MTC in Thyroglossal Duct Cyst (Case Discussion)

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CASE REPORT

- A 26-yr-old woman presented with cough
- A soft, mobile, 2-cm anterior neck mass
- No thyroid abnormalities or cervical lymphadenopathy
- Diagnosis: Thyroglossal duct carcinoma (TGDC)
THYROGLOSSAL DUCT CYST
THROGLOSSAL DUCT CYST

- The most common congenital abnormality of thyroid development, involving around 7% of the population
Many cystic remnants of the thyroglossal tract are never detected clinically

A postmortem study of 200 adults found a 7% incidence of TGDC

Ellis PD, van Nostrand AW  
Laryngoscope. 1977;87(5 Pt 1):765
The presence of thyroid tissue, in a TDC, is due to its embryologic origin and ranges from 1.5% to 62% of cases.
EMBRYOLOGY
THYROID GLAND

- Endoderm of the floor of mouth between the 1st and 2nd archs.
- Descends as a bilobed diverticulum from the foramen cecum around the 4th week to rest by the 7-8th week.
THYROGLOSSAL DUCT CYST

- Most common congenital midline mass
- Asymptomatic mass at or below the hyoid bone that elevates with tongue protrusion.
- Ectopic thyroid tissue vs. thyroglossal duct cyst?
THROGLOSSAL DUCT CARCINOMA

- Thyroglossal duct carcinomas are very uncommon findings, occurring in approximately 1% of all the thyroglossal duct cysts, often incidentally discovered following surgical excision.
Thyroglossal Duct Remnants - can have thyroid tissue or form Thyroglossal Duct cysts anywhere along path.
The standard operation for thyroglossal duct cyst is to resect the cyst in continuity with the midportion of the hyoid bone and a core of tissue from the hyoid upwards toward the foramen cecum, an operation known as the Sistrunk procedure.
SISTRUNK TECHNIQUE

- Complete excision of the cyst and the thyroglossal duct with central hyoidectomy
- Cyst recurrences are rare
DIAGNOSIS OF TGDC IS MADE AFTER SURGICAL REMOVAL OF THE THROGLOSSAL DUCT CYST FOR OTHER REASONS

19 patients of 26 cases in Pellegriti et al series
Most thyroglossal carcinomas are located in the thyrohyoid region
TDCs

- 60% between the hyoid bone and the thyroid cartilage
- 13% in the substernal region
- 24% above the hyoid bone including the submental site
- 2% intra-lingual
AGE DISTRIBUTION

- 31% occur before the age of 10 years
- 20% in the second decade
- 14% in the third decade
- 35% after 40 years of age.
FNA

- True-positive rate 53%
- True-negative rate 47%

Rosenberg TL, Brown JJ, Jefferson GD
Med Clin North Am. 2010;94(5):1017
Thyroid Cancer in Thyroglossal Duct Cysts Requires a Specific Approach due to Its Unpredictable Extension
THYROGLOSSAL DUCT CARCINOMA: REVIEW OF RECENT LITERATURE

## Thyroglossal Duct Carcinoma: Review of Recent Literature

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TGDC Carcinoma

- Uncommon, 1%
  - 94% Thyroid- Papillary
  - 6% Squamous Cell

- TGDC Carcinoma or a Metastatic Cystic Thyroid Carcinoma in a Midline Lymph node?

No cases of medullary thyroid cancer (MTC)
MEDULLARY CARCINOMA IN A LINGUAL THYROID

- Although there is no data on the occurrence of C-cell rests in the tongue, the presence of such cells in the thymus and parathyroid indicates that their migration is occasionally subject to aberration.

- This aberrant migration might be the source of C-cells in this case.

Singapore Med J 2008; 49(3): 251-253
Yaday S, Singh I, Singh J, Aggarwal N
In this case

Tgdc MTC + PTC
lung MTC
NECK MASS IHC

- Amyloid  +
- Cacitonin  +
- Chromogranin +
- NSE  +
LUNG MASS IHC

- P63 +
- Thyroglobin (NEG)
- CK7 +
- Chromogranin (weakly positive)
- Calcitonin (positive)
ATA mtc guideline
Recommendation grade B

- IHC analysis:
  - calcitonin, chromogranin, CEA
  - absence of thyroglobin
Mixed medullary-follicular carcinoma is a rare tumor of the thyroid. Less than 40 cases have been reported in the literature.

mTC + pTC
IHC: calcitonin positive
Thyroglobin negative

PTC variant of mtc
MTC

- 70% of patients with palpable thyroid nodule have cervical Mets and 10% have distant Mets
Recent Data (After Operation)

- CEA 1.1 ng/dl  (double check)
- Calcitonin less than 1micIU/ml (double check)
- Ca, P, Alb normal
- TFT, normal
NON-SECRETORY MTC

- The prognosis of these patients varied between intermediate –term survival (12.5 years) and rapid progression leading to death within 1.75 years after diagnosis.
CLINICAL BEHAVIOR MTC IS UNPREDICTABLE

Some patients with distant mets may live for several years
TGDC

- 80% PTC
- 8% Follicular variant of PTC
- 6% Squamous cell cancer
- 6% FTC, Hurthle cell, and anaplastic
WHAT IS THE DIAGNOSIS

- Papillary carcinoma
- Medullary carcinoma
- Papillary + Medullary carcinoma
- PTC variant of MTC
- Mucoepidermoid carcinoma
- Pyramidal lobe Tumor
- Lymph node Mets

- Dual ectopia of thyroid tissue in remote structure: esophagus, mediastinum, heart, aorta, adrenal, pancreas, gallbladder, and skin
The distinction between thyroglossal duct remnant cancer and pyramidal lobe tumors requires specific pathological criteria, as pointed out by LiVolsi et al.

LiVolsi VA, Perzin KH, Savetsky L 1974 Cancer 34:1303–1315
Mucoepidermoid carcinoma of the thyroid is rare and may originate from ultimobranchial remnants or follicular epithelium
DIAGNOSIS IN THIS CASE:

PTC VARIANT OF MTC IN THYROGLOSSAL MASS
MTC OF THE LUNG
Thyroid Cancer in Thyroglossal Duct Cysts Requires a Specific Approach due to Its Unpredictable Extension
The surgeon’s decision to remove the thyroid gland depends on whether one believes TGDC cancer is a de novo process or whether it represents a metastasis from carcinoma within the thyroid gland.

This decision is also dependent on the adjunctive treatment plans, i.e., whether subsequent treatment with radioiodine for metastatic disease is required.
Additional approaches to treatment are more controversial and relate to varying concepts of the origin of thyroglossal duct carcinoma.
Some argue that these lesion arise de novo within the cyst, whereas others believe the tumor represents a metastasis from an occult primary carcinoma of the thyroid.
DE NOVO HYPOTHESIS

- Ectopic thyroid nests are present in as many as 62% of thyroglossal duct surgical specimens.

- Parafollicular cells are absent in ectopic thyroid tissue, and we could find no cases of medullary carcinoma of a thyroglossal duct cyst.

- The finding of unsuspected carcinoma of the thyroid gland in 11–33% of patients with a TGDC who underwent thyroidectomy provide support for the metastatic theory.
Plaza et al. proposed an algorithm for treatment

- Patients less than 45 yr of age
- Tumors less than 1.5 cm confined to the cyst
- Normal thyroid in ultraonography
- No suspicious lymph node

Simple Sistrunk procedure

Plaza CP, Lo´pez ME, Carrasco CE, Meseguer LM, Perucho Ade L
Ann Surg Oncol 13:745–752
- Total thyroidectomy
- Compartment-oriented neck dissection only if lymph node metastases are found on ultrasound or during surgery
- Radioiodine
A SLIGHTLY DIFFERENT APPROACH

Stratified patients into “low risk” and “high risk”

Patel et al
J Surg Oncol 79:134–139; discussion 140–141
Simple Sistrunk
- Younger than 45 yr
- Tumor less than 4 cm
- No Soft tissue extension
- No Metastases

Total Thyroidectomy
- Older than 45
- Tumor larger than 4 cm
- Soft tissue extension
- Nodal or distant Metastases

J Surg Oncol 79:134–139; discussion 140–141
CONCLUSION

- TGDC are the most common developmental anomalies of the thyroid gland
- 75% of midline neck tumors in children
  - 7% in adults
- Carcinoma of the thyroglossal duct remnant is reported in less than 1%
- Diagnosis: epithelial lining of the duct or cyst
- Extent of surgery: individualized
THANK YOU