



Tomas Joaquin C. Mendez, MD
Cecilia Gretchen S. Navarro-Loecin, MD

Department of Otolaryngology
Head and Neck Surgery
St. Luke's Medical Center, Quezon City

Double Ectopic Thyroid Gland in a 10-Year-Old Filipino Boy

ABSTRACT

Objective: To present a case of a double ectopic thyroid gland in a 10-year-old boy and discuss the pros and cons of the different management options that were available.

Methods:

Design: Case Report
Setting: Tertiary Private Hospital
Patient: One

Results: A 10-year-old boy presented with hoarseness and easy fatigability for 6 years. Rigid endoscopy and CT scan showed an infraglottic mass originating from the anterior tracheal wall causing obstruction. Biopsy revealed thyroid tissue with atypia. Thyroid scintigraphy showed uptake in the submental and midline anterior neck. Thyroid hormone levels were consistent with hypothyroidism. Levothyroxine returned hormone levels to normal and resulted in complete regression of the mass with no symptoms of dyspnea, stridor or bleeding.

Conclusion: The management of ectopic thyroid presents a challenge as there are no guidelines for optimal treatment. Thyroid hormone insufficiency is a frequent occurrence and emphasis must be given to its monitoring. Surgery in a critical airway lesion such as this may be reserved for cases where the patient experiences dyspnea and stridor or lack of response to thyroid hormone treatment.

Keywords: *Ectopic thyroid, direct laryngoscopy, thyroid hormone, levothyroxine*

Ectopic thyroid results from an aberrancy in the normal migration pathway of the thyroid gland due to an arrest in the descent and/or an interruption in the pathway causing maturation and development in other locations other than the true final anatomic position.¹ Although a lingual thyroid is the most frequent location of ectopic thyroid tissue, other locations such as the sublingual region, tracheal, submandibular, lateral neck, palatine tonsils, and axilla also exist,² and ectopic thyroid tissue in distant sites such as the ovary and GI tract have also been reported.² Intratracheal thyroid tissue represents only 7% of all intraluminal tracheal masses.³ Dual ectopic thyroid is extremely rare especially when no thyroid gland is seen in the normal anatomic position with very few cases reported in the literature.⁴ We report one such case.

CASE REPORT

Our patient presented at birth with a weak, breathy cry and an incidental finding of a submental mass. He was generally stable with no episodes of cyanosis and no need for intubation or admission into intensive care. As the boy grew older, his hoarseness persisted, described as breathy in quality with associated easy fatigability.

Correspondence: Dr. Cecilia Gretchen S. Navarro-Loecin
Department of Otolaryngology, Head and Neck Surgery
2nd Floor, St. Luke's Medical Center, Quezon City
Eulogio Rodriguez Sr., Avenue, Quezon City 1112
Philippines
Phone: (632) 723-0101 local 6530
Email: slmcenthns@gmail.com

The authors declare that this represents original material that is not being considered for publication or has not been published or accepted for publication elsewhere in full or in part, in print or electronic media; that the requirements for authorship have been met by the authors, and that the authors believe that the manuscript represents honest work.

Disclosures: The authors signed a disclosure that there are no financial or other (including personal) relationships, intellectual passion, political or religious beliefs, and institutional affiliations that might lead to a conflict of interest.

Presented at the Philippine Society of Otolaryngology Head and Neck Surgery Interesting Case Contest (2nd Place). June 30, 2016. JY Campos Hall B Unilab Bayanihan Center, Pasig City.



Creative Commons (CC BY-NC-ND 4.0)
Attribution - NonCommercial - NoDerivatives 4.0 International

Philipp J Otolaryngol Head Neck Surg 2018; 33 (1): 47-50

© Philippine Society of Otolaryngology – Head and Neck Surgery, Inc.

Four years prior to consult, an ear, nose and throat (ENT) specialist evaluated the submental mass and hoarseness, discovering a reddish, vascular, infraglottic mass on flexible nasopharyngolaryngoscopy. Partial excision of the submental mass yielded histopathologic results consistent with nodular colloid goiter. The mother was advised that thyroid hormone levels and further biopsy of the infraglottic mass were needed but they did not follow-up. There was no interim progression of symptoms.

Due to persistence of hoarseness, a second opinion was sought four months before consult and flexible nasopharyngolaryngoscopy still showed an infraglottic mass. Non-contrast neck computed tomography (CT) scan showed a 0.8 x 0.8 cm homogeneously enhancing nodule in the anterior commissure of the vocal folds and infraglottic area originating from the anterior tracheal wall, extending downwards and causing

infraglottic obstruction, as well as another focus in the region of the floor of the mouth, anterior to the hyoid bone at the level of the anterior belly of the digastric muscle. There was no visible thyroid gland at the level of the thoracic inlet. (Figure 1 A, B) The mother was advised excision of the infraglottic mass due to impending upper airway obstruction.

The boy was brought to us for a third opinion by now complaining of easy fatigability, hoarseness and difficulty catching up in school. On examination, there was no palpable thyroid gland in the anterior neck, but there was a 3 cm submental surgical scar. Rigid endoscopy showed a reddish brown vascular mass originating from the anterior tracheal wall occupying around 2/3 of the trachea at the infraglottic level with a posterior airway patency of around 20-25%. There was good bilateral vocal fold movement but the mass prevented apposition of the vocal folds. (Figure 2 A, B) An ultrasonogram to confirm CT scan

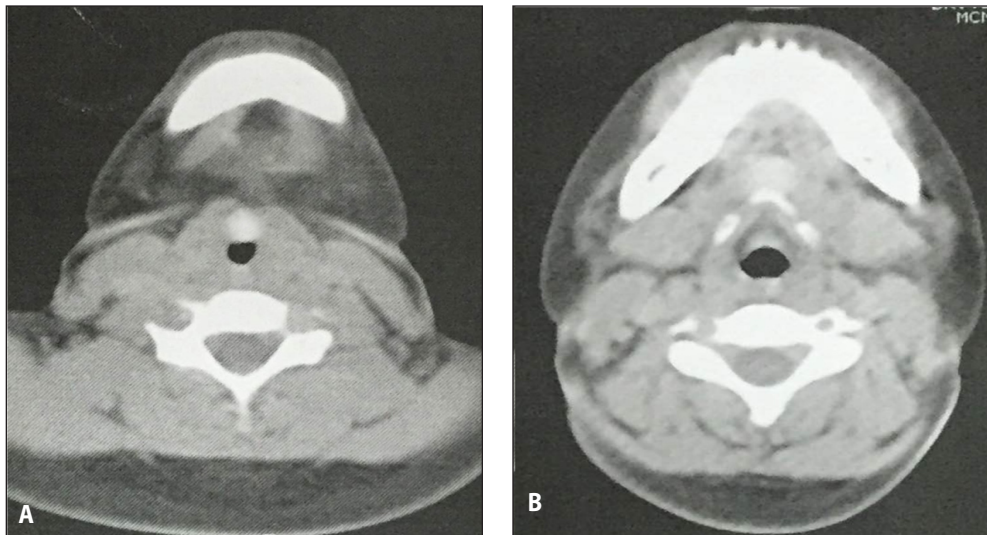


Figure 1. Non-contrast Neck CT Scan prior to hormonal therapy, **A.** Representative axial cut at the subglottic level showing homogeneously enhancing nodule; and **B.** Representative axial cut at the level of the hyoid bone showing remnant thyroid tissue anterior to the hyoid body.

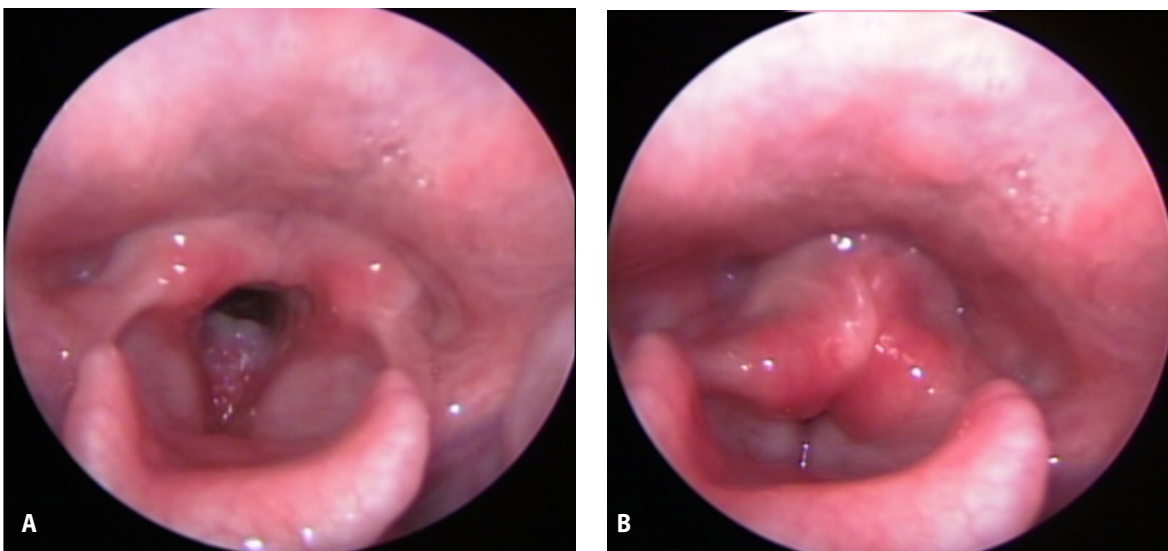


Figure 2. Flexible nasopharyngolaryngoscopy **A.** Abduction, showing the infraglottic mass occupying around 75% of the tracheal lumen; and **B.** Compensatory adduction of the false vocal folds on phonation.

findings revealed no thyroid gland in the normal anatomic location. Thyroid function tests were consistent with hypothyroidism. Iodine-131 thyroid scintigraphy showed two foci of increased tracer uptake, arranged in vertical configuration in the anterior neck. The superior 2.1 x 2.1 cm focus appearing submental in location on lateral view was suggestive of ectopic thyroid tissue. The inferior 2.3 x 1.6 cm focus in the midline anterior neck most likely represented functioning thyroid tissue. (Figure 3) Direct laryngoscopy punch biopsies of the infraglottic

mass in multiple quadrants yielded histopathologic results consistent with thyroid tissue with atypia after staining with PAX-8. (Figures 4 A, B) Hormone replacement was initiated with levothyroxine 100mcg/day with no episodes of stridor, dyspnea or upper airway bleed. The voice improved in four months with good phonation and no apparent hoarseness or easy fatigability. The infraglottic mass regressed completely. (Figure 5)

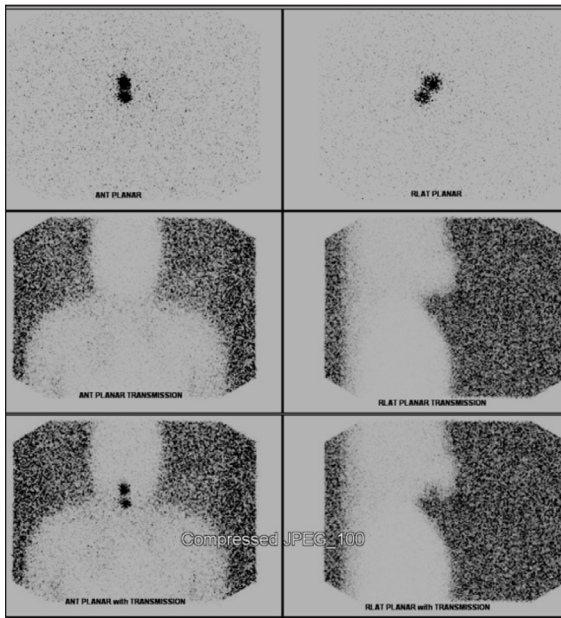


Figure 3. Thyroid scintigraphy scan (I-131) prior to hormonal therapy showing the two foci of avid radiotracer uptake.

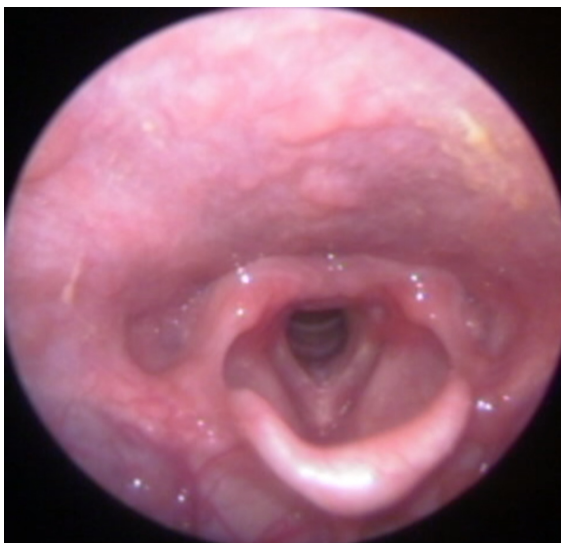


Figure 5. Flexible nasopharyngolaryngoscopy showing complete regression of the infraglottic mass after normalization of thyroid hormones.

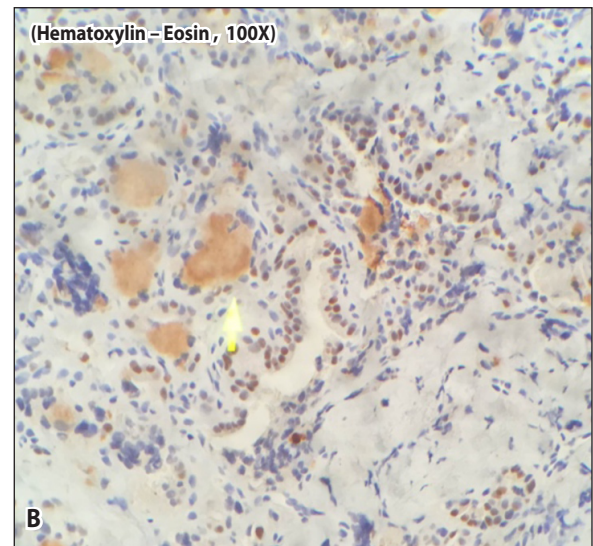
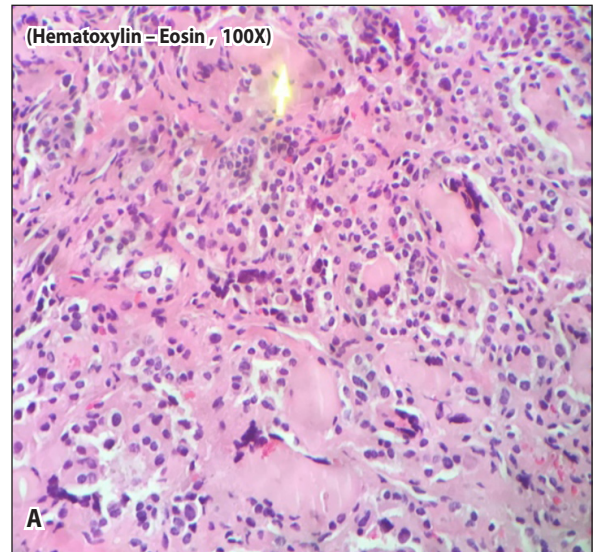


Figure 4. A. Histopathologic slide (Hematoxylin - Eosin), low power view (100x), infraglottic biopsy. The arrow points to eosinophilic staining of colloid material seen in the tissue sample; **B.** PAX-8 tissue stain confirming the presence of thyroid tissue. The arrow points to colloid material with avid stain uptake.

DISCUSSION

The thyroid gland is the first endocrine organ to develop in the human body, beginning 3-4 weeks AOG.² Dual ectopic thyroid development occurs during the descent and formation of the thyroid gland, which according to two predominating theories of development allow the implantation or invasion of thyroid tissue in other locations.⁵

The prevalence of ectopic thyroid in other countries ranges from 1/100,000 to 1/300,000 births being more common in females of Asian origin.⁶ Dual ectopic thyroid tissue is even rarer especially with no orthotopic thyroid gland.⁴ Studies also suggest that up to 11% of intratracheal thyroidal tissue may undergo malignant transformation if left untreated most commonly papillary thyroid carcinoma.⁴

Our patient presented with submental and infraglottic double ectopic thyroid glands. Infraglottic intraluminal involvement may be explained by two theories of development. According to the malformation theory of Zierussen,⁵ developing tracheal cartilage splits the thyroid gland, allowing intraluminal development of an ectopic rest of thyroid tissue. In the ingrowth theory of Paltauf, thyroid tissue develops intratracheally through direct invasion of thyroid tissue through the trachea.⁵ Our case is interesting because our patient had an intratracheal component that did not appear to originate from any normal thyroid tissue. Moreover, he presented unusually with hoarseness and easy fatigability only, as patients with upper airway obstruction often have biphasic stridor, dyspnea and cough.⁶

Migration control as well as normal thyroid development and differentiation are controlled by gene expression. Control and regulation of thyroid migration are a function of the *FOXE1* gene.² Further studies are needed to elucidate the relationship of *FOXE1* allele mutations and thyroid ectopy⁷ including studies of other genes such as *Titf1/Nkx2-1*, *PAX8* and *Hhex* that are involved in intrinsic thyroid development and maturation.⁸

The most important diagnostic tests include a neck ultrasonogram and thyroid scan preferably a Technetium 99m pertechnetate scan.⁶ The thyroid scan provides valuable information on the presence of a normal anatomic thyroid gland and offers more information to differentiate a suspicious ectopic thyroid mass from other differential diagnoses.⁶ A CT or MRI are helpful for pre-operative evaluation since these evaluate presence of cartilage invasion and/or position of the intratracheal lesion.⁹ Flexible endoscopy should be done in cases of airway involvement such as with our patient. Unlike our patient, intratracheal thyroid tissue commonly presents as a reddish-brown submucosal mass in the posterolateral trachea.⁶

The management of ectopic thyroid with or without an orthotopic thyroid gland is a challenge as there are currently no established guidelines for treatment.⁶ Hypothyroidism further complicates treatment. The treatment strategy centers on the patient's presentation and symptoms depends on the location of the ectopic thyroid and its biochemical characteristics, age and the qualitative characteristics of

the mass itself.⁶ Some studies support the efficacy of TSH suppression alone for asymptomatic/mild obstructive symptom patients with or without thyroid hormone derangements, as TSH suppression alone is proven to significantly decrease the size of the ectopic thyroid masses and stabilize the thyroid hormones.² TSH suppression also decreases the risk for malignant transformation and prevents further growth of the mass.² Surgical excision is recommended for masses with severe obstructive symptoms, bleeding, ulceration, degeneration, or biopsy proven malignancy.⁶ An infraglottic thyroid, as found in our patient, may be removed using an open cricoid approach, CO₂ laser, or harmonic scalpel if there are severe obstructive symptoms.² Radioactive iodine is reserved for those who are unstable to undergo surgery.⁶

Management revolves around the patient as a whole in cases like these. Children are especially sensitive to the effects of hypothyroidism and the importance of thyroid hormone stabilization cannot be overemphasized. Surgery may be reserved for critical airway compromise, and watchful waiting with close follow up is currently the best option.

REFERENCES

1. Singh GB, Kumar D, Ranjan S, Tomer S. A rare case of double ectopic thyroid without orthotopic thyroid gland. *Int J Pediatr Otorhinolaryngol Extra*. 2015 Mar; 10(2): 28-30. DOI: <http://dx.doi.org/10.1016/j.pedex.2015.01.003>.
2. Ibrahim NA, Fadeyibi IO. Ectopic thyroid: etiology, pathology and management. *Hormones (Athens)*. 2011 Oct-Dec; 10(4): 261-269. PubMed PMID: 22281882.
3. Oliver VJ, Rico RR, Morillo AD, Ruiz EF, Ruiz JM, Arcos JAP, Baro GR. Tejido tiroideo ectopico intralaringeo. Presentacion de un caso clinico y revision de la literatura. *Acta Otorrinolaringologica*. 2001; (53): 54-59.
4. Kumar Choudhury B, Kaimal Saikia, U, Sarma D, Saikia M, Dutta Choudhury S, Barua S, Dewri S. Dual ectopic thyroid with normally located thyroid: a case report. *J Thyroid Res*. 2011; 2011: 159703. DOI: 10.4061/2011/159703; PubMed PMID: 21765986; PubMed Central PMCID: PMC3134180.
5. Ramalingam KK, Ramalingam R, Dhote K, Murthy S. Ectopic thyroid: a rare cause of tracheal obstruction. *Indian J Otolaryngol Head Neck Surg*. 2005 Jul; 57(3): 252-5. DOI: 10.1007/BF03008026; PubMed PMID: 23120184 PMCID: PMC3451353.
6. Nousseios G, Anagnostis P, Goulis DG, Lappas D, Natsis K. Ectopic thyroid tissue: anatomical, clinical, and surgical implications of a rare entity. *Eur J Endocrinol*. 2011 Sep; 165(3): 375-382. DOI: 10.1530/EJE-11-0461; PubMed PMID: 21715415.
7. Van Vliet G, Deladoey J. Sublingual thyroid ectopy: similarities and differences with Kallmann syndrome. *F1000 Prime Rep*. 2015 Feb 3; 7:20. DOI: 10.12703/P7-20; PubMed PMID: 25750738; PubMed Central PMCID: PMC4335790.
8. De Felice M, Di Lauro R. Thyroid development and its disorders: genetics and molecular mechanisms. *Endocr Rev*. 2004 Oct; 25(5): 722-746. DOI: 10.1210/er.2003-0028; PubMed PMID: 15466939.
9. Zander DA, Smoker WR. Imaging of ectopic thyroid tissue and thyroglossal duct cysts. *Radiographics*. 2014 Jan-Feb; 34(1): 37-50. DOI: 10.1148/rg.341135055; PMID: 24428281.