Ectopic Intratracheal Thyroid and Parathyroid Tissue Presenting with Airway Obstruction

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INTRODUCTION

Ectopic thyroid tissue is a rare phenomenon, resulting from errors in embryogenesis of the thyroid gland. It can occur at any point along the gland’s migratory pathway from the foramen cecum to its final pretracheal position. The prevalence of ectopic thyroid is roughly 1 in 10,000, though this is felt to be an underestimate, as it is often asymptomatic and found only upon autopsy. Ectopic thyroid tissue is most commonly found in the sublingual, sublingual, thyroglossal, laryngotracheal, and lateral neck sites.

Intratracheal thyroid tissue is more commonly related to invasive thyroid carcinoma, so EITT is easily misdiagnosed as tracheal invasion. We present a case in which an intratracheal mass was initially felt to be secondary to invasive thyroid carcinoma and ultimately found to be EITT. This is the first reported case demonstrating both intratracheal thyroid and parathyroid tissue.

CASE REPORT

A 56-year-old man presented to an outside pulmonologist for evaluation of respiratory difficulty and was found to have a tracheal mass. CT neck and chest demonstrated an intratracheal mass that appeared to arise off the left posteroatorial wall with significant airway narrowing (Figures 1A and B). An orthotopic thyroid gland was noted. He underwent endobronchial ultrasound-assisted biopsy of the mass, which was consistent with thyroid tissue. Immediately following this biopsy, the patient developed stridor and required intubation.

He was thought to have thyroid carcinoma with tracheal invasion and was scheduled for a total thyroidectomy and tracheal resection on 5/30/13. Intraoperatively, no communication was found between the thyroid gland and the trachea and all frozen sections demonstrated benign thyroid tissue. Therefore, the total thyroidectomy was aborted until final pathology could be analyzed. Final pathology was consistent with an orthotopic thyroid gland containing an adenomatoid nodule and benign thyroid tissue from the tracheal biopsies.

Direct laryngoscopy demonstrated near-total obstruction of the trachea just distal to the vocal folds, due to a submucosal mass arising mostly from the left tracheal wall. The patient subsequently underwent several procedures to address this mass. On 7/8/13, the stent was removed and direct laryngoscopy was performed, with excision of a small amount of granulation tissue. On 8/26/13, we performed direct laryngoscopy with CO2 laser ablation and balloon dilation of the tracheal mass, which extended from just inferior to the vocal folds to just proximal to the tracheotomy. The patient was decannulated in the office on 9/26/13. The final procedure, direct laryngoscopy with repeat CO2 laser ablation and balloon dilation, was performed on 10/9/13. This procedure addressed an area of residual bulk along the anterior tracheal wall just proximal to the prior tracheotomy site. (See Figures 2-4 for office and intraoperative views of trachea throughout treatment course).

Final pathology following CO2 laser excision of the intratracheal mass was consistent with benign thyroid and parathyroid tissue (Figures 5A and B). At his most recent follow-up, four months after his final procedure, the patient denied dyspnea or dysphagia and is back to work with full duties. Both vocal folds remain fully mobile with excellent voice.

DISCUSSION

Ectopic intratracheal thyroid tissue (EITT) represents from 1% to 6.7% of all endotracheal tumors. The first case was described by Ziemsen in 1875. Since then, there have been fewer than 140 cases reported in the literature. EITT most commonly presents in women (3:1 ratio) between the ages of 30 and 50. It is most commonly seen in the endodideto regions, as well as in central Europe. Roughly 75% of EITT cases are associated with orthotopic thyroid glands.5,7

Two theories exist regarding how EITT arises. The first suggests that the developing thyroid gland is divided by the developing trachea and its cartilaginous rings. An alternative theory is that there is an ingrowth of thyroid tissue into the developing tracheal lumen.8

Most often, EITT presents as a smooth, broad-based submucosal mass extending from the lateral or posteroatorial tracheal wall into the lumen. It is most commonly seen in the subglottis and upper trachea, but may arise anywhere from the glottis to the carina. Hemorrhage, ulceration, and multiple nodules are more indicative of carcinoma and are typically not seen in EITT.7

Symptomatically, EITT usually presents as gradually progressive dyspnea, cough, and stridor. It is frequently mistaken for asthma, with only partial response to therapy. The airway obstruction can progress to life-threatening levels. It may present following removal of the orthotopic thyroid gland, with resultant physiologic hyperfunction of the ectopic tissue. Hormonal changes associated with menses and pregnancy have also been noted to worsen the associated dyspnea due to alternating hyperplasia and involution. Patients are typically euthyroid, due to a normally functioning orthotopic thyroid.8

CT and MRI may help in delineating the mass and identifying any connection between the intratracheal mass and the orthotrophic thyroid, which may be more indicative of invasive thyroid carcinoma. However, several cases have noted an appearance consistent with invasive thyroid carcinoma on imaging for EITT.2,10,11

Treatment of choice is excision, which can be achieved with endoscopic and laser debulking, with ablative medical thyroid hormone suppression. Alternatives include radioactive ablation and thyroid hormone suppression.1 There has been a range of reported surgical approaches. Most authors have had success with endoscopic laser ablation. This approach is associated with careful dissection of the mass away from the underlying tracheal mucus.5,7 Still others report performing tracheal resection with end-to-end anastomosis or staged tracheal wall reconstruction.12 Our patient was ultimately successfully treated with endoscopic laser debulking.

Our patient’s pathology demonstrated the first reported case of intratracheal ectopic parathyroid tissue. During embryogenesis, the third pair of pharyngeal pouches differentiates into the inferior parathyroid glands and the thyrothymic tract, which, while the fourth pair develops into the superior parathyroid glands and the lateral anlage of the thyroid gland. Errors in descent may yield ectopic parathyroid glands. Ectopic parathyroid glands have been reported in 4–16% of patients. Most frequently, these are found in the mediastinum, thyrohyoid ligament, or within the thyroid. Less often, they are noted in the aortopulmonary window, pericardium, muscosa of the pyriform sinus, within the carotid sheath, or in the posterior cervical triangle.2,11 They have not previously been reported within the trachea.

CONCLUSIONS

Ectopic intratracheal thyroid tissue is a rare entity. This is the first case reported of both intratracheal thyroid and parathyroid tissue. The treatment of choice is surgical excision. Endoscopic laser debulking is a successful method of treatment in cases of airway obstruction. EITT is an important diagnosis to consider, as it is easily mistaken for asthma or invasive thyroid carcinoma.

REFERENCES


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