Thyroid Hemiagenesis: An Incidental Discovery during Treatment for Papillary Thyroid Carcinoma

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Abstract

Thyroid hemiagenesis (THA) is a rare congenital abnormality in which one of the thyroid lobes fails to develop normally. The prevalence rates range from 0.02% to 0.25%, with no gender preponderance and most commonly involve the left lobe. We report a case of left THA in a 37-year-old female that was found incidentally during total thyroidectomy for right papillary thyroid carcinoma. Most patients with THA are asymptomatic, however, may have subclinically raised thyroid-stimulating hormone levels or be predisposed to developing thyroid disorders such as nodular goiter, hyperthyroidism, Grave's disease, Hashimoto's thyroiditis, hyperparathyroidism, or malignancy. Usual imaging techniques such as ultrasonography, computed tomography, and magnetic resonance imaging allow the confirmation of diagnosis and provide clearer glandular and regional visualization. Additional modalities such as thyroid scintigraphy enable the functional assessment and identification of any suspicious nodules and aid in decision-making. THA is a very uncommon condition and the diagnosis is primarily made through imaging. Clinicians should be aware that this condition exists along with documented associations of concomitant thyroid disorders and malignancy.

Keywords: Case report, papillary thyroid carcinoma, thyroid agenesis, thyroid dysgenesis

Introduction

Thyroid hemiagenesis (THA) is a rare congenital abnormality in which one of the thyroid lobes fails to develop and was first described by Robert B. Todd in 1852.[1] The true prevalence of this condition is not known as most patients remain asymptomatic. Due to the rarity of this condition, there are no guidelines or protocols for the clinical implications of this condition or any of its associated disorders. We present a case of incidental detection of THA during surgery for papillary thyroid carcinoma (PTC) within the contralateral intact lobe and a review of the literature on this rare condition.

Case Report

A previously healthy 37-year-old female presented with a gradually enlarging anterior neck swelling over 7 months. She was asymptomatic and had no significant risk or family history of thyroid malignancies. She was clinically euthyroid and physical examination revealed a nontender, firm 3 cm × 3 cm swelling at the anterior neck that moved with deglutition. There was no palpable regional lymphadenopathy and general systemic examination was unremarkable.

Her routine and biochemical blood works were normal, with a thyroid-stimulating hormone (TSH) of 1.177 μIU/mL (normal value: 0.35–4.94) and T4 of 12.67 pmol/L (normal value: 9.01–19.05). Ultrasonography of her neck revealed a thyroid nodule at the right lobe measuring 2 cm × 3 cm × 3.3 cm with suspicious features (American College of Radiology-Thyroid Imaging Reporting and Data Systems 5) [Figure 1a]. The contralateral left thyroid lobe was unremarkable measuring 0.9 cm × 1.3 cm × 2.5 cm [Figure 1b]. The lesion also revealed an increase in Doppler flow within as opposed to the overlying thyroid tissue [Figure 1c]. Ultrasound-guided fine-needle

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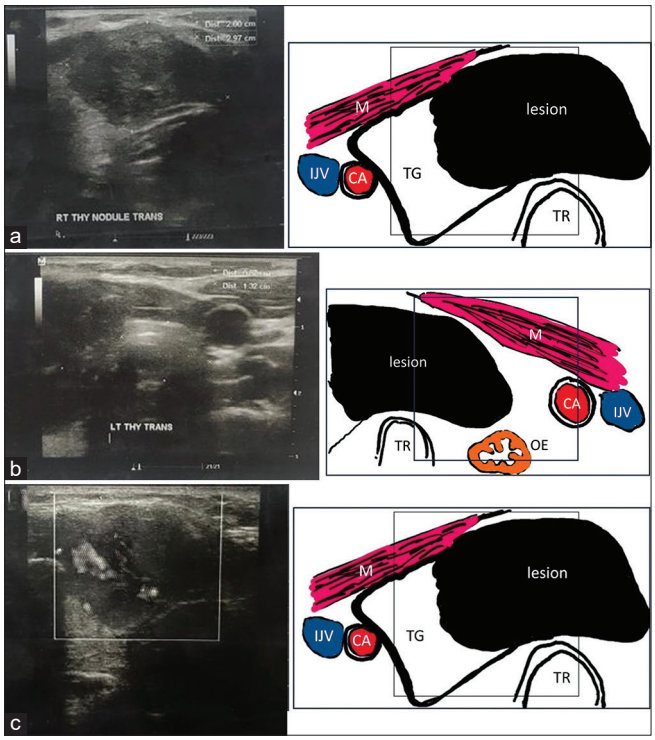

Figure 1: (a) Ultrasonography revealed a very hypoechoic almost entirely solid lesion at the right thyroid lobe involving the isthmus measuring 2.0 cm x 3.0 cm (AP x W) with an increase in Doppler flow (not shown). An illustration displaying the ultrasonographic findings. (b) There was an absence of left thyroid tissue at the left thyroid bed. Left thyroid tissue is not seen at its expected location anterior to the oesophagus. The lesion seen earlier crosses the midline extending slightly to the left. (c) Transverse view across the lesion showing an increase in Doppler flow within as opposed to the overlying thyroid tissue. An illustration displaying the ultrasonographic findings. CA: Carotid artery, IJV: Internal jugular vein, M: Muscle; OE: Oesophagus, TG: Thyroid gland, TR: Trachea

aspiration of the right thyroid nodule revealed cytology consistent with PTC (Bethesda VI).

She underwent open total thyroidectomy with intraoperative nerve monitoring (IONM) through a standard anterior neck collar incision. Intraoperatively, there was a large hard thyroid nodule at the thyroid isthmus involving the medial segment of the right lobe measuring 3 cm × 2 cm [Figure 2]. During contralateral dissection, there was no glandular tissue in the left thyroid bed. A 1.5 cm × 0.5 cm piece of adipose-like tissue was seen connected to the isthmus in place of the left lobe [Figure 3]. Vessels that are likely synonymous with the superior pole vessels were seen supplying this piece of tissue. The left superior and inferior parathyroid glands were identified in their usual anatomical positions along with the recurrent laryngeal nerve in the trachea-oesophageal groove which was confirmed with IONM stimulation. There were no enlarged central compartment lymph nodes.

The specimen was sent for histopathological examination and the final report of PTC was made (pT2). The left lobe consisted of only adipose tissue with no thyroid tissue seen. The patient made an uneventful recovery and started on oral levothyroxine

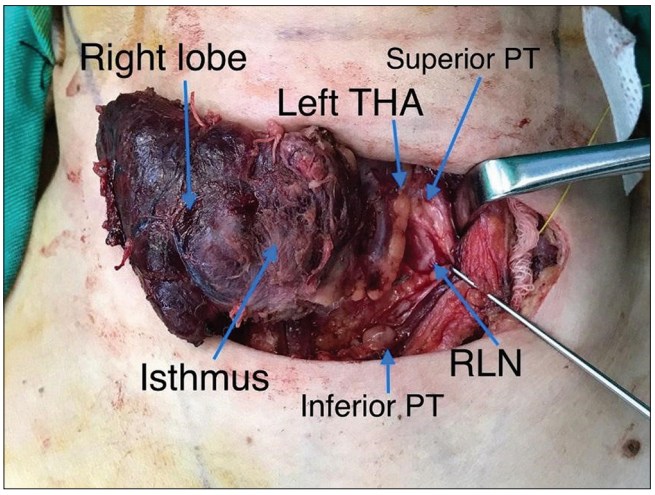

Figure 2: Intraoperative total thyroidectomy specimen. PT: Parathyroid gland, RLN: Recurrent laryngeal nerve

supplementation. She was also referred for adjuvant radioactive iodine ablation and continues to be monitored periodically.

Discussion

The true prevalence of THA in the population is not known, but several epidemiological studies with modest recruitment have reported a prevalence rate of between 0.02% and 0.25%.[2-4] In one of the largest published single-population-based thyroid screenings of almost 300,000 individuals in Japan, the prevalence of congenital thyroid agenesis and THA was 0.02% with no statistically significant gender predominance.[4] In all of these studies, left-sided agenesis was most frequently documented followed by right and isthmus agenesis. Patients with THA remain largely asymptomatic throughout their lives and detection is usually incidental or during the investigation for associated thyroid disorders. The size of the intact lobe appears to be significantly increased compared to the ipsilateral lobe volume in bilobed thyroids and may be likely due to compensatory hyperplasia.[4] Mean TSH and T3 levels were found to be higher than controls, but interestingly, there was no significant difference in T4 levels.[4-6] This has led to the postulation that in these patients, a different set point on the hypothalamus–pituitary–thyroid axis exists due to chronic TSH hyperstimulation to maintain normal hormone levels and function.

Ultrasonography remains the imaging modality of choice for the detection of thyroid disorders "as" it is sensitive "to identify" the anatomy of the lobe and "visualise" any obvious structural abnormalities, relatively affordable without risk of radiation, albeit being "an" operator dependent.[6] Additional modalities such as thyroid scintigraphy would be able to provide functional imaging of the thyroid gland along with any ectopic thyroid tissue as well as suspicious nodules not otherwise detected by ultrasonography.[7] Computed tomography is sometimes used to provide better delineation of the anatomy and adjacent structures.

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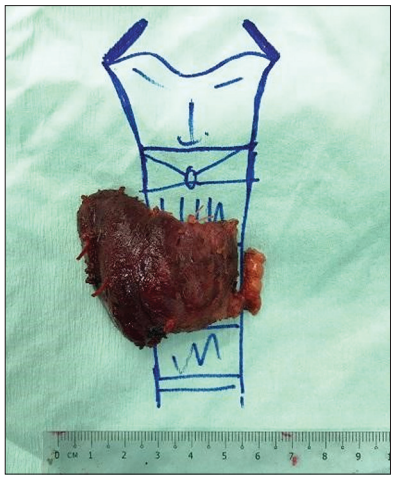

Figure 3: Postoperative image of total thyroidectomy specimen

Patients with THA have been reported to have an increased incidence of concomitant thyroid disorders including nodular goiter, hyperthyroidism, Grave's disease, Hashimoto's thyroiditis, and even hyperparathyroidism.[8,9] TSH has goitrogenic properties and has been associated with the development of nodular or diffuse goiters, which is also linked with an increased risk of malignancy.[10] Ruchala *et al*. and Szczepanek-Parulska *et al*. have both reported a significant number of THA patients with positive thyroid autoantibodies compared to control groups and this has led to the opinion that these patients may be more likely to develop thyroid disorders throughout the course of their lives.[5,8] Thyroid malignancy has been reported in 23 cases of patients with THA, with PTC being the predominant type.[9] However, due to the small number of cases in this already small cohort of THA patients, there is insufficient evidence to conclude a definitive association with the development of malignancy.

Conclusion

The fact remains that THA is a very uncommon condition and the diagnosis is primarily made via imaging. Clinicians should be aware that this condition exists along with documented associations of concomitant thyroid disorders and malignancy.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent form. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Quain R, Wheatstone C. The Cyclopaedia of Anatomy and Physiology. Vol. 1836. London: Sherwood, Gilbert, and Piper; 1836. p. 1-1126.
2. Korpal-Szczyrska M, Kosiak W, Swieton D. Prevalence of thyroid hemiagenesis in an asymptomatic schoolchildren population. Thyroid 2008;18:637-9.
3. Gursoy A, Anil C, Unal AD, Demirer AN, Tutuncu NB, Erdogan MF. Clinical and epidemiological characteristics of thyroid hemiagenesis: Ultrasound screening in patients with thyroid disease and normal population. Endocrine 2008;33:338-41.
4. Suzuki S, Midorikawa S, Matsuzuka T, Fukushima T, Ito Y, Shimura H, *et al.* Prevalence and characterization of thyroid hemiagenesis in Japan: The Fukushima health management survey. Thyroid 2017;27:1011-6.
5. Ruchala M, Szczepanek E, Szaflarski W, Moczko J, Czarnywojtek A, Pietz L, *et al.* Increased risk of thyroid pathology in patients with thyroid hemiagenesis: Results of a large cohort case-control study. Eur J Endocrinol 2010;162:153-60.
6. Mikosch P, Gallowitsch HJ, Kresnik E, Molnar M, Gomez I, Lind P. Thyroid hemiagenesis in an endemic goiter area diagnosed by ultrasonography: Report of sixteen patients. Thyroid 1999;9:1075-84.
7. Gurleyik G, Gurleyik E. Thyroid hemiagenesis associated with hyperthyroidism. Case Rep Otolaryngol 2015;2015:829712.
8. Szczepanek-Parulska E, Zybek-Kocik A, Woliński K, Czarnocka B, Ruchała M. Does TSH trigger the anti-thyroid autoimmune processes? Observation on a large cohort of naive patients with thyroid hemiagenesis. Arch Immunol Ther Exp (Warsz) 2016;64:331-8.
9. Lesi OK, Thapar A, Appaiah NN, Iqbal MR, Kumar S, Maharaj D, *et al.* Thyroid hemiagenesis: Narrative review and clinical implications. Cureus 2022;14:e22401.
10. Pedrinola F, Rubio I, Santos CL, Medeiros-Neto G. Overexpression of epidermal growth factor and epidermal growth factor-receptor mRNAs in dyshormonogenetic goiters. Thyroid 2001;11:15-20.