for 75% and males for 25% of cases, giving a female-to-male ratio of 3:1 [3]. Since thyroid disorders develop predominantly in women, women may have more opportunity to receive thyroid examination and the ratio of females to males may not be conclusive. Clinically, thyroid hemiagenesis may present as a unilateral neck mass caused by compensatory hypertrophy of the contralateral lobe. In fact, thyroid hemiagenesis is usually found when the patient has another type of thyroid disorder. The associated diseases in the remaining thyroid lobe include benign adenoma, multinodular goiter, hyperthyroidism, chronic thyroiditis, and, very rarely, carcinoma [4-7]. The most common pathology involved in thyroid hemiagenesis is hyperthyroidism [1, 4, 5].

The diagnosis of thyroid hemiagenesis should be considered in any patient with unilateral absence of function as assessed by thyroid scintigraphy. Thyroid scan may be unable to distinguish thyroid hemiagenesis from a large cold nodule involving the entire lobe, unilateral inflammatory disease, or a hot nodule suppressing the contralateral lobe [8]. Thyroid hemiagenesis with an isthmus present appears as a hockey-stick shape on thyroid scintigraphy [4]. As in the present case, the ectopic prelaryngeal thyroid is easily recognized by scintigraphy. Ultrasonography, computerized tomography, or magnetic resonance imaging should also be applied for diagnostic confirmation of thyroidal hemiagenesis. The present case

demonstrates that ultrasonography is a very convenient means of showing the absence of the thyroid gland by transverse section over the trachea and both sides of the thyroid. Since half of the thyroid gland is sufficient to maintain normal thyroid function [22], patients with thyroid hemiagenesis usually have normal thyroid function. Although treatment of thyroid cancer with total thyroidectomy is still somewhat controversial, this procedure facilitates the detection of thyroid cancer metastasis and reduces the radioiodine ablation dose. Preoperative diagnosis of an ectopic prelaryngeal thyroid is helpful for the surgeon in completing a total thyroidectomy. Preoperative recognition of thyroid hemiagenesis in patients undergoing a thyroidectomy is important to avoid unnecessary contralateral neck exploration, which may increase the risk of parathyroid or recurrent nerve injury.

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