Papillary carcinoma arising from thyroglossal duct cyst with thyroid and lateral neck metastasis

Song-I Yang, Kwang-Kuk Park, Jeung-Hoon Kim*

Department of Surgery, Kosin University College of Medicine, Busan, Republic of Korea

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A B S T R A C T

INTRODUCTION: Thyroglossal duct carcinomas (TGDC) are rare, with approximately 274 reported cases since the first report in 1915. The prevalence of carcinomas in surgically removed thyroglossal duct cyst (TG) is less than 1%. The usual recommended treatment for this condition is the Sistrunk operation, but controversies remain regarding the need for total or partial thyroidectomy.

PRESENTATION OF CASE: A 28-year-old woman was admitted to our hospital with the symptoms of painless midline neck swelling and growing mass. A preoperative computed tomography (CT) showed a 4 cm sized heterogeneous mass at the infrahyoid anterior neck. Ultrasonography of the neck additionally showed suspicious metastatic lymph node at right level VI, both level VI. The patient underwent a Sistrunk operation. The frozen section revealed papillary carcinoma arising from TGDC and also revealed metastatic papillary carcinoma in the right thyroid, at right level III and level VI. Total thyroidectomy, right modified radical neck dissection and central neck dissection were performed. The thyroid gland and TGDC were confirmed papillary carcinoma. The dissected neck lymph nodes revealed metastatic papillary carcinoma.

DISCUSSION: The usual recommended treatment for TGDC is the Sistrunk procedure. There is controversy regarding whether total or partial thyroidectomy should be performed.

CONCLUSION: Physicians should be aware of extended operation, including thyroidectomy and/or neck node dissection for TGDC with metastatic lesion of thyroid and neck node.

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1. Introduction

Thyroglossal duct cyst (TDC) results from a developmental anomaly of the thyroid gland and accounts for 70% of congenital neck masses.1 The majority of patients present within the first two decades of life, but nearly a third of cases may manifest in young adulthood. (i.e. >20 years of age)2 TDC usually manifests as an enlarging painless neck mass in children or young adults, and in most circumstances diagnosis can be made by history and physical examination.3 However, thyroglossal duct carcinoma (TGDC) may be clinically indistinguishable from benign TGD, and the diagnosis in most cases is incidental after surgical resection. Fine needle aspiration cytology under ultrasound guidance may enhance preoperative diagnosis. The standard treatment is a Sistrunk procedure, but there is controversy regarding whether total or partial thyroidectomy should be performed.

2. Presentation of case

A 28-year-old woman presented with a painless midline neck swelling which had been progressively increasing in size for one year. On examination, an oval-shaped mass located in the upper part of the anterior neck was found. The mass moved with swallowing as well as on tongue protrusion. Tongue mobility was intact and no abnormalities were observed at the base of the tongue. Thyroid function test was within normal range and other baseline blood investigations were also normal. A clinical diagnosis of thyroglossal cyst was made. Ultrasonography of the neck revealed a cystic mass with internal calcified solid portion at anterior neck, measuring 3.4 cm × 2.8 cm, and it did not communicate with the thyroid gland. Ultrasonography of the neck additionally revealed suspicious metastatic lymph node at right level VI, both level VI. Computed tomography (CT) showed a 4 cm sized heterogeneous mass at the infrahyoid anterior neck, (Fig. 1) However, a clear demarcation was seen between the mass and the thyroid gland, and there were regional lymphadenopathy on right level III. The patient underwent Sistrunk operation. The frozen section revealed papillary carcinoma arising in a TGDC (Fig. 2) and also revealed metastatic papillary carcinoma in a right thyroid, right level III and level VI. Therefore, total thyroidectomy, central neck dissection and right modified radical neck dissection were

*Corresponding author at: Department of Surgery, Kosin University College of Medicine, 34 Amnam-dong, Seo-gu, Busan 602-703, Republic of Korea.
Tel.: +82 51 990 6462; fax: +82 51 246 6093.
E-mail addresses: tonybina@daum.net (S.-I. Yang), minga000@daum.net (K.-K. Park), kkparkys2@gmail.com (J.-H. Kim).

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performed. Histopathological examination identified papillary carcinoma that was confined to the cyst (Fig. 3). Non-neoplastic cystic spaces were partially lined by flattened epithelial cells. Pathologic analysis showed a 7 mm papillary carcinoma in the left lobe of the thyroid gland that lacked extrathyroidal extension (Fig. 4). Postoperative recovery was uneventful. Follow-up at 6 and 36 months after surgery did not reveal any clinical evidence of tumor recurrence, and serum thyroglobulin levels were within normal ranges.

3. Discussion

Thyroglossal duct cysts develop from persistence of the mid portion of the thyroglossal duct which is an embryonic structure that traces the path of the descent of the thyroid gland. The duct is normally obliterated at around the 8th–10th week of gestation, but if the duct fails to involute completely, the remaining epithelial tissue can develop a TGDC. Previous studies have suggested that this failure to involute occurs in approximately 7% of the population.4 Typically, TDC manifest as an enlarging, painless,
midline neck mass in children or young adults that moves upward with tongue protrusion.\textsuperscript{2,3,5} Rarely, the cyst may localize to the floor of the mouth.\textsuperscript{6,7} Malignant transformation of TDC to TGDC is uncommon, and the incidence of TGDC varies from 0.7% to 1% of all cases of TDC.\textsuperscript{8,9} According to Widstrom et al.,\textsuperscript{10} the criteria for diagnosis of primary carcinoma of the thyroglossal duct includes the following: histological identification of TGDC by demonstration (i.e. epithelial lining of ducts with normal thyroid follicles within walls of the cysts), normal thyroid tissue adjacent to the tumor, and histopathological examination of the thyroid gland showing no sign of primary carcinoma.\textsuperscript{11} The histologic findings of thyroglossal duct carcinoma are most commonly papillary carcinoma (75–80%), but other thyroid tumors such as follicular, Hurthle cell, and mixed papillary-follicular carcinomas have been reported.\textsuperscript{12} Imaging diagnostic techniques, including ultrasound, scintigraphy and CT, are usually unable to preoperatively diagnose malignant disease\textsuperscript{11} and fine needle aspiration yields a correct result in only 66% of the cases.\textsuperscript{13} Nearly 4% of thyroglossal duct carcinomas were found to be locally invasive, while 11% were found to include metastasis to cervical lymph nodes. Contemporaneous history of thyroid carcinoma was described in about 20% of thyroglossal duct carcinomas.\textsuperscript{14} Reported treatment options for cancers arising in TDC have included mass resection, Sistrunk’s procedure (a more involved resection removing the cyst, the body of the hyoid bone, and a cone of the base of the tongue muscle up to the foramen caecum), or a Sistrunk’s procedure involving total (or subtotal) thyroidec- tomy. Sistrunk’s procedure is now generally recommended for surgical treatment of benign thyroglossal duct cysts in children and adults.\textsuperscript{15} This procedure ensures complete resection of the lesion, which can often be found extending around the body of the hyoid bone and up to the base of the tongue. The lesion follows the embryonic trajectory of the endodermal pouch of the forert that gives rise to the thyroid gland (i.e. the thyroglossal duct).\textsuperscript{4,16} The Sistrunk’s procedure can thus be considered as a first-line treatment for papillary carcinoma arising in TDCs. In a retrospective study of 57 cases, Patel et al. concluded that the addition of total thyroidecotomy to the Sistrunk operation did not have a significant impact on recurrence and survival.\textsuperscript{14} But some authors propose a more radical approach which involves local excision of the tumor in addition to total thyroidecotomy because of occurrence of multicentric papillary carcinomas throughout the entire thyroid gland.\textsuperscript{17,18} A previous study proposed an algorithm for treatment of papillary carcinoma in TDC which involved a simple Sistrunk procedure in patients less than 45 years of age with tumors less than 1.5 cm that are confined to the cyst and who show an ultrasonographically normal thyroid gland and no suspicious lymph nodes.\textsuperscript{19} Total thyroidecotomy with neck dissection performed only when lymph node metastases are found on ultrasound or during surgery followed by radioiodine is recommended for those not fitting the criteria detailed above. A previous study reported many similarities between papillary carcinoma in TDC and general papillary carcinoma, suggesting that TDC carcinoma is similar to other papillary carcinomas in terms of its multifocality, lymph node metastasis, and prognosis.\textsuperscript{20} These studies recommend the Sistrunk’s procedure to be sufficient for carcinoma in TDC measuring up to 1 cm, with total thyroidecotomy, and central compartment neck dissection for larger tumors, keeping in mind that a lateral neck dissection may be needed secondarily in some cases, for these midline tumors.\textsuperscript{20} In the present case, the mass was 3.5 cm and nodal involvement of the tumor was seen on preoperative radiological and intraoperative histopathological studies, which indicated papillary carcinoma. Therefore, a Sistrunk’s operation, total thyroidecotomy, right modified radical neck dissection, and central compartment neck dissection were performed in this case. The prognosis for papillary TGDC carcinoma is excellent, with occurrence of metastatic lesions occurring in less than 2% of cases.\textsuperscript{9} Follow-up consists of physical examination, ultrasound of the surgical region and thyroid, and total body scintigraphy.

### 4. Conclusion

Because of the rarity of TGDC, this diagnosis may be missed, drastically affecting the appropriateness of the treatment provided. Surgeons should be aware of TGDC in surgical planning and postoperative treatment and should include this pathology in differential diagnosis of anterior midline neck mass. Regular follow-up to detect any recurrence in the thyroid gland is essential, particularly in cases where the thyroid gland is not resected.

### Conflict of interest statement

The authors have no conflict of interests to declare.

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Consent from patient available.

### Authors’ contributions

SIY collected the information, researched the literature, and wrote the article. KKP and JHK helped with the literature research and in preparing the manuscript. JHK helped in the literature research and edited the final version of manuscript. All authors read and approved the final manuscript.

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