

Original Article



Thyroid Cancer Arising from Thyroglossal Duct Cyst

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Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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ABSTRACT

Purpose: Thyroglossal duct cyst is the most common congenital anomaly of the neck, and malignancy arising from a thyroglossal duct cyst occurs only in approximately 1% of patients. Ultrasound examination and fine-needle aspiration should be performed to make an accurate diagnosis before surgery. Sistrunk operation is known as a surgical treatment for thyroglossal duct cyst. We describe the clinical characteristics of thyroglossal duct cyst carcinoma in eight cases and the treatment methods for each.

Methods: This is a retrospective study of thyroglossal duct cyst detected in patients undergoing surgery at two separate tertiary medical institutions in Korea from July 2006 to August 2020.

Results: Among 346 patients with thyroglossal duct cyst, eight were diagnosed with thyroglossal duct cyst carcinoma. In four out of these eight patients, cysts and thyroid nodules were observed, and thyroidectomy and Sistrunk operation were performed together. Modified radical neck dissection was performed only in two patients with confirmed lymph node metastasis before surgery. All four patients who underwent total thyroidectomy and Sistrunk operation received radioactive iodine treatment and did not receive radiation therapy. None of the patients had distant metastasis.

Conclusion: Thyroglossal duct cyst carcinoma is rare, and an initial evaluation is critical in determining the extent of surgery. Fine needle aspiration should be performed in patients who are clinically diagnosed with thyroglossal duct cyst, and a thyroid ultrasound scan should also be performed.

Keywords: Thyroglossal duct cyst; Thyroid cancer; Fine-needle aspiration

INTRODUCTION

Thyroglossal duct cysts (TGDCs) are the most common congenital anomaly of thyroid development that occur in infancy and childhood and mostly present as midline neck masses (1). In general, the thyroid gland develops as a midline endodermal invagination in the foramen caecum during the third week of fetal development. At 7–8 weeks of intrauterine development, the thyroid gland reaches its normal position under the thyroid cartilage and descends through the thyroglossal duct (TGD). The TGD disappears physiologically at 10 weeks of fetal development. The remnants of the TGD will remain as cysts, ducts, or ectopic tissue (2,3).

Although the prevalence of TGDC is approximately 7%, malignancy arising from a TGDC occurs only in approximately 1% of patients with TGDC, and papillary thyroid cancer (PTC) is the most common type (4). This malignant tumor was first reported by Brentano in 1911 (5). When a clinical diagnosis of a TGDC is made, it should be closely examined by using ultrasound scan, computed tomography (CT) scan, and fine-needle aspiration (FNA) test. Examinations for TGDC carcinoma before surgery are performed to determine the optimal surgical extent and to plan radioactive iodine (RAI) treatment (6).

In general, the Sistrunk operation is the surgical procedure of choice for TGDC. This method of removing TGDC, the central part of the hyoid bone, and the tissues around the thyroglossal tract is the surgery of choice for low-risk patients. However, high-risk patients need additional surgical resection beyond this range (2,7).

In this report, a series of a rare case of papillary carcinoma arising from a TGDC is presented, its clinical characteristics are investigated, and an appropriate treatment method is sought along with a literature review.

MATERIALS AND METHODS

A retrospective chart review study was conducted at two separate tertiary medical institutions in Korea. The clinical records were reviewed between July 2006 and August 2020, and patients with a histological diagnosis of carcinoma of TGDC were identified. The patients' demographics, such as age, sex, surgical method, surgical extent, preoperative FNA results, site of TGDC, type of carcinoma, clinical course, postoperative radio-iodine ablation, and follow-up, were analyzed. This study was approved by the Institutional Review Board of Gangnam Severance Hospital (approval No. 3-2020-0415).

RESULTS

During the research period, 346 patients underwent TGDC surgery in two separate tertiary care centers; eight patients were diagnosed with papillary carcinoma arising from TGDC and underwent surgery. The female-to-male ratio was 7:1. The average age at the time of surgery was 42.25 years (range, 26–68 years). In four patients, the masses were noticeable or palpable, while the masses in the other four patients were accidentally discovered upon ultrasound examination of the thyroid gland. Two patients underwent FNA due to TGDC prior to surgery, one of which was reported as PTC and the other as inappropriate. Four patients (50%) had a coexisting thyroid carcinoma, all of which were PTC. The average tumor size was 1.6 mm (range, 0.6–2.5 mm). Four patients received RAI treatment and no recurrence was reported. The average follow-up period was 42 months (range, 1–94 months) (**Tables 1 and 2**).

All patients were included in an imaging study followed by surgery. In one patient, only the thyroid gland was imaged, and no cyst was observed. Of the eight patients, three had normal thyroid glands, and the five patients had nodules. According to histopathological findings, out of the five patients who underwent thyroidectomy, four patients had papillary thyroid cancer and one patient had noninvasive follicular thyroid neoplasm with papillary-like nuclear features (NIFTP).

Carcinoma Arising from TGDC and Sistrunk Operation

Table 1. The image finding of TGDC, the thyroid gland, and LNs

No	Operation	Extent of surgery	Age	Sex	Imaging finding		
					TGDC	Thyroid	LNs
1	MRND, left	BTT with CCND Lt. MRND Shaving off of trachea & RLN	52	M	Cyst not imaged	Rt. thyroid with 1.5 cm lesion at mid portion Lt. thyroid with 1.4 cm lesion (extrathyroidal invasion) at mid portion close to the esophagus and the trachea Lt. isthmus with 0.4 cm lesion	0.8 cm LN at Lt. submental area 0.5 cm, 0.7 cm, 1.0 cm LNs at Lt. level IV
2	Sistrunk operation	Sistrunk operation BTT with CCND	29	F	3 cm sized low-density cystic mass with calcification in anterior neck	Rt. thyroid with 1.0 cm lesion at upper portion	No nodes
3	Excision of thyroglossal duct cyst	Sistrunk operation BTT with CCND	26	F	3 cm cystic lesion with solid component in midline of anterior neck	Lt. thyroid with 1.1 cm lesion at mid portion	No nodes
4	Sistrunk operation	Sistrunk operation BTT with CCND Left. MRND	53	F	1.7 cm cystic lesion in Rt. midline anterior neck	Lt. isthmus with 1.0 cm lesion Lt. thyroid with 0.3 cm at upper portion	1.1 cm metastatic LN at Lt. level III
5	Sistrunk operation	Sistrunk operation	28	F	1.4 cm cystic lesion with calcification, solid component	Normal thyroid	No nodes
6	Sistrunk operation	Sistrunk operation	48	F	1.7 cm cystic lesion with calcification	Normal thyroid	No nodes
7	Sistrunk operation	Sistrunk operation	34	F	1.5 cm ovoid hypoechoic Cystic lesion	Normal thyroid	No nodes
8	Sistrunk operation	Sistrunk operation Rt. thyroidectomy with CCND	68	F	1.1 cm cystic lesion with calcification	Rt. thyroid with 2.2 cm lesion at mid portion Lt. thyroid with multiple benign nodules	No nodes

PTC = papillary thyroid carcinoma; CCND = central compartment lymph node dissection; TGDC = thyroglossal duct cyst; BTT = bilateral total thyroidectomy; MRND = modified radical neck dissection; RLN = recurrent laryngeal nerve; LN = lymph node.

Table 2. Clinicopathologic features of the patients in this case series

No	Extent of surgery	Location	Symptoms	FNA		Tumor size (cm)	TGDC pathology	Thyroid pathology	RAI	RTx	Recurrence	Follow-up (mon)
				Cyst	Thyroid							
1	BTT with CCND Lt. MRND Shaving off of trachea & RLN	Central neck	Exam	No	Rt.: VI, Lt.: V	1.2	PTC arising in thyroglossal duct cyst	Rt. thyroid: PTC (1.3 cm) Lt. thyroid: PTC (1.1 cm) Metastatic carcinoma in 4 out of 34 lymph nodes	Yes	No	No	94
2	Sistrunk operation BTT with CCND	Central neck	Mass	No	Rt.: VI	2.2	PTC arising in thyroglossal duct cyst	Rt. thyroid: PTC (1.0 cm) Lt. thyroid: Free	Yes	No	No	80
3	Sistrunk operation BTT with CCND	Central neck	Mass	No	Lt.: VI	2.5	Thyroglossal duct cyst containing PTC	Rt. thyroid: Free Lt. thyroid: PTC (1.3 cm)	Yes	No	No	39
4	Sistrunk operation BTT with CCND Lt. MRND	Central neck	Exam	No	Lt.: VI	1.5	PTC	Rt. thyroid: Free Lt. thyroid: PTC (1.0 cm, 0.3 cm)	Yes	No	No	25
5	Sistrunk operation	Central neck	Mass	Cell paucity	No	0.7	PTC arising in thyroglossal duct cyst	No lesion	No	No	No	8
6	Sistrunk operation	Central neck	Mass	PTC	No	1.8	PTC arising in thyroglossal duct cyst	No lesion	No	No	No	28
7	Sistrunk operation	Central neck	Exam	No	No	0.7	PTC	No lesion	No	No	No	57
8	Sistrunk operation Rt. thyroidectomy with CCND	Central neck	Exam	No	Rt.: II, Lt.: II	0.6	PTC arising in thyroglossal duct cyst	Rt. thyroid: NIFTP (1.5 cm)	No	No	No	1

FNA = fine-needle aspiration; TGDC = thyroglossal duct cyst; RAI = radioactive iodine; RTx = radiation therapy; BTT = bilateral total thyroidectomy; CCND = central compartment lymph node dissection; MRND = modified radical neck dissection; RLN = recurrent laryngeal nerve; PTC = papillary thyroid cancer; NIFTP = noninvasive follicular thyroid neoplasm with papillary-like nuclear features.

Among all cases, only Sistrunk operation was performed in three cases. In four cases, cystic lesion and thyroid nodules were identified on ultrasound images, and thyroidectomy and Sistrunk operation were performed together. In the first case, thyroid nodules and lateral neck lymph node were seen in the ultrasound image, but cystic lesion was not visible. Furthermore, lateral neck node metastasis was confirmed by FNA test before surgery. Thus, total thyroidectomy and modified radical neck dissection (MRND) were performed. In another patient, lateral neck node metastasis was also confirmed and MRND was performed.

Four patients who underwent total thyroidectomy and Sistrunk operation did not have malignant disease or recurrence between 25 and 94 months (average 60 months). All patients received RAI treatment, and none received radiation therapy (RTx). No patients had distant metastasis.

DISCUSSION

TGDC is the most common midline neck cyst and is a congenital anomaly. It accounts for 70% of congenital neck masses (8). In general, TGDC is benign but has been reported to undergo malignant transformation in approximately 1% of cases (9,10). Weiss and Orlich (9) reported that thyroid cysts and the thyroid gland were associated with thyroid papillary cancer in 11.4% of 115 cases, and Chrisoulidou et al. (11) reported the same in 66% of six cases. In our study, four (50%) out of eight patients with TGDC had papillary thyroid cancer. Carcinoma arising from TGDC has an excellent prognosis and a metastasis rate of less than 2% (12).

In terms of sex distribution at the two tertiary institutions in our series, women outnumbered men. Türkyilmaz et al. (13) also reported a high distribution among women. However, some studies have reported that TGDC has the same distribution between men and women (14). In our study, seven out of eight patients with carcinomas arising from TGDC were women. This distribution might be due to geographic differences in Korea, where thyroid papillary cancer more commonly occurs in women than in men.

The main cause of patients visiting the hospital for TGDC is a mass that is visible, palpable, or infected. In cases where a mass is present, it is usually solitary, midline, and moves with swallowing, and when infected, it causes pain, fever, dysphagia, dyspnea, or could lead to a fistula (15). Among the patients we analyzed, four patients (50%) had been treated because of the presence of a palpable mass.

A detailed clinical history must be obtained to make an accurate diagnosis before surgery. Making a diagnosis before surgery is difficult; therefore, imaging tests, such as ultrasound, neck CT, and magnetic resonance imaging, which help to determine whether the thyroid tissue is normal, are mainly performed. In general, an imaging test cannot confirm a malignant disease; therefore, an FNA test is performed as well. However, due to the hypocellularity of samples, only 66% of cases show accurate results (16). In our study, FNA test was performed in only two of the seven cases where a cystic lesion was detected upon preoperative imaging examination. A diagnosis of PTC was made in one case, and in the other case, a diagnosis could not be made due to hypocellularity.

Much controversy exists over the extent of surgical treatment for carcinomas arising from TGDC. Some consider a Sistrunk operation as sufficient, while others recommend both a Sistrunk operation and a total thyroidectomy (12,14,17). Sistrunk operation was introduced

in 1920 by Sistrunk as a procedure for removing structures that make up a cyst, its tract, the middle part of the hyoid bone, and the base of the tongue (18). The recurrence rate was reduced to 3%–6% compared with that of the previous techniques in which a simple incision is made to drain the cyst and remove the middle part of the hyoid bone along with cyst. Currently, the Sistrunk operation is the standard surgical technique that is performed (13,19). Sistrunk operation is a safe procedure with few complications. The major complications (abscess, hematoma, nerve palsies etc.) are known to be extremely rare (15).

Most surgeons determine surgical strategies based on risk stratification and perform Sistrunk operation in clinically low risk patients with normal thyroid glands. Total thyroidectomy is also performed in high-risk patients and in patients with a positive thyroid FNA test. This is because performing total thyroidectomy generally is a way to cause lifelong hormone replacement therapy and possible complications in low-risk patients. In a review study involving more than five cases, the average rate of Sistrunk operation and total thyroidectomy performed together was 46.3% (20).

In our study, 50% (four out of eight cases) were had thyroid cancer. Sistrunk operation and total thyroidectomy were performed in three cases, right total thyroidectomy and Sistrunk operation were performed together in one case, and only Sistrunk operation was performed in three cases. In two cases, modified radical neck dissection was performed because a metastasis node was present in the lateral part of the neck.

CONCLUSION

Carcinomas arising from TGDCs are rare and were reported in only eight cases out of the 346 cases we reviewed. An initial evaluation, which is important in determining the extent of surgery, should be properly performed because of lack of a clear guideline on the extent of surgery or the need for adjuvant therapy, such as RAI or RTx. Therefore, in order to make accurate preoperative diagnosis of malignancies and to determine the extent of surgery, TGDC, thyroid ultrasound scans and FNA tests should be performed for patients who have been clinical diagnosed with TGDC.

The analysis of the results of the cases reviewed here suggests that the surgical method and conservative management suitable for each patient should be performed according to the preoperative examination results of TGDC, and continuous observation and examination are required.

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