Case Report

Thyroid Embryonic Anomalies Involving the Medial and Lateral Anlagen: Two Surgical Case Reports

Ferdinand Rico and John Lung

Department of Surgery, Texas Tech University Health Sciences Center, 1400 S Coulter, Amarillo, TX, USA 79106

Correspondence should be addressed to Ferdinand Rico; ferdinand.rico.md@gmail.com

Received 28 September 2019; Accepted 19 November 2019; Published 5 December 2019

Academic Editor: Boris Kirshtein

Copyright © 2019 Ferdinand Rico and John Lung. This is an open access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

1. Introduction

The thyroid gland develops from the median bud of the pharynx from the foramen cecum to the thyroid isthmus. An ectopic thyroid tissue is where the thyroid gland is located other than anterior to the trachea in the lower neck due to aberrations in embryogenesis [1]. Most patients with ectopic thyroids do not have symptoms, but obstructive symptoms and hypothyroidism have been reported in cases [2]. The prevalence of an ectopic thyroid is estimated around 1 per 100,000 to 300,000 people and occurs in 1 in 4,000-8,000 people with thyroid disease [3]. Ectopic thyroid tissue lateral to the normal midline location is rare. The exact definition of lateral ectopic thyroid tissue is not defined consistently in the literature with some reports defining it as lateral to the carotid sheath and jugular vein [4]. Others define it as an ectopic thyroid tissue embedded in or deep to the strap muscles that appears off the midline [5]. We present a case of a morbidly obese patient with an enlarged heterogeneous left lobe of the thyroid on imaging that had two ectopic masses near the left thyroid and a right persistent thyroid rest. The second case depicts a possible retained or a missed substernal thyroid that was accidentally discovered after a prior left thyroid lobectomy 4 years ago. We discuss the preoperative workup, surgical anatomy, postoperative management of both patients, and implications for future surgical practice.

2. Case Description

2.1. Case 1. A 43-year-old female was referred and evaluated in clinic with a chief complaint of a slowly enlarging neck mass. She had a past medical history of hypertension, morbid obesity, sleep apnea, and asthma. She was afebrile and denied any dyspnea, chest pain, or dysphagia. Focused physical exam noted a palpable left neck mass that moves with deglutition without clinical tracheal deviation. Upon review of her preoperative workup, her thyroid ultrasound revealed an enlarged heterogeneous left lobe of the thyroid. Computerized tomography (CT) imaging of the neck and thorax...
revealed a bilateral multinodular thyroid goiter with a left substernal anterior mediastinal extension and a 12.8 mm calcification on the left lower area. The adjacent trachea showed narrowing and rightward displacement due to the thyroid mass (Figure 1). A thyroid uptake (Iodine-123) scintigraphy showed a cold defect in the inferior pole of an enlarged left lobe (Figure 2).

Ultrasound-guided fine-needle aspiration cytology (US-FNAC) of the left thyroid nodule reported a possible papillary carcinoma.

Her operation started with a left thyroidectomy which was then converted to a total thyroidectomy with the use of intraoperative nerve monitoring (IONM) and IOUS. The left thyroid lobe was dissected with difficulty due to its larger left lobar size (10 × 7 × 5 cm), hypervascularity, and multinodularity (Figure 3). It was dissected meticulously and bluntly and was completely excised close to its capsule. The plane of dissection was clean, smooth, and left with no visible residual mass. After removal of the left thyroid, palpation revealed additional masses. A separate large left substernal mass (6 × 5 × 3.5 cm) was identified, transcervically dissected, and completely excised. Another large left mass was palpated directly posterior to the previously removed left lobe. The fat plane was opened up. This mass was also meticulously dissected and completely excised. This posteriorly located mass characteristically had a teardrop shape which appeared as a long remnant tract on the superior end. The mass measured about 8 × 5 × 2 cm and compressed laterally the carotid sheath. All of these masses were safely and completely excised (Figure 3). The recurrent laryngeal nerve (RLN) on the left was identified, well visualized, protected, and left intact with the aid of IONM. On further intraoperative evaluation including IOUS, the right thyroid lobe was identified as enlarged, hypervascular, and with palpable nodules. The pyramidal lobe was also observed as prominently enlarged. The lower pole of the thyroid was noted to have a tail-like extended protrusion distinctive of a persistent grade II thyroid rest. All enlarged lobes were dissected en bloc with the pyramidal lobe. The right multinodular thyroid lobe measured 8 × 4 × 3 cm in size (Figure 3).

A French #10 Jackson-Pratt drain was applied. She was discharged a few days later with supplemental prescriptions of calcium, vitamin D3, and levothyroxine.

2.2. Case 2. A 65-year-old female presented with a past medical history of hypertension, asthma, hypothyroidism, and GERD. Past surgical history included a right breast lumpectomy, cholecystectomy, 4 cesarian sections, hysterectomy, and a left thyroid lobectomy 4 years ago. She was referred after a CT scan was done which showed an incidental finding of a left substernal anterior mediastinal exophytic mass measuring 9 × 4 × 3 cm with rightward tracheal deviation (Figure 4). She was euthyroid and denied any compressive aerodigestive signs and symptoms. US-FNAC was consistent with a left substernal thyroid mass. A well-planned elective substernal thyroidectomy through median sternotomy was scheduled thereafter.
3. Discussion

Surgical removal of an enlarged thyroid is needed when obstructive symptoms are present or with a high suspicion for malignancy. The incidence of asymptomatic patients with thyroid nodules is increasing due availability of imaging such as ultrasound [6]. Management of an enlarged thyroid depends on American Thyroid Association recommendations based on characteristics of the nodule and the patient’s age, sex, radiation exposure, and other factors [7]. Radioiodine ablation is a nonsurgical option in both benign and malignant diseases with long-term complications including hypothyroidism [8].

Ectopic thyroid has been diagnosed positively by clinicians through preoperative imaging in our review of the literature [9]. Surgeons must be well adept in the knowledge of thyroid and parathyroid embryogenesis, with its diverse aberrance of embryologic, histologic, and anatomic development. All of this knowledge coupled with the surgeon’s experience and intuition is of utmost importance intraoperatively to prevent a retained or missed thyroid, especially if positive for malignancy.

Our first surgical case was intraoperatively interesting. Two left ectopic thyroid masses were found. A left substernal mass and another mass posterior to the left thyroid were developed. The right thyroid with its tail-like extension or protuberance was also considered aberrant. Only on further inspection and palpation along with IOUS were further ectopic thyroid masses and aberrant thyrothymic rests identified. Thus, in all partial or complete thyroidectomy cases,
surgeons should appreciate the normal anatomy of the thyroid. Moreover, the knowledge of embryogenesis is of utmost importance in recognizing ectopic thyroid masses. Knowledge of embryogenesis and anatomy is required to excise ectopic glands during the same operation, preventing a retained or missed thyroid gland.

Our second case describes a patient who had a left thyroid lobectomy 4 years ago. The primary care physician ordered a neck and chest CT recently for a suspected goiter. Incidentally, a left substernal anterior mediastinal mass was found compressing and significantly deviating the trachea to the right. US-FNAC showed a thyroid mass. This was suggestive of a missed and therefore a retained ectopic thyroid (a persistent median thyroid rest anlage or an unfused aberrant lateral anlage) during the initial left thyroid lobectomy operation.

A good surgical dissection and exposure aids in a safer operation. It starts from skin incision along the Langer lines. After the avascular subplatysmal plane is dissected, transection of the midline investing layer of superficial cervical fascia is next done to expose the visceral cervical space of the neck. Lateral retraction of strap muscles is needed for wide exposure of thyroid vessels especially on the superior area to identify and preserve the superior laryngeal nerve. Identification of the Tubercle of Zuckerkandl (TZ) is mandatory with its close anatomic relationship and variability with the RLN and parathyroid gland (PtG). This will prevent RLN injury and preserve the PtG without devascularization. The TZ has four grades: #0, unrecognizable; #1, thickening of the lateral edge of the thyroid lobe; #2, less than 1 cm; and #3, greater than 1 cm [10]. Prior to closure, final post procedure reevaluation with palpation and/or use of IOUS is needed not to miss any other mass. All of these dissection steps allow good visualization of the thyroid itself and/or any ectopic aberrant thyroid.

In normal embryogenesis, thyroid anlagen migrate from the floor of the primitive foregut to its location on the midline of the neck in front of the trachea [11]. If the developing thyroid does not follow this migration, the gland will develop ectopically [1]. Over 50% of thyroid dysgenesis cases have a correlation with an ectopic thyroid. Several genetic abnormalities were associated with impaired thyroid development including FoxE1, TTF-1, TTF-2, Pax-8, and HoxA3 [12, 13]. The thyroglossal duct is attached to the foramen cecum and thyroid gland during migration and normally involutes. Although debated among embryologists, two anlagen (median and lateral) are needed in the development of the thyroid. Normally, the median and lateral anlagen fuse during embryological development. According to some embryologists, the lateral thyroid anlage is derived from the ultimobranchial body, a descending diverticulum of the fourth pharyngeal pouch (as it embryologically descends along with the inferior parathyroid) [13]. The presence of a lateral thyroid anlage can explain the nonmidline ectopic thyroid tissue in the neck, since an arrest of migration of the lateral thyroid anlage would result in the failure of fusion with the medial anlage [11]. Our posterior ectopic thyroid and substernal ectopic thyroid could represent a persistent abnormality of the lateral thyroid anlage. Embryologic abnormalities and developmental variations of median thyroid anlagen, lateral thyroid anlagen, both anlagen, and neither anlage have been detailed extensively in previous literature [12].

Thyroid rests can also form, which are isolated normal tissue below the lower pole of thyroid in the line of the thyrothymic tract or in the upper anterior mediastinum. They are remnants from the thyroid descent of the median anlage during normal embryogenesis. Our dissected right thyroid lobe has a type II thyroid rest. A classification of thyroid rests from types I-IV was proposed in relation to the thyroid (Figure 5) [14].
After removing an ectopic thyroid, pathology is needed to exclude the possibility of a well-differentiated metastasis that replaced a lymph node [15]. In our case, all tissue samples were negative for any malignancy. There are few reported cases of dual or triple ectopic thyroids [1, 16–24]. Our case was not detected on preoperative workup including CT scan, ultrasound, and thyroid scintigraphy. Excellent surgical knowledge of the anatomy and embryology of the thyroid is necessary not to cause surgical complications including bleeding, RLN injury, relative or absolute hypoparathyroidism, and a missed ectopic thyroid [25]. The first case depicts excellent neck dissection and exposure of an enlarged hypervascular thyroid, with safe preservation of RLN (with use of IONM) and PtG. After a routine thyroidectomy prior to closure, surgeons should intraoperatively palpate the operative site including the retrosternal anterior mediastinal area through the neck incision. For the same reason, surgeons should use IOUS. In the second case, an incidental CT finding of retrosternal mass after a past surgical history of left thyroid lobectomy was found to be a thyroid mass.

In summary, an experienced and intuitive neck surgeon should know the normal thyroid anatomy and its embryonic development. A surgeon should identify any congenital aberrant anomalies in the perioperative period and prevent future complications. Surgeons must keep in mind the possibility of missing an ectopic thyroid that could be malignant.

**Conflicts of Interest**

The authors declare that there is no conflict of interest regarding the publication of this paper.

**References**

[1] M. Chawla, R. Kumar, and A. Malhotra, "Dual ectopic thyroid: case series and review of the literature," *Clinical Nuclear Medicine*, vol. 32, no. 1, pp. 1–5, 2007.

[2] A. Soscia and G. Guerra, “Parapharyngeal ectopic thyroid: the possible persistence of the lateral thyroid anlage. Clinical case report,” *Surgical and Radiologic Anatomy*, vol. 26, no. 4, pp. 338–343, 2004.

[3] J. S. Yoon, K. C. Won, I. H. Cho, J. T. Lee, and H. W. Lee, “Clinical characteristics of ectopic thyroid in Korea,” *Thyroid*, vol. 17, no. 11, pp. 1117–1121, 2007.

[4] T. P. Caccetta, A. Kumar, and H. Ishak, “Lateral aberrant thyroid tissue presenting as a lateral neck mass,” *ANZ Journal of Surgery*, vol. 75, no. 12, pp. 1123-1124, 2005.

[5] D. A. Zander and W. R. K. Smoker, “Imaging of ectopic thyroid tissue and thyroglossal duct cysts,” *Radiographics*, vol. 34, no. 1, pp. 37–50, 2014.

[6] J. K. Hoang, J. E. Langer, W. D. Middleton et al., “Managing incidental thyroid nodules detected on imaging: white paper of the ACR incidental thyroid findings committee,” *Journal of the American College of Radiology*, vol. 12, no. 2, pp. 143–150, 2015.

[7] B. R. Haugen, E. K. Alexander, K. C. Bible et al., “2015 American Thyroid Association management guidelines for adult patients with thyroid nodules and differentiated thyroid cancer: the American Thyroid Association guidelines task force on thyroid nodules and differentiated thyroid cancer,” *Thyroid*, vol. 26, no. 1, pp. 1–133, 2016.

[8] N. Ibrahim and I. Fadeyibi, "Ectopic thyroid: etiology, pathology and management," *Hormones*, vol. 10, no. 4, pp. 261–269, 2011.

[9] H. Prado, A. Prado, and B. Castillo, "Lateral ectopic thyroid: a case diagnosed preoperatively," *Ear, Nose, & Throat Journal*, vol. 91, no. 4, pp. E14–E18, 2012.

[10] M. R. Pelizzo, A. Toniato, and G. Gemo, "Zuckerkandl’s tuberculum: an arrow pointing to the recurrent laryngeal nerve (constant anatomical landmark)," *Journal of the American College of Surgeons*, vol. 187, no. 3, pp. 333–336, 1998.

[11] G. Guerra, M. Cinelli, M. Mesolella et al., "Morphological, diagnostic and surgical features of ectopic thyroid gland: a review of literature," *International Journal of Surgery*, vol. 12, pp. S3–S11, 2014.
[12] A. Agarwal, A. K. Mishra, C. P. Lombardi, and M. Raffaelli, "Applied embryology of the thyroid and parathyroid glands. Surgery of the Thyroid and Parathyroid Gland, G. W. Randolph, Ed., pp. 51–78, Saunders, Philadelphia, PA, 2nd edition, 2013.

[13] M. De Felice and R. Di Lauro, "Thyroid development and its disorders: genetics and molecular mechanisms," Endocrine Reviews, vol. 25, no. 5, pp. 722–746, 2004.

[14] W. R. Sackett, T. S. Reeve, B. Barraclough, and L. Delbridge, "Thyrothymic thyroid rests: incidence and relationship to the thyroid gland," Journal of the American College of Surgeons, vol. 195, no. 5, pp. 635–640, 2002.

[15] R. C. Clay and S. S. Blackman, "Lateral aberrant thyroid: metastasis to THE lymph nodes from primary carcinoma of the thyroid gland," Archives of Surgery, vol. 48, no. 3, pp. 223–228, 1944.

[16] M. Rahalkar, A. Rahalkar, and S. Solav, "A rare case of triple thyroid ectopia," Indian Journal of Endocrinology and Metabolism, vol. 18, no. 2, pp. 238–240, 2014.

[17] H. J. Kwon, S. M. Jin, S. H. Lee, and D. H. Kim, "Adenomatous hyperplasia arising from dual ectopic thyroid," Clinical and Experimental Otorhinolaryngology, vol. 2, no. 3, pp. 155–158, 2009.

[18] B. K. Choudhury, U. K. Saikia, D. Sarma et al., "Dual ectopic thyroid with normally located thyroid: a case report," Journal of Thyroid Research, vol. 2011, Article ID 159703, 4 pages, 2011.

[19] S. R. Konde, H. Singh, A. Pawar, and A. Sasane, "Triple ectopic thyroid," Medical Journal Armed Forces India, vol. 68, no. 2, pp. 173–175, 2012.

[20] K. Oikawa and R. Kuramoto, "Triple ectopic thyroid: a case report and review of literature," Journal of Thyroid Disorders & Therapy, vol. 02, no. 02, article 1000126, 2013.

[21] R. Kumar, S. Khullar, R. Gupta, A. Marwah, and M. A. Drm, "Dual thyroid ectopy: case report and review of the literature," Clinical Nuclear Medicine, vol. 25, no. 4, pp. 253–254, 2000.

[22] A. Sood, R. K. Seam, M. Gupta, D. Raj Sharma, and P. Bhardwaj, "Dual ectopic thyroid: a case report with review of literature," Iranian Journal of Radiology: a quarterly journal published by the Iranian Radiological Society, vol. 8, no. 1, pp. 29–32, 2011.

[23] S. H. Baik, J. H. Choi, and H. M. Lee, "Dual ectopic thyroid," European Archives of Oto-Rhino-Laryngology, vol. 259, no. 2, pp. 105–107, 2002.

[24] N. Hod, L. Mindlin, M. Cohenpour, and T. Horne, "Double ectopic thyroid," Pediatric Radiology, vol. 32, no. 12, pp. 859–861, 2002.

[25] N. Bhattacharyya and M. P. Fried, "Assessment of the morbidity and complications of total thyroidectomy," Archives of Otolaryngology–Head & Neck Surgery, vol. 128, no. 4, pp. 389–392, 2002.