Intratracheal ectopic thyroid: Case report and review

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Abstract

Intratracheal ectopic thyroid tissue is a rare abnormality that can cause airway obstruction. The symptoms can easily be confused with those of bronchial asthma. We describe the case of a 40-year-old man with subglottic thyroid tissue and multinodular goiter who had been misdiagnosed earlier with bronchial asthma. After the correct diagnosis was established, the lesion was excised via an external approach. We also discuss the clinical features and management of intratracheal thyroid tissue.

Introduction

Ectopic thyroid tissue can be located in the midline at any site from the base of the tongue to the mediastinum. The trachea is one of the rarest locations. When intratracheal thyroid does occur, the most common location is the subglottic space. The most common manifestation is airway obstruction. The first sign may be a wheeze, which can be easily mistaken for bronchial asthma. The rarity of this entity makes the diagnosis even more difficult. We describe a case of intratracheal ectopic thyroid tissue in a man who had indeed been misdiagnosed with asthma.

Case report

A 40-year-old man came to our institution’s bronchopneumonology unit with a 1-month history of gradually worsening shortness of breath and exertional dyspnea. He was diagnosed with bronchial asthma and prescribed bronchodilator treatment, but he failed to respond. As his symptoms worsened, the diagnosis came into doubt, and further testing was performed.

Flexible bronchoscopy detected a subglottic mass attached to the left side of the tracheal wall. On magnetic resonance imaging (MRI), the mass appeared to be a multinodular and heterogenous lesion; it originated on the left posterior side of the trachea at the level of the first and second tracheal cartilages (figure 1, A). The patient was referred to the ENT department, where he reiterated that his shortness of breath became worse with exercise. He had no history of lung disease. He had undergone a left lobe thyroidectomy for multinodular goiter 21 years earlier, but he had not undergone any postoperative follow-up. He had no history of heart disease.

Clinically, the patient exhibited stridor on inspiration and expiration. Videolaryngoscopy revealed a paralytic left vocal fold. The mass could be seen under the fold (figure 1, B). It was multinodular and covered with normal-appearing mucosa. The diameter of the tracheal passage was approximately 3 mm at the level of the mass. The patient also had a palpable mass on the right side of the neck that felt like an enlarged thyroid gland. This mass, which had also been demonstrated on MRI, appeared to be a multinodular enlargement of the right lobe of the thyroid gland (figure 1, C). Finally, a multinodular remnant of the left lobe could be seen in the left tracheoesophageal groove. A radionuclide thyroid scan detected multinodular hyperplasia with normoactive nodules on the right side and the remnant thyroid tissue on the left side. The lesion did not uptake any radionuclide substance. The patient was scheduled for surgery. Preoperative blood biochemistry revealed a deep hypothyroidism, as the patient’s thyroid-stimulating hormone (TSH) level exceeded 100 μU/ml.

Tracheotomy with local anesthesia was performed between the third and fourth tracheal rings. Next, the patient underwent direct rigid laryngobronchoscopy under general anesthesia. Biopsies were taken from the overlying mucosa and from the intratracheal mass. The lesion was fragile, and it bled easily. Findings on histopathologic evaluation of the biopsied material were consistent with thyroid tissue. The pathologist suggested an excisional biopsy of the tumor. Thyroid hormone replacement therapy was started, and the patient’s TSH level fell.

We chose an open approach for total resection of the mass. The trachea and thyroid cartilage were exposed, and the tracheal lumen was entered by anterior splitting of the first three tracheal rings. The mass, which was attached to the left posterolateral tracheal wall, was detached.
from the first and second tracheal cartilages along with the overlying mucosa. Profuse bleeding necessitated the transfusion of 1 unit of blood. The mucosal defect was closed with a split-thickness skin graft. A stent was made from the finger of a surgical glove and filled with sponge. The stent was placed in the lumen and fixed to the skin with sutures and buttons.

The patient’s postoperative course was not complicated, and the stent was removed after 10 days under endoscopic guidance. Histopathologic examination of the resected mass revealed normal thyroid tissue, and the definitive diagnosis was ectopic thyroid tissue in the trachea (figure 2). A total thyroidectomy was performed 4 months later. No signs of recurrence were evident at the 1-year follow-up.

Discussion
The first reported case of intratracheal ectopic thyroid tissue was published by Ziemssen in 1875. Since then, more than 100 cases have been reported. More than 90% of these cases have occurred in Central European countries. Intratracheal thyroid tissue accounts for 7% of all intratracheal tumors. Two-thirds of these cases occur in women.

Etiology. The mechanism for the development of ectopic intratracheal thyroid tissue is not known for certain. There are two long-standing theories. Von Bruns proposed that the thyroid gland becomes divided by the growing tracheal cartilages and that a portion of the thyroid tissue is left inside the trachea between the cartilage and the tracheal mucosa. This view was supported by Falk, who studied sections of 21 larynges obtained from newborns and premature infants and found thyroid tissue in the trachea in 9 of them (42.9%). The other theory was suggested by Paltauf, who speculated that thyroid tissue may penetrate the tracheal cartilage and who demonstrated a connection between the thyroid gland and thyroid tissue in the trachea. These two theories, which were both proposed at the end of the 19th century, are still being debated.

Ectopic intratracheal thyroid tissue may be more common than what has been reported because it is possible that most patients remain asymptomatic until a change in their endocrine status occurs. The tissue probably remains silent until an increase in TSH level occurs. A high TSH level stimulates both the thyroid and the intratracheal thyroid tissue. Our patient was hypothyroid. The multinodular
goiter in his right lobe, the hypertrophied remnant of the left lobe, and the intratracheal thyroid might all have been consequences of his elevated TSH level. If a patient with ectopic intratracheal thyroid tissue maintains a normal TSH level, the thyroid may remain asymptomatic. In fact, an autopsy study by Ferlito et al showed that even large and multiple plaques of intratracheal ectopic tissue may remain asymptomatic. However, this is not the case for all patients. Several authors have reported obstruction of the airway by intratracheal thyroid tissue in patients with a normal TSH level and a normal thyroid gland. Still, Waggoner reported that 74% of cases of intratracheal thyroid tissue were associated with goiters. Moreover, Central Europe, where most cases of intratracheal thyroid tissue have been reported, is an area where goiter is endemic. Finally, Donegan and Wood proposed that ectopic intratracheal thyroid tissue may be hereditary.

**Diagnosis.** Diagnosis may pose a problem. The physician must have a high degree of suspicion. As mentioned, many patients with ectopic intratracheal thyroid tissue, including ours, are misdiagnosed with bronchial asthma and treated accordingly. Bronchoscopy is the diagnostic method of choice. If the lesion is smooth and covered with normal-appearing mucosa, it is safe to perform a biopsy. However, care should be taken during a biopsy because ectopic thyroid tissue may bleed profusely. In fact, Randolph et al advised against performing biopsies for just this reason. Without a biopsy, however, a diagnosis is extremely difficult to establish.

One option is to assume the tumor is benign and perform a complete excision. But the possibility that the lesion is malignant makes this option less than optimal; intratracheal thyroid tissue has been reported to become malignant in 11% of cases. Therefore, we feel that the best course is to obtain a biopsy with appropriate precautions. One such precaution is to perform a tracheotomy beforehand. This step prevents aspiration of the bleeding caused by the biopsy, and it prevents the complete airway obstruction that can occur as a result of postoperative edema. Thoren reported the case of a patient who died as a result of reactive swelling following laryngoscopy.

Computed tomography (CT) and MRI are valuable diagnostic tools. A finding of a clear separation of the intratracheal thyroid tissue and the thyroid gland itself makes a diagnosis of ectopic intratracheal thyroid tissue more likely. CT and MRI can also detect additional pathologies, such as multinodular goiter. Radioactive iodine scanning is not very useful for diagnosing intratracheal thyroid tissue because the uptake by the thyroid gland masks the uptake of the intratracheal thyroid tissue. A radioactive iodine scan may be helpful in determining if the thyroid gland is functioning and if any other ectopic thyroid tissue is present.

**Management.** Surgery is the mainstay of treatment for intratracheal thyroid tissue. Chanin and Greenberg described the case of an infant with intratracheal thyroid tissue who was treated with thyroid-suppression therapy (levothyroxine 0.05 mg/day) for 10 years. However, because this patient was a newborn, the airway enlarged during therapy because the ectopic intratracheal thyroid tissue did not. Such is not the case with an adult, of course. Most reported ectopic intratracheal thyroid tissues were removed through a laryngotraceal fissure. We preferred this approach in our patient. Because this type of approach may result in airway stenosis, we exercised extreme care in making sure that we did not damage the tracheal cartilages. Afterward, we placed a skin graft over the mucosal defect. An allograft can be used instead of a skin graft. For our patient, we needed only a very small skin graft. With this technique, donor-site morbidity is minimal. Also, using a skin graft is more economical than using an allograft. We also placed a stent for 10 days. Using a stent is important for preventing stenosis.

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