



Benign Intratracheal Thyroid: A Systematic Review of 43 Cases With Five New Case Reports

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Objective/Hypothesis: To examine the clinical features of benign intratracheal thyroid (ITT) and their management strategies and outcomes.

Study Design: Case series study.

Methods: This systemic review was conducted in two international academic centers. This review includes 43 patients: one new case from the Massachusetts Eye and Ear Infirmary, four new cases from Beijing Tongren Hospital, and 38 previously published cases. We analyzed these 43 cases and summarized the patients' epidemiological data, clinical features, and treatment regimens.

Results: ITTs were less common in men than in women (male:female ratio of 3:10). ITT was observed in patients as young as neonates and as old as 85 years. Orthotopic thyroid nodules were present in 55.8% of the patients with ITT. Malignancy was incidentally found in 4.6% of all ITTs. Imaging examinations showed that the ITTs were typically attached to the posterolateral/lateral tracheal wall of the first, second, or third tracheal rings. Tissue attachment between the ITT and normal thyroid lobes was seen in 59.5% of the patients. Thirty-seven patients underwent surgery: 30 underwent open surgery, and seven underwent endoscopic debulking resections. One neonate received thyroid suppression therapy. One patient with ITT and papillary thyroid cancer was treated with radiotherapy and ultimately died after recurrence.

Conclusions: Surgical resection is an effective treatment for benign ITT. We hypothesized that abnormalities during the embryonic development of Berry's ligament might play a role in ITT pathogenesis.

Key Words: Intratracheal thyroid, pathogenesis, surgical treatment, endotracheal tumor, ectopic thyroid.

Level of Evidence: NA

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INTRODUCTION

Intratracheal thyroid gland tissue (ITT) is extremely rare and accounts for 7% of all primary endotracheal tumors.¹ Patients with ITT may present with a long

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H.M.—design, conduct, analysis of research. J.A.B.—conduct, presentation of the research. A.J.I., P.L., Q.Z., G.W.R.—presentation of research. J.F.—design of research.

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stridor and cough duration and often experience delayed diagnosis. ITTs are more commonly found in women than in men and most often affect patients of 30–40 years of age.² ITT is traditionally considered to be ectopic thyroid tissue. During the first 3–6 weeks of normal embryonic development, thyroid progenitor cells descend to the pretracheal region from the foramen cecum as the thyroglossal duct descends caudally, forming the normal thyroid gland.³ Nevertheless, the mechanism of ITT pathogenesis is unclear owing to the rarity of ITT. In addition, the diagnosis and management of ITT are not well established. In order to help address these gaps in clinical knowledge, we report new ITT cases in combination with a detailed review of the literature.

A patient with ITT from the Massachusetts Eye and Ear Infirmary (MEEI) was diagnosed in 2018, and four patients with ITT from Beijing Tongren Hospital (BTH) were diagnosed in 2016–2020 (all previously unreported cases) are presented here. In addition, 38 patients with ITT were found in the literature. To the best of our knowledge, this constitutes the largest published group of patients with ITT.

MATERIALS AND METHODS

Literature Search

A comprehensive electronic literature search was performed to identify the relevant articles, according to the PRISMA

guidelines. The PubMed and Embase databases were searched for relevant studies published from January 1958 to December 2020 using the keywords “ectopic thyroid,” “intratracheal thyroid,” and “intralaryngotracheal thyroid.” The papers selected for this systematic review were ITT case reports with information about the symptoms, imaging examinations, pathology, and treatments. The population was the patients with a diagnosis of ITT. There was no specific intervention and no comparator. The outcomes were patients’ epidemiological data, clinical features, and treatment regimens. If more than one report was based on the same case, the most recent case report was selected. Only the articles written in English were included. Patients with malignant nodules in the orthotopic thyroid gland with tracheal invasion were excluded, as these cases are difficult to distinguish from true ITTs.

Data Analysis

Data including age, sex, symptoms, tracheal site of the ITT, thyroid function, pathological findings for the ITT, pathology of the normal orthotopic thyroid (NT), treatment, connection between the ITT and NT, and follow-up were analyzed.

A review of the medical records from January 1990 to December 2020 was performed at the MEEI and BTH. The cases were selected using the same inclusion and exclusion criteria as for the literature search. Approval was obtained from the

institutional review boards of the MEEI and BTH. All five new cases provided informed consent for inclusion in this paper.

RESULTS

According to the PRISMA guidelines, we searched PubMed and Embase up to December 2020 with the search strategy: (((ectopic thyroid) AND (intratracheal thyroid)) OR (intratracheal thyroid)) OR (intralaryngotracheal thyroid), and 649 papers were found. Then we read the 649 abstracts, and only 114 papers with specific intratracheal thyroid case reports were selected. We identified 38 patients reported from 114 cases of ITT (Fig. 1); two were excluded for not being reported in English, 71 were excluded because of lacking detailed information, two were excluded because being redundant older reports of the same patient, and one was a report of a patient with thyroid cancer with tracheal invasion. Of the remaining 38 reports, all have orthotopic thyroid glands which could be distinguished from “ectopic thyroid.” A summary of the clinical details of the 38 ITT patients are shown in Table I.^{1,2,4-39}

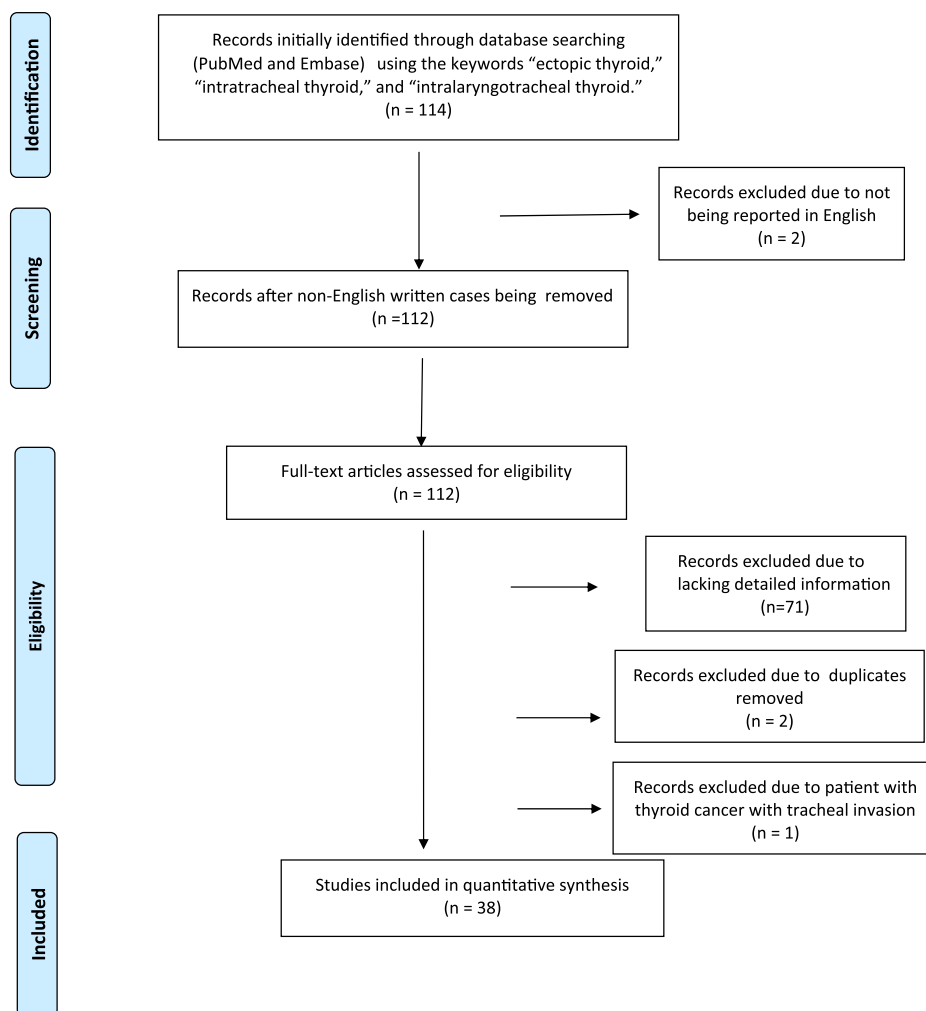


Fig. 1. Selection process of the cases.

TABLE I.
Clinical Data for 38 Cases With Intratracheal Thyroid Identified in the Literature.

Source	Age/Sex	Symptoms	Tracheal site	Connection Between ITT and NT Present	Pathology	Treatment	Follow-up
Waggoner ⁴	19 years/F	Wheeze, respiratory obstruction	Right, posterolateral, subglottic	NA	Benign	Surgery and RAI	No recurrence reported
Beck ⁵	0.5 years/F	Incidental finding by autopsy	Left, posterolateral, subglottic	Yes	Benign	No treatment	Incidentally diagnosed during autopsy
Dowling et al. ²	38 years/F	Wheeze, cough	Left, posterolateral, subglottic	Yes	Benign	Surgery	No recurrence reported
Randolph et al. ⁶	16 years/F	Respiratory distress	Left, posterolateral, subglottic	Yes	Benign	Surgery	No recurrence reported
Bone et al. ⁷	44 years/F	Progressing dyspnea	left, lateral, subglottic	Yes	Benign	Surgery	No recurrence reported
Myers and Pantangco ⁸	56 years/F	Progressive dyspnea	Right, posterolateral, subglottic	Yes	Benign	Surgery	No recurrence reported
Rotenberg et al. ⁹	47 years/F	Hemoptysis, chronic bronchitis	Right, lateral, subglottic	NA	Incidental PTC	Surgery	No recurrence reported
Donegan and Wood ¹⁰	31 years/F	Wheeze	Left, lateral, subglottic	Yes	Benign	Surgery and suppression therapy	No recurrence reported
Hazarika et al. ¹¹	60 years/F	Progressive respiratory distress	Left, anterolateral, subglottic	NA	Benign	Surgery	No recurrence reported
Ferlito et al. ¹²	77 years/M	Incidental finding by autopsy	Anterior, subglottic	NA	Benign	No treatment	Incidentally diagnosed during autopsy
Chanin and Greenberg ¹³	5 days/M	Respiratory distress	Left, posterolateral, subglottic	NA	NA	Suppression therapy for 10 years	No recurrence reported
Osammor et al. ¹⁴	57 years/M	Progressive hoarseness and breathlessness	Left, posterolateral, subglottic	NA	Benign	Surgery	No recurrence reported
al-Hajjaj ¹⁵	30 years/F	Dry cough, difficulty in breathing	Right, lateral, subglottic	NA	Benign	Surgery and suppression therapy	No recurrence reported
Ogden and Goldstraw ¹⁶	43 years/F	Stridor, dyspnea	Posterior, subglottic	Yes	Benign	Surgery	No recurrence reported
Soylu et al. ¹⁷	32 years/F	Severe dyspnea	Left, posterolateral, subglottic	NA	Benign	Endoscopic surgery	No recurrence reported
Muysoms et al. ¹⁸	62 years/F	Dyspnea, dry cough	Right, posterolateral, subglottic	Yes	Benign	Surgery	No recurrence reported
See et al. ¹⁹	33 years/M	Slowly progressive stridor	Left, posterolateral, subglottic	No	Benign	Surgery and RAI	No recurrence reported
Brandwein et al. ²⁰	24 years/F	A gagging or choking sensation	Left, posterolateral, subglottic	NA	Benign	Surgery	No recurrence reported
Hari et al. ²¹	64 years/M	Incidental finding because of nasal polyposis	Right, posterolateral, supracarina	No	Incidental PTC	Radiotherapy and salvage surgery	Died of PTC recurrence and metastasis
Døssing et al. ¹	29 years/F	Gradual progression of dyspnea	Right, posterolateral, subglottic	No	Benign	Surgery	No recurrence reported
Byrd et al. ²²	54 years/M	Incidental finding by MRI	Right, anterolateral, subglottic	NA	Benign	NA	NA
Bowen-Wright and Jonklaas ²³	19 years/F	Shortness of breath	Left, lateral, subglottic	NA	Benign	Endoscopic surgery and thyroid stimulating hormone suppression therapy	No recurrence reported
Ramalingam et al. ²⁴	48 years/M	Difficulty in breathing	Right, posterolateral, subglottic	Yes	Benign	Surgery	No recurrence reported
Abboud et al. ²⁵	39 years/F	Dyspnea and cough	Right, posterolateral, subglottic	NA	Benign	Surgery	No recurrence reported
Karakullukçu et al. ²⁶	40 years/M	Shortness of breath	Left, posterolateral, subglottic	NA	Benign	Surgery	No recurrence reported
Sung et al. ²⁷	33 years/F	Gradually progressive dyspnea	Left, posterolateral, subglottic	Yes	Benign	Surgery	No recurrence reported
Khan et al. ²⁸	81 years/F	Cough and shortness of breath	Right, posterolateral, subglottic	NA	Benign	Surgery	No recurrence reported

(Continues)

TABLE I.
Continued

Source	Age/Sex	Symptoms	Tracheal site	Connection Between ITT and NT Present	Pathology	Treatment	Follow-up
Verburg et al. ²⁹	41 years/F	Dyspnea	Left, posterolateral, subglottic	Yes	Benign	Endoscopic surgery, RAI, and suppression therapy	No recurrence reported
Hall et al. ³⁰	85 years/F	Persistent orthopnea and stridor	Left, posterolateral, subglottic	NA	Benign	Endoscopic surgery and suppression therapy	No recurrence reported
Love et al. ³¹	79 years/F	Dyspnea and stridor	Right, posterolateral, subglottic	Yes	Benign	Surgery	No recurrence reported
Serraj et al. ³²	30 years/F	Progressively worsening dyspnea	Right, posterolateral, subglottic	Yes	Benign	Surgery	No recurrence reported
Hussain et al. ³³	42 years/F	Episodes of dyspnea	Left, posterolateral, subglottic	NA	Benign	Surgery	No recurrence reported
Rispoli et al. ³⁴	17 years/M	Cough and dyspnea	Left, lateral, subglottic	NA	Benign	Endoscopic surgery	No recurrence reported
Furnas et al. ³⁵	1 day/F	Inadequate regular respiration	Left, posterolateral, subglottic	Yes	Benign	No	Died of airway obstruction secondary to ITT
Rahman et al. ³⁶	29 years/M	Breathing difficulty	Right, posterolateral, subglottic	Yes	Benign	Endoscopic surgery	No recurrence reported
Pantha et al. ³⁷	15 years/F	Difficulty in breathing	Left, posterolateral, subglottic	Yes	Benign	Surgery	No recurrence reported
Paknezhad et al. ³⁸	73 years/F	Denied any associated symptoms	Right, lateral, subglottic	No	Benign	Endoscopic surgery	No recurrence reported
Liu et al. ³⁹	61 years/M	Slight hemoptysis	Left, lateral, subglottic	Yes	Benign	Surgery	No recurrence reported

ITT = intratracheal thyroid; NA = not available; NT = normal orthotopic thyroid; Pathology = pathological diagnosis of the ITT; PTC = papillary thyroid cancer; RAI = radioactive iodine.

The five new patients with ITT were identified from the MEEI and BTH. One case was diagnosed at the MEEI in July 2018, and four patients were diagnosed at the BTH between April 2016 and December 2020 (Table II). For the first four cases, local anesthesia and tracheotomy were performed first, and intraoperative pathological examination was performed. Surgery was completed after benign thyroid tissue was confirmed by intraoperative frozen sections. For case 5, after local anesthesia and tracheotomy, the biopsy was done by endoscopy. Surgical treatment under secondary anesthesia was performed after benign thyroid tissue was confirmed by postoperative pathological results.

Case 1

The patient was a 67-year-old woman who was initially admitted for a complaint of 3 years' duration of cough, dyspnea, and stridor, which worsened in the previous 2 months. A computed tomography (CT) scan revealed an intratracheal mass protruding through the tracheal wall and connecting to the right thyroid lobe (Fig. 2A). Flexible laryngoscopy showed a broadly based submucosal neoplasm at the level of the first tracheal ring (Fig. 2B). An awake tracheostomy was performed, and direct laryngoscopy with frozen biopsies resulted as "thyroid tissue." An open approach was then undertaken

to excise the ITT. First, a right thyroid nodule enucleation was done, which revealed a "benign goiter" on frozen pathological analysis. The tracheal lumen was then entered by performing an anterior split of the cricoid cartilage and the first three tracheal rings (Fig. 2C,D). The mass was detached from the first and second tracheal cartilage rings. As the pedicle between the ITT and NT was very thin, it was not excised. The recurrent laryngeal nerve (RLN) was identified, but the mass could be detached without damaging the RLN. A tracheal T-tube stent was placed. The patient was successfully decannulated 6 months postoperatively. The postoperative period was uneventful, with no voice complaints or recurrence reported during the 4 years of follow-up, and no further treatment was necessary.

Case 2

The patient was a 44-year-old woman who presented with complaints of dyspnea for 10 years. Laryngoscopy revealed a smooth, pink mass in the upper trachea, and CT imaging demonstrated a mass attached to the right posterolateral wall of the trachea (Fig. 3). An awake tracheostomy was performed. After confirming the diagnosis of normal ITT by direct laryngoscopy and biopsy, the patient underwent an open excision of the mass. During resection, a thin stalk was seen connecting the right

TABLE II.
Clinical Data of Patients With Intratracheal Thyroid From BTH and MEEI.

Case	Age/Sex	Tracheal Site	Connection Between ITT and PNT	Thyroid Function	Pathology	Treatment	Hospital
1	67 years/F	Right, posterolateral, subglottic	Yes	Normal	Benign	Surgery	BTH (2016)
2	44 years/F	Right, posterolateral, subglottic	Yes	Normal	Benign	Surgery	BTH (2017)
3	61 years/F	Left, posterolateral, subglottic	Yes	Normal	Benign	Surgery	BTH (2017)
4	63 years/F	Right, posterolateral, subglottic	Yes	Normal	Benign	Surgery	BTH (2018)
5	53 years/F	Right, posterolateral, subglottic	Yes	Normal	Benign	Surgery	MEEI (2018)

BTH = Beijing Tongren Hospital; ITT = intratracheal thyroid; MEEI = Massachusetts Eye and Ear Infirmary; NT = normal orthotopic thyroid.

thyroid lobe to the ITT through the trachea. A right hemithyroidectomy was performed, and the RLN was identified, protected, and preserved. A 1.0 × 0.5 cm cartilage defect in the tracheal wall was identified, which was repaired primarily. Pathology revealed benign thyroid tissue as reviewed by two experienced pathologists. The patient was successfully decannulated 3 months after surgery. There was no evidence of recurrence during the 3 years of follow-up.

Case 3

A 61-year-old woman presented with complaints of a mild cough and increasing shortness of breath for 4 months. A smooth, subglottic, submucosal mass was detected by laryngoscopy. CT imaging indicated that the mass was attached to the left posterolateral wall of the trachea, and multiple thyroid nodules were detected by ultrasound (Fig 4A,B). Awake tracheostomy was performed. The patient underwent a left hemithyroidectomy

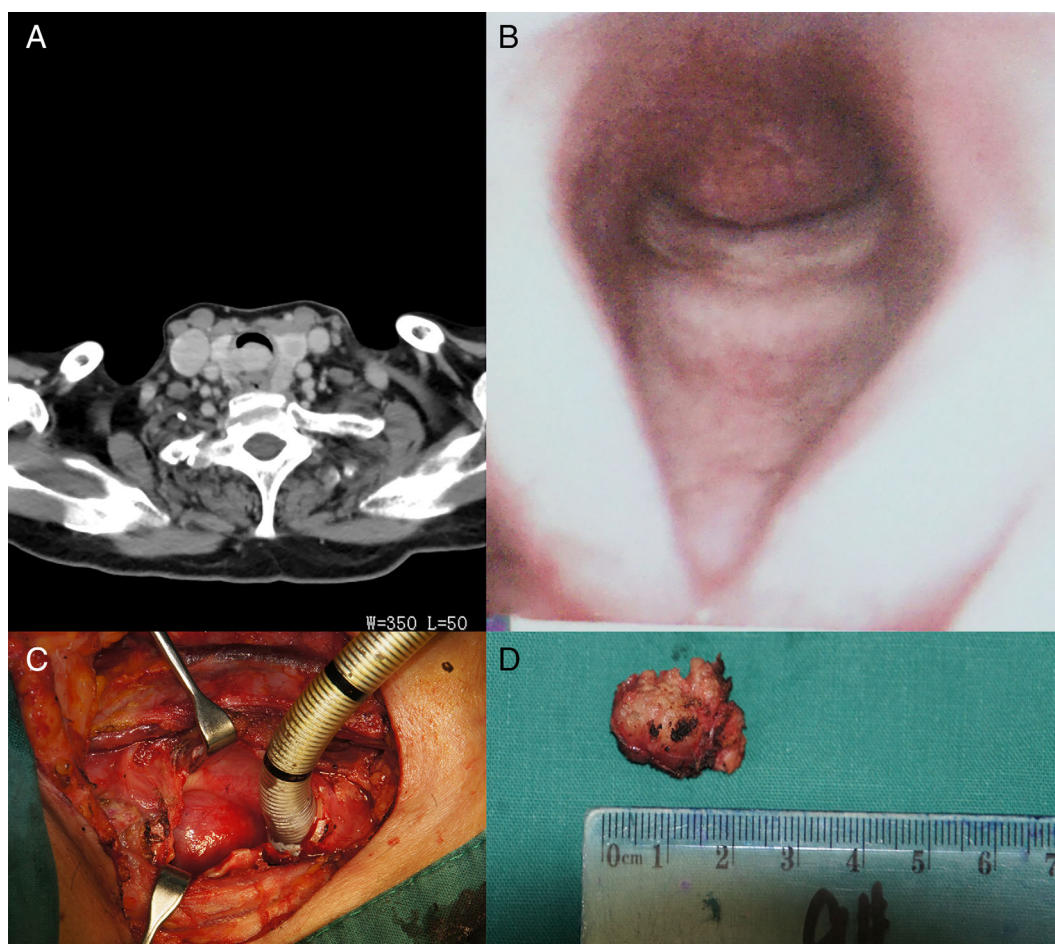


Fig. 2. Images for case 1. (A) Computed tomography scan showing a large, homogenous, and smooth 2 × 1.5 cm intratracheal mass attached to the right posterolateral tracheal wall. The mass protruded through the tracheal wall and was connected to the right thyroid lobe. (B) View of the intratracheal thyroid (ITT) through the transnasal fiberoptic scope. A submucosal broadly based neoplasm was seen at the level of the first tracheal ring, and more than 90% of the tracheal lumen was occupied by the mass. Mobility of both vocal cords was normal. (C) Exposure of the ITT during the surgery (right: inferior; left: superior). (D) The resected specimen.



Fig. 3. Images for case 2. A mass attaching to the right posterolateral tracheal wall without clear borders to the trachea.

and excision of the ITT with RLN identification, protection, and preservation. The pedicle was resected with the surrounding cartilage. Frozen biopsy demonstrated benign thyroid tissue. The tracheal defect caused by the pedicle was small and was closed primarily. Tracheal

reconstruction was undertaken, and a tracheal T-tube was placed. Decannulation was achieved 6 months postoperatively. This patient had no postoperative complications or recurrence.

Case 4

A 63-year-old woman presented with complaints of cough and increasing shortness of breath for 1 month. She had undergone a partial thyroidectomy with nodule enucleation for benign goiter 14 years before. A mass obstructing 90% of the tracheal lumen was observed in the subglottic region by laryngoscopy. CT scan showed a mass on the right posterolateral tracheal wall (Fig. 5A,B). Multiple thyroid nodules in the NT were identified by ultrasound. Awake tracheostomy was performed first, and then a right hemithyroidectomy was performed with RLN identification and preservation under general anesthesia. After confirming the diagnosis of ITT through frozen biopsy, the trachea was split in the midline to expose the ITT. The ITT was excised along with a small portion of the second tracheal ring cartilage, and the trachea was closed primarily. The right sternothyroid muscle was used to bolster the tracheal incision, and a T-tube was placed. The patient underwent decannulation 6 months later. This patient had no postoperative complications or recurrence.

Case 5

A 53-year-old female presented with complaints of shortness of breath and stridor worsening over the prior year. CT imaging revealed dehiscence in the right posterolateral aspect of the cricoid cartilage, with protrusion of

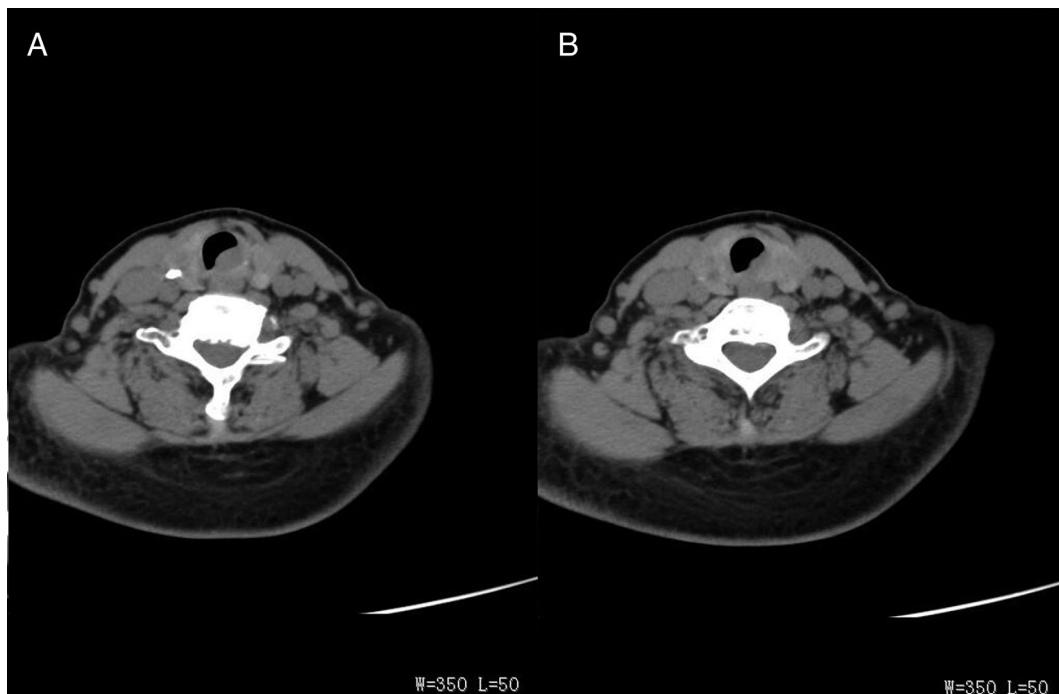


Fig. 4. Images for case 3. (A) A mass attaching to the left posterolateral tracheal wall with clear border to the trachea. (B) A connection could be seen between the intratracheal thyroid and left thyroid lobe, and multiple thyroid nodules were detected.

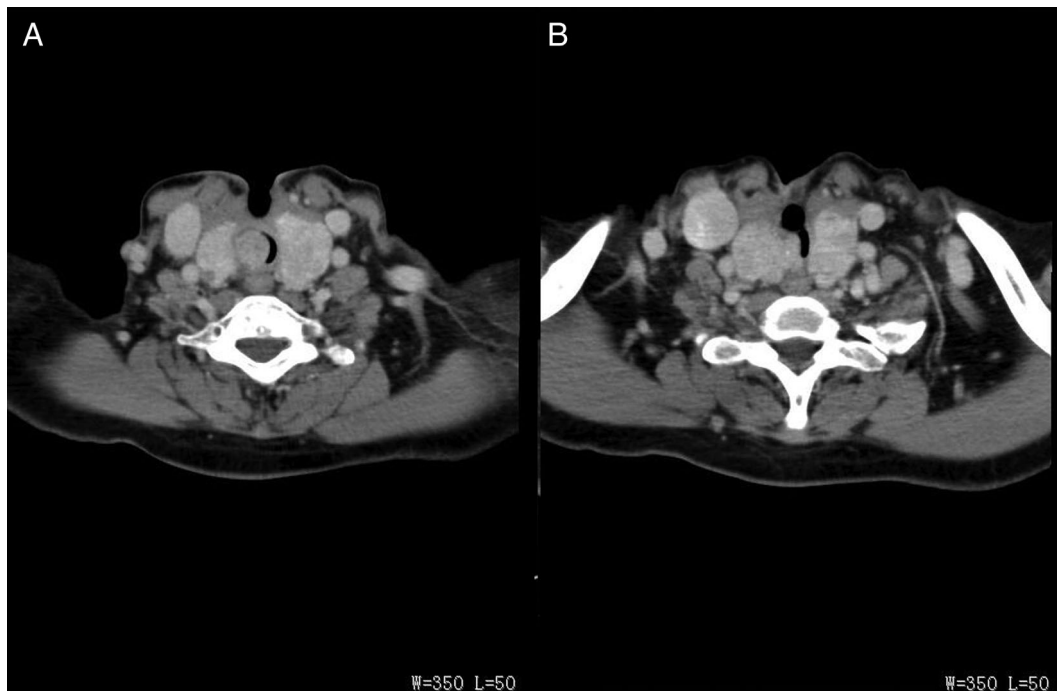


Fig. 5. Images for case 4. (A) A $1.3 \times 1.4 \times 1.7$ cm sharply demarcated mass on the right posterolateral tracheal wall, occupying 90% of the tracheal lumen. (B) A stalk between the intratracheal thyroid and the right thyroid lobe could be seen, which penetrated into the trachea through the space between the first and second tracheal ring.

thyroid tissue into the subglottic airway. Emergency direct laryngoscopy revealed a firm, submucosal mass emanating from the right posterior subglottis, obstructing 80% of the airway at this level. An urgent tracheostomy was performed. Biopsy demonstrated thyroid tissue with no evidence of malignancy. Definitive surgery was performed, which included right hemithyroidectomy. The right RLN was found to be intricately involved in the airway dehiscence area, necessitating intentional nerve resection. RLN resection was expected and discussed with the patient preoperatively. The ectopic thyroid tissue was dissected from the outer part of the tracheal mucosa, keeping the mucosa intact. The integrity of the tracheal wall was verified by endoscopy. The muscles were carefully closed over and used to bolster the resected area. On final pathology, two foci of papillary thyroid carcinoma were identified in the right lobe. Both were deemed incidental and distinct from the area of ITT. The patient was decannulated postoperatively, and the tracheostoma healed. There was no report of recurrence or complication from treatment to the time of this report.

Combined Cases

The data of the 43 ITT patients (38 from the literature and five patients) are presented below. The results are also summarized in Tables I and II.

Demographics

There were 32 (74.4%) women and 11 (25.6%) men, with a median age of 42. Sixteen (37.2%) patients had a history of thyroid surgery.

Presentation

Stridor (11.63%), breathlessness (18.60%), and dyspnea (41.86%) were the major complaints of patients with ITT, and most patients experienced these symptoms for years before diagnosis. Symptoms became severe in four (10.3%) patients, who were either pregnant or adolescents.

Imaging and Laboratory

NT was present in all 43 (100%) patients. In all patients, benign ITT appeared as a submucosal, smooth mass with a broad base. The ITTs were located in the upper trachea (from the first to third tracheal rings) in 42 (97.7%) patients and above the carina in one (2.3%) patient. Thyroid function results were available for 23 (53.5%) patients; thyroid stimulating hormone levels were normal in 17 patients (17/23, 73.9%). Ten (23.3%) patients underwent radionuclide thyroid scans before surgical treatment, revealing minimal radionuclide uptake in four (4/10, 40.0%) patients and moderate uptake in six (6/10, 60.0%) patients. In the five new cases, preoperative imaging or intraoperative manifestations all clearly showed that ITT was related to the position of Berry's ligament in the extratracheal normal thyroid gland lobe, suggesting that Berry's ligament might be involved in the occurrence of ITT.

Pathological Diagnosis

Pathological diagnosis was available in 42 (97.7%) patients. The patient without a pathological diagnosis was a newborn. Malignancy was incidentally identified in the ITT in 4.6% of the cases. Benign nodules in the NT

were detected in 24 (55.8%) patients. Two (4.6%) patients had incidental papillary thyroid microcarcinoma,^{21,24} and one (2.3%) patient had incidental medullary thyroid cancer.²²

Treatment

Thirty-seven (86.0%) patients underwent surgery, of whom 30 (30/37, 81.1%) had open surgery with an initial tracheostomy to secure the airway. Surgical debulking was performed endoscopically in seven (7/37, 18.9%) patients, and a stent was placed in eight (8/37, 21.6%) patients. A fibrous connection between the ITT and NT was seen by preoperative examinations or intraoperatively in 22 patients (22/37, 59.5%). Six patients did not undergo surgery. Some patients underwent radioactive iodine (RAI)/suppression therapy, as shown in Table I.

Follow-up

Four (9.3%) patients died during follow-up. One newborn received successful suppression therapy for more than 10 years. No recurrence or tracheal stenosis was reported in the remaining 38 (88.4%) patients.

DISCUSSION

To the best of the authors' knowledge, this study included the largest number of ITT patients. The clinical characteristics of the ITT cases were examined, highlighting some epidemiology, pathogenesis, and management features of ITT.

Less than 150 cases of ITT have been reported worldwide.^{4,5,6,7} Most studies have been published as single case reports, and almost all epidemiological data about ITT are cited from studies published at least 50 years ago.

Benign ITT is difficult to identify at an early stage because of its concealed site and is most often misdiagnosed as other airway pathology, such as asthma. In this study, the male-to-female ratio was 3:10, which is consistent with other reports.²⁰ The age of onset ranged widely from 1 day to 85 years, widening the range previously reported in the literature.²⁰ The prevalence of incidental malignancy in this study was 4.6%, which was below the widely cited 11% reported in 1962.² Still, the 11% prevalence could be an overestimation due to the limited pathological techniques available before 1962.^{20,23} Sixteen (37.2%) patients had a history of thyroid surgery before ITT was diagnosed.

Pathogenesis of ITT

Ectopic thyroid has been classically subdivided into two groups. The first group constitutes patients with ectopic thyroid tissue on the descending route of the thyroglossal duct from the foramen cecum to the pretracheal region. In the second group, ectopic thyroid foci are located away from the descending route, such as associated with the heart, pancreas, and esophagus.^{40,41,42}

As described in this study, benign ITT does not appear to belong to either of the two ectopic subgroups described above, and its underlying mechanism is

controversial. Malformation and ingrowth are the two historical hypotheses for ITT development.²² The malformation theory proposes that a portion of thyroid tissue is incorporated in the trachea from the NT during embryonic development, while the ingrowth theory proposes that thyroid tissue grows into the trachea during the late fetal or postnatal period. Neither theory explains why this process usually occurs in the lateral or posterolateral upper trachea, as seen in this study.

Our study provides clues for the pathogenesis of benign ITT. There were three infants with ITT, and Falk also reported finding thyroid tissue protruding into the trachea in neonatal autopsies.²⁶ This suggests that the embryonic period might be a crucial period for ITT development. In 40 (93.0%) of our patients, the ITT was attached to the posterolateral or lateral tracheal wall of the first to third rings. It is also the site of Berry's ligament, which is responsible for attaching the thyroid to the trachea. A stalk was identified in 22 (59.5%) of the patients in this study, representing the connection between the NT and the ITT. We speculate that ITT might result from the protrusion of Berry's ligament from the area of lateral tracheal dehiscence.

Although it can be speculated that excessive development of Berry's ligament with lateral tracheal dehiscence could be an etiology for ITT, the findings in two previously published cases could not be explained with this theory. In one patient, thyroid tissue was found in the esophagus, protruding into the trachea.¹⁶ In the second patient, the ITT was in the supracarinal region.²¹ In these two patients, the lesion might be an ectopic thyroid, similar to patients with ectopic thyroid tissue associated with the heart.

Management of ITT

Benign ITT should be differentiated from other intratracheal tumors, such as adenoid cystic carcinoma and thyroid cancer with intratracheal invasion, and biopsy plays a critical role in the diagnosis. Tracheostomy under local anesthesia is usually required to relieve dyspnea. Imaging studies such as CT scans can provide helpful anatomic information to guide the surgery.

Surgery through an open approach is the conventional treatment for ITT and provides an optimal surgical view to excise the mass and perform concurrent hemithyroidectomy. Nevertheless, there is no consensus on treating the stalk that connects the ITT to the NT. If no stalk is seen by the naked eye, it could be recommended to remove the ITT completely. If the stalk is clear and very thin, we recommend transecting the stalk at the insertion site. If the stalk is thick or has no clear margin with the trachea, excising the ITT with a small portion of the tracheal wall could be recommended in most patients, as was performed in cases 2, 3, 4, and 5 of the present study. Tracheal reconstruction is then required. Tension sutures, end-to-end anastomosis, and primary repair with overlaying strap muscles are all acceptable choices depending on the circumstance. In case 1, we also attempted excision of the ITT while leaving a pedicle tissue behind, as it was noted to be quite thin; no evidence of recurrence was observed during the 4 years of follow-up. We,

therefore, do not consider benign ITT an absolute indication for a hemithyroidectomy, even when there is a clear pedicle between the thyroid lobe and the ITT.

Laser therapy during endoscopic surgery has been used in ITT treatments since 1993.¹⁷ Because endoscopic surgery is a debulking operation, it is often performed in patients with advanced age, poor general health, and those who decline open approach surgery. In these patients, the main reason for endoscopic surgery was to obtain an accurate diagnosis, and suppression therapy was usually required postoperatively. If malignancy were identified, other adjuvant treatments might be employed. Although the tracheal injury is minimized with endoscopic surgery, hemostasis is challenging. However, with improved instrumentation and hemostatic techniques, endoscopic surgery may be a promising future treatment for benign ITT. The decision to place a stent is based on a patient's condition and the surgeon's experience; T-tubes, silastic sheets, and titanium mesh have all been used successfully in previous studies.

Primary treatment with thyroid suppressive therapy has been reported in only one neonate, and the mass significantly decreased in size.¹³ RAI may increase the risk of secondary malignancy and address the airway issues. RAI is not recommended routinely for benign ITT.

Of course, this study has limitations. From 114 initially identified cases, only 38 could be included. In addition, only the data from the articles could be analyzed, and the descriptions of the symptoms were not uniform.

CONCLUSION

ITT cases are extremely rare, and the diagnosis and management of these lesions pose unique challenges to surgeons. Surgery through an open approach is the conventional treatment for ITT, yielding low postoperative complication rates and adequate control of the airway symptoms. It could be hypothesized that Berry's ligament plays an important role in ITT development. Still, the full mechanism of ITT pathogenesis remains unclear and represents an area for future research.

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