INTRALARYNGOTRACHEAL GOITER:
A CASE PRESENTATION AND REVIEW OF THE LITERATURE

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ABSTRACT

Thyroid tissue located within the upper airway has received only sparse attention in the English literature. Aberrant benign thyroid masses may present as either an unrelated autopsy finding or with symptoms such as dyspnea, hemoptysis or adult-onset asthma. There is no place for medical management in the therapy of these lesions, although the appropriate surgical procedure is not clearly established.

In this article a patient with respiratory distress and subglottic goiter is presented. She had subtotal thyroidectomy performed for goiter about 10 years ago. She was managed with tracheotomy and submucosal resection of subglottic thyroid tissue, followed by iodine ablation therapy and thyroid hormone replacement.


Keywords: Subglottic goiter, Laryngotraheal goiter, Ectopic thyroid tissue.

INTRODUCTION

Ectopic thyroid masses may be encountered in four locations: the lingual area, sublingual (supra- and infrahyoid) area, thyroglossal duct, and the intralaryngotraheal area. Ectopic thyroid tissue has also rarely been discovered within the pericardium, esophagus, and as caudal as the diaphragm. Thyroid tissue that is located within the upper airway (lumen of trachea) however, has received only sparse attention in the English-language literature. In the commonly used textbooks of medicine, pediatrics and surgery, little mention is made of intratracheal goiter. Its rarity often results in delayed recognition, and this can pose challenging diagnostic and management problems.

CASE REPORT

A twenty-five year old woman presented with progressive dyspnea from 1 month ago. She had respiratory distress with significant limitation of activity, occasional stridor, and spasmodic cough, but no history of hemoptysis. At the age of fifteen she had right total and left subtotal thyroidectomy for huge colloid goiter which had developed gradually within seven years. For recurrence of the anterior neck mass she had a second operation performed 3 years ago. The pathologic results of both operations were colloid goiter but she did not receive any thyroid hormone replacement.

On physical examination she had a scar of two collar incisions in the neck with a soft mass measuring about 3 cm on the left side of the trachea without any cervical lymphadenopathy. Cervical X-ray revealed pressure and deviation of the trachea (Fig. 1). CT scan of the neck revealed an extrinsic and intrinsic mass in the laryngotraheal region which was enhanced with contrast. The intrinsic mass had a smooth border and had narrowed more than 85 percent of the tracheal lumen (Fig. 2-a,b). Bronchoscopy showed a smooth submucosal mass in the left posterolateral subglottic area with intact mucosa. Cervical exploration was done through a lower previous collar incision on Jan 10, 1997.

The extrinsic mass which was a remnant of the left upper lobe of the thyroid was excised. There was no intraluminal connection or invasion of the thyroid cartilage. Tracheotomy was done through a midline incision on tracheal rings 1-4. Submucosal resection of the subglottic mass was per-

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formed by curettage. The bleeding was controlled by topical epinephrine application and tamponade. The tracheal opening was closed and supported by the right strap muscle. For assurance of complete therapy, she was treated by radioiodine ablation therapy and thyroid hormone replacement. During more than 2 years of follow-up the patient has had no respiratory problem and no gross mass in the subglottic area.

DISCUSSION

Incidence and genesis

Intralaryngotracheal goiter is, admittedly, a condition of considerable rarity. The investigation of Wegelin and Falk would indicate that the presence of thyroid tissue in this location is common, and its potential development into a goiter is not a remote possibility. Most of the reported cases came to clinical attention because of mechanical obstruction of the airway. Ziemssen described intralaryngotracheal thyroid for the first time in 1875. By 1975 approximately 115 examples of benign intra-airway thyroid tissue had been recorded, mostly in the European literature, and 90% of the affected subjects were of middle European descent. No more than ten cases have been reported in the English language literature. This tumor does not appear commonly, although it is said to constitute 7 percent of all intratracheal tumors.

Ectopic endotracheal thyroid affects females more often than males at a rate of 3:1. It presents in the third to fifth decade and is usually found in natives of endemic goiter.

Fig. 1. Cervical X-ray with deviation of the trachea.

Fig. 2-a,b. CT scan of neck with contrast showing enhanced subglottic mass.
zones of the world, accounting for the high proportion of cases in the German literature. There are two theories addressing the embryologic basis. The malformation theory proposes that the thyroid gland, arising during early embryonic life, is encroached upon and divided by the later developing laryngeal and thyroid cartilages. The ingrowth theory claims that the maturing thyroid gland may grow into incompletely formed laryngotracheal cartilages in the late fetal or postnatal period. It has been suggested that both theories may be correct. Certainly there have been examples of both subtypes reported in the literature.

Clinical features

Aberrant benign thyroid masses may present as either an unrelated autopsy finding or with symptoms such as dyspnea, hemoptysis, or adult onset asthma. Intratracheal goiter usually leads to respiratory embarrassment, which may become more pronounced with respiratory infections, at puberty or with pregnancy. Cyclic respiratory symptoms associated with menstruation have been reported. Rarely the first symptoms appear in the neonatal period. While the average duration of symptoms before diagnosis has been about four or five years, the range is much greater and encompasses such extremes as a few weeks in Ziemssen’s case and 27 years in a case described by Wurster.

Dowling states that 74 percent of all intratracheal thyroids are associated with goiters. Not infrequently the respiratory obstruction has been erroneously attributed to the external goiter, and thyroidectomy has failed to bring relief. It is not uncommon to find case histories in which aberrant thyroid masses had undergone physiologic hypertrophy in a recently thyroidectomized patient and produced severe, progressive dyspnea.

Diagnosis

In any patient with a narrowed tracheal lumen it must be ascertained whether the reduction in caliber is the result of extrinsic compression of the trachea or an intrinsic tracheal mass. An intratracheal thyroid mass may be obvious on the routine chest film as a smooth narrowing of the tracheal air shadow. In the differential diagnosis of laryngotracheal thyroid tissue one must consider and exclude any of the causes of extrinsic and intrinsic tracheal obstruction, i.e., vascular or lymphatic compression, thymic tumor, laryngotracheal chondroma, chondrosarcomas, and adenoid cystic mucoepidermoid and squamous cell carcinomas, etc. Of course before intratracheal thyroid can be diagnosed, the physician must suspect its existence. Intraluminal thyroid tissue may be found anywhere between the glottis and the tracheal bifurcation, and they usually present as broad-based, smooth, rounded masses protruding from the left posterolateral subglottic wall. The overlying mucosa usually is intact, but there may be evidence of chronic irritation. Multiple nodules and ulceration with hemorrhage are not common and should arouse suspicion of a malignancy. Less frequently, the lesion presents on the right side, anteriorly, more distally in the trachea, on the vocal cords, or on the aryepiglottic folds. For simplicity of terminology, the lesion will be referred to as intratracheal goiter, although many cases have been found in the distal larynx or, less commonly at the level of the vocal cords.

In more than two-thirds of cases, a goiterous enlargement of the normally placed thyroid is coexistent, often with a connecting tongue of tissue between it and the ectopic tissue. Lack of familiarity with this anatomic arrangement can lead to an erroneous impression of direct invasion by differentiated follicular carcinoma of the thyroid gland. True ectopic thyroid tissue may also undergo malignant degeneration in this site as well as in other ectopic sites.

Therapy

Different treatment modalities including hormonal suppression, radioactive iodine, and radiotherapy have been considered. There is only one report of shrinkage of intralaryngotracheal goiter in an infant by levothyroxine. Most authors believe there is no place for medical management in the therapy of these stenotic lesions, although the actual surgical procedure varies with the tissue diagnosis. Most authors feel that laryngo- or tracheo-fissure with submucosal dissection is the procedure of choice for benign tumors. As much tracheal mucosa as possible should be spared and rarely is there a need for a postoperative stent. Malignant tumors arising in intratracheal thyroid tissue are treated like other cancers in this location.

REFERENCES

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