



PAPILLARY CARCINOMA IN THYROGLOSSAL DUCT CYSTS (TGDC)

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ABSTRACT

Background: Thyroglossal duct cysts (TGDC) are common midline neck swelling presenting mainly in childhood. The presence of malignancy in thyroglossal duct cyst is much less common accounting for only 1% of all cases. We report three cases of papillary carcinoma in a thyroglossal duct cyst in adults.

Methods: We present a series of three patients with TGDC carcinoma. All three patients presented with midline painless neck swelling and underwent Sistrunk procedure. Histopathological examination revealed papillary carcinoma in thyroglossal cyst, following which all three patients underwent total thyroidectomy and radioactive iodine therapy. The thyroid lobes were found to be normal. Tumor or thyroglossal duct cyst recurrence did not occur in any of the patients in follow-up.

Results And Conclusions: Thyroglossal duct cyst malignancy is usually diagnosed incidentally following excision of TGDC. Total thyroidectomy for tumors of thyroglossal ducts could be justified by the high incidence of associated papillary carcinomas of the thyroid, RAI therapy could be given and better follow up by serum thyroglobulin. In these cases, a radical therapeutic attitude results in better patient outcome.

KEYWORDS : Papillary, Carcinoma, Thyroidectomy, Thyroglossal Cyst

INTRODUCTION

The thyroid gland develops as an out pouching from the primitive foregut at the level of foramen caecum which descends into the neck^[1]. It leaves behind an epithelial tract known as the thyroglossal tract, which usually disappears at during 5th to 8th week of gestation^[4]. Incomplete obliteration of this tract results in the formation of Thyroglossal Duct Cyst (TGDC).

A TGDC is the most common anomaly in thyroid development^[2]. Seventy percent of midline neck masses during childhood and 7% in adults are TGDCs^[3]. Approximately 1% of TGDC are found to contain malignancy, with around 270 reported cases so far^[5]. The most common primary thyroglossal duct cyst carcinoma is papillary carcinoma (75–80%), but other tumor types of primary tumor do occur, such as mixed papillary-follicular carcinomas (7%), squamous cell carcinoma (5%), follicular carcinoma (1.7%); Hurtle cell carcinoma and anaplastic carcinoma (0.9%) have also been reported^[6]. We report three such cases of papillary thyroid carcinoma arising from a TGDC.

METHODS

We performed a retrospective analysis of records to identify patients treated for TGDC carcinoma at our institute of last 15 years. Criteria for inclusion in this study were presence of TGDC carcinoma and primary treatment for this disease being conducted at our institution. Patient demographics, clinical features, preoperative diagnostic tests, the extent of surgery, and adjuvant therapy after surgery for each patient were reviewed. Final pathologic details following definitive therapy were recorded. The patients were followed up for recurrence by yearly analysis of serum thyroglobulin.

Case Description**Case 1**

A 45-year-old man with ongoing treatment for schizophrenia on Flupenthixol and clonazepam for 10 years, presented with a midline neck swelling for the last three months. Ultrasound of the neck showed 4 x 3 cm hypochoic lesion with no internal vascularity suggestive of TGDC. The patient underwent Sistrunk procedure. Gross examination of the specimen revealed partially cystic mass with papillary projection and partially solid. Microscopic examination was suggestive papillary carcinoma of Thyroglossal duct. Total thyroidectomy was done. Thyroid gland showed no malignancy in HPE. The patient subsequently underwent Radioactive Iodine ablation (RIA).

Case 2

A 65-year man presented with swelling in the anterior neck that had been progressively increasing in size for the past 3 months. He had no other complains with no significant medical history. Ultrasound of the neck revealed a 3cm x 2cm TGDC. The patient underwent Sistrunk

procedure and histopathological examination of the excised cyst revealed papillary thyroid carcinoma. The patient further underwent total thyroidectomy without neck dissection and radioactive iodine ablation. The patient is disease free till 8 years of follow up.

Case 3

A 32-year woman presented with 3cm x 2cm swelling in the neck for the past 2 years. She had no other complaint. On clinical examination thyroid and cervical lymph nodes were non palpable. Ultrasound confirmed the diagnosis of TGDC. She underwent Sistrunk procedure and histopathology showed papillary carcinoma of thyroid. She further underwent total thyroidectomy. Final histopathology of thyroid showed a benign cystic lesion of 1cm x 2cm. She underwent radioactive iodine ablation and she is disease free on follow up for 4 years.

Table 1: Patient Demographics And Presentation

	Age	Sex	Comorbidity	duration	size	Comp ressive sympt oms	sit e	Lym ph node mets	Follow up
Case 1	45	M	schizop hrenia	3 months	4 x 3cm	None	IH	No	1 year
Case 2	32	F	None	2 years	3 x 2cm	None	IH	No	4 year
Case 3	65	M	None	3 months	3x 2cm	None	IH	No	8 year

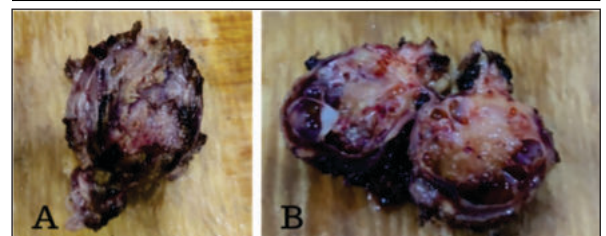


Figure 1: Gross(A) and cut section (B) photographs of the thyroglossal cyst. Cut section showed complex cyst with solid and cystic components.

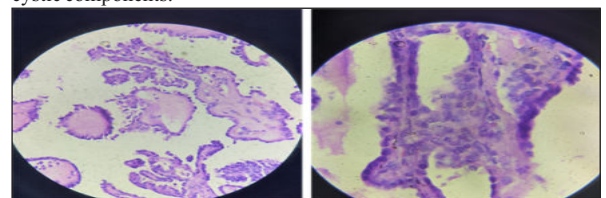


Figure 2: Microphotograph Showing Papillary Tumor Arising From

TGDC wall having follicular and papillary pattern

DISCUSSION

The TGDC carcinoma was first described by Bretano in 1911 and Uchermann in 1915^[6]. The appearance of a carcinoma in thyroglossal duct cyst is rare with incidence is 1%. TGDC typically presents in a pediatric population, and most cases present before age 30 years; contrastingly, TGDC carcinoma tends to present later in life, with a mean age of 39.5 years. Furthermore, thyroid cancers are 2.9 times more common in females than males, this predilection also extends to TGDC carcinoma.^[5]

There are two theories to explain the origin of TGDC carcinoma. Firstly, the de novo theory is based on the fact that in 62% of cases, ectopic thyroid tissue can be identified histopathologically, and this is supported by the absence of a medullary carcinoma in the TGD as it arises from parafollicular cells. The second is the metastatic theory which suggests that thyroglossal duct carcinoma is metastatic from an occult primary thyroid gland, as papillary carcinoma is multifocal in nature.^[7,8]

The usual symptom of TGDC is a midline neck swelling which moves with deglutition and protrusion of tongue. Malignancy should be suspected if the thyroglossal duct cyst is hard, irregular, fixed, rapidly growing, and associated with palpable neck lymph nodes^[9,10]

The definitive management of thyroglossal duct cyst carcinoma remains controversial, in regard to the treatment strategy for the thyroid gland. While some authors consider Sistrunk's procedure to be curative in most cases, others recommend that total thyroidectomy should be performed in case of thyroglossal duct cyst carcinoma, due to the high incidence of associated papillary or mixed carcinomas in the thyroid gland. According to Balalaa et al., total thyroidectomy is indicated, whether or not the thyroid is involved clinically or radiologically, as can facilitate the detection of metastases, and the risk of injury to the recurrent laryngeal nerve or parathyroid glands in skilled hands is very uncommon^[11]. The rationale for adding thyroid resection to every patient with a thyroglossal duct cyst carcinoma is based on three main considerations, the presence of concomitant thyroid malignancy in the main thyroid gland, the utilization of radioactive iodine as an adjuvant treatment, and the role of serum thyroglobulin, as a follow-up marker for thyroid cancer.s

Some authors advocate a surgical approach that is customized according to the patient and is based on risk group stratification. Accordingly, Sistrunk's procedure is performed as the only procedure in case of clinically and radiologically normal thyroid gland in low-risk patients with low-risk tumors, for patients who are age <45 years, who have no history of radiation exposure, a tumor size <4.0 cm, no soft tissue invasion, no distant or lymphatic metastases, and the absence of aggressive tumor histology.

Tharmabala and colleagues propose a risk staging strategy into three classifications: for low-risk, observe and wait; for moderate risk, total thyroidectomy, hormonal suppressing treatment, and radioactive iodine; for the high-risk group, lymph node dissection in addition to the other treatments^[12].

In this case, the authors chose Sistrunk's procedure and total thyroidectomy followed by radioactive iodine ablation as a treatment strategy. All three patients were from disease free till the last follow up.

CONCLUSION

We report 3 cases of papillary thyroid carcinoma arising from a TGDC. All three patients underwent Sistrunk operation first and subsequent total thyroidectomy followed by radioactive iodine ablation. All three patients are stable and disease-free since the time of surgery up until the last follow-up. Although the thyroid gland was found to normal in all cases, we recommend total thyroidectomy in all patients presenting with TGDC carcinoma. It is justified by high incidence of associated papillary carcinomas of the thyroid, RAI therapy could be given and better follow up by serum thyroglobulin. A radical therapeutic attitude results in safer patient outcome.

DECLARATIONS

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Ethics: Written informed consent was obtained from the patient for publication of this case report and accompanying images.

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