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Thyroglossal Duct Carcinoma with Concurrent Papillary Thyroid Carcinoma: A Case Report

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ABSTRACT

Objective: To report a case of thyroglossal duct carcinoma with concurrent papillary thyroid carcinoma

Methods:

Design: Case Report
Setting: Tertiary National University Hospital
Patient: One

Results: A 46-year-old woman was diagnosed with thyroglossal duct carcinoma after undergoing a Sistrunk procedure. Due to presence of thyroid nodules, the patient underwent second stage thyroidectomy with central neck dissection which revealed papillary thyroid carcinoma.

Conclusion: Thyroglossal duct carcinomas are rare entities and there is no current consensus regarding their management. Difficulties arise in the diagnosis of these tumors as they present similarly to benign thyroglossal duct cysts. Most cases are diagnosed postoperatively. Proper preoperative assessment including head and neck examination, biopsy, and radiologic imaging is necessary to recognize patients who could benefit from more aggressive management.

Keywords: *thyroglossal carcinoma; thyroglossal duct cyst; papillary thyroid carcinoma*

Thyroglossal duct cysts are the most common developmental anomalies of the thyroid gland. They stem from the persistence of the tract of the migrating thyroid gland as it descends from its origin in the foramen cecum to its adult position inferior to the cricoid cartilage.[1] The thyroglossal duct cyst may contain remnants of thyroid tissue from which thyroid malignancies may arise. Thyroglossal duct carcinomas are rare malignancies accounting for 1% of all cases of thyroglossal duct tumors and may occur concurrently with thyroid malignancies.[2] We present a case of papillary thyroid carcinoma in a thyroglossal duct cyst with a concurrent thyroid papillary carcinoma.

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CASE REPORT

A 46-year-old woman presented with a three-year history of slowly growing anterior neck mass with no associated hypothyroid or hyperthyroid symptoms. There was no family history of carcinoma and the patient had no exposure to radiation. On physical examination, there was a 7 x 6 x 5.9 cm firm anterior neck mass located in the midline which moved with tongue protrusion. *(Figure 1)*

A neck ultrasonogram revealed a cystic mass in the anterior neck consistent with a thyroglossal duct cyst and normal thyroid glands. Fine needle aspiration biopsy revealed benign cyst contents and the patient underwent a Sistrunk procedure. Intraoperatively, there was note of a 6 x 5 x 4 cm cyst with solid areas adherent to the strap muscles *(Figure 2)* and an enlarged pre-laryngeal node. *(Figure 3)* Final histopathology results revealed classical variant papillary thyroid carcinoma, 4.2 cm in greatest tumor dimension, with extension to adjacent soft tissues. The enlarged pre-laryngeal node was positive for tumor.

On repeat neck ultrasonography done after the Sistrunk procedure, two cystic nodules were noted in the right thyroid lobe. The patient underwent a second stage total thyroidectomy with central neck dissection. Intraoperatively, the right thyroid lobe measured 4 x 3 x 2 cm with multiple nodules palpable in the lobe. The left thyroid lobe measured 4 x 2 x 1 cm with no masses noted within the lobe. *(Figure 4)* There were no visible or palpable enlarged central lymph nodes.

Final histopathologic examination of the thyroidectomy specimen revealed multifocal classical variant papillary carcinoma of the right thyroid lobe and multinodular colloid goiter of the left thyroid lobe and isthmus. Level 6 lymph nodes were negative for tumor. The patient was scheduled to undergo radioactive iodine therapy but was lost to follow up.

DISCUSSION

The thyroid gland initially arises as a mass from a depression at the foramen cecum during the 5th week of development. The thyroid gland migrates inferiorly and reaches its final adult position at the level below the cricoid cartilage during the 7th week of development.[1]

The most common developmental anomalies of the thyroid gland are thyroglossal duct cysts which occur in 7% of the adult population. A thyroglossal duct cyst may develop if the tract of the descending thyroid fails to degenerate. The cyst may contain remnants of thyroid tissue from which malignancies may arise.[3]

Thyroglossal duct carcinomas are rare entities with an incidence of <1% of all cases of thyroglossal duct cyst.[2] Concurrent thyroglossal duct carcinomas and thyroid carcinomas have been reported to occur at a rate between 0 – 25%.[4] However, the true incidence is difficult to ascertain as not all patients undergo thyroidectomy. The most

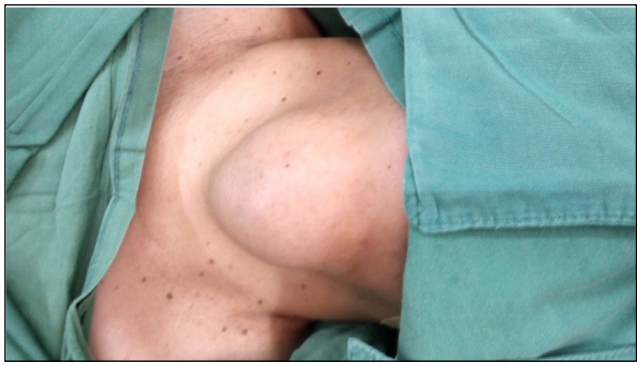

Figure 1. Preoperative picture of the patient showing a midline anterior neck mass

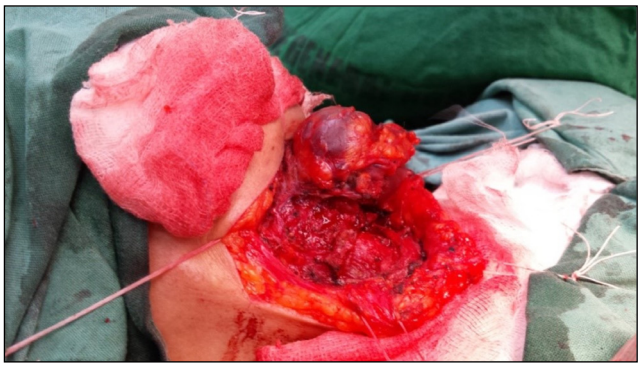

Figure 2. Thyroglossal duct cyst seen intraoperatively

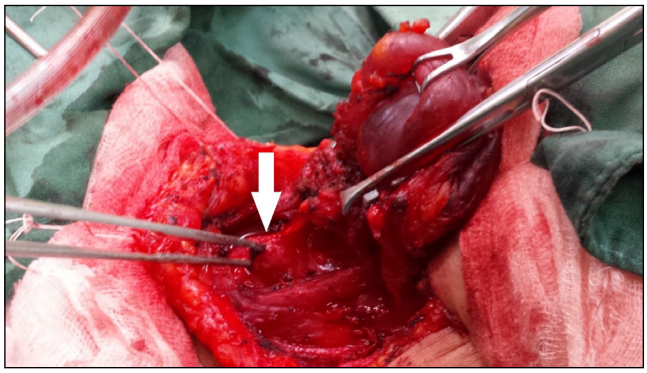

Figure 3. Arrow pointing to an enlarged pre-laryngeal node seen intraoperatively

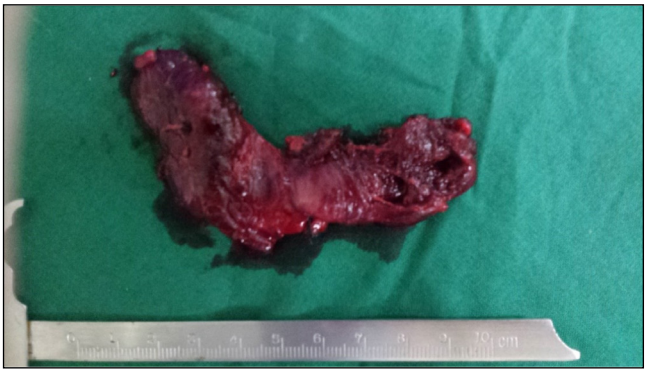

Figure 4. Resected thyroid gland

common histologic type of carcinoma found in thyroglossal duct cysts are papillary carcinomas but other types of carcinoma, such as mixed papillary-follicular, squamous cell, anaplastic, and Hurthle cell have been reported.[5] There are no reported cases of medullary carcinomas found in thyroglossal duct cysts as medullary carcinomas arise from the parafollicular cells which have a different embryologic origin from the thyroid.[6] This supports the theory that thyroglossal duct carcinomas arise de novo from the cyst itself. Two theories accounting for the origin of thyroglossal duct carcinomas are development of the carcinoma from thyroid tissue in the cyst and metastasis from a primary cancer in the thyroid gland.[7]

Fine needle aspiration biopsy and neck ultrasound are standard diagnostic tests used for preoperative evaluation of midline neck masses. However, fine needle aspiration biopsy is often inaccurate when diagnosing thyroglossal duct carcinomas.[8] Yang *et al*. reported a true positive rate of 53% and false negative rate of 47% for diagnosing thyroglossal duct carcinomas using fine needle aspiration biopsy. Ultrasound guided aspiration biopsy can improve accuracy of results.[9] On ultrasonography, presence of a central solid component, calcifications, irregular borders, and thickened cyst walls are suggestive for carcinoma. However, they may be indistinguishable from benign TGDCs.[5] Thyroglossal duct carcinomas are usually diagnosed postoperatively upon histopathologic examination of the operative specimen.

Frozen section examination of the specimen should be considered when malignancy is a consideration. Danilovic *et al*. recommended use of frozen section to diagnose papillary carcinoma due to poor sensitivity of FNAB. In their study, frozen section correctly diagnosed all cases of thyroglossal duct carcinomas based on suspicious findings on ultrasound.[10]

There is no current standard for treatment of thyroglossal duct carcinomas. Sistrunk procedure is usually done for thyroglossal duct carcinomas with some studies supporting its use as sufficient for treatment. The cure rate for papillary thyroglossal duct carcinomas has been reported at 95% when treated with Sistrunk procedure alone. Addition of total thyroidectomy with lymph node dissection increases the cure rate to 100%.[11] Some studies suggest performing a total thyroidectomy owing to the incidence of concurrent thyroid carcinoma with thyroglossal duct carcinomas.[12] This highlights the importance of adequate preoperative evaluation for the patients. Radioactive iodine therapy has also been advised for patients with thyroglossal duct carcinomas however there is still no consensus for its use.[13]

Recent studies have proposed a risk stratification system to screen for high risk patients. More aggressive treatment is recommended if the patient has any of the following features: (1) age greater than 45 years, (2) history of radiation exposure, (3) thyroid mass seen on imaging studies, (4) cervical lymphadenopathies evident clinically or on imaging studies, (5) tumor size greater than 1.5 cm in diameter, and (6) cyst wall invasion and positive margins on histopathologic examination.[5]

In conclusion, thyroglossal duct carcinomas are rare entities and there is no current consensus regarding its management. Difficulties arise in the diagnosis of these tumors as they present similarly to benign thyroglossal duct cysts. Most cases are diagnosed postoperatively. Proper preoperative assessment including proper head and neck examination, biopsy, and radiologic imaging is necessary to recognize patients who could benefit from more aggressive management.

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