

# Primary Papillary Carcinoma Arising in a Thyroglossal Duct Cyst

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We report a case of papillary carcinoma arising in a thyroglossal duct cyst, presenting with an anterior neck mass of a 31-year-old woman. The tumor was judged to be a primary lesion on the basis of intraoperative examination of the thyroid and pathologic findings of the mass. One year later, a small nodular mass in the left thyroid gland and lymph node enlargement of the right cervical lymph node were noted by follow-up imaging studies. Total thyroidectomy, right modified radical neck dissection and central neck dissection were performed. The thyroid gland revealed nodular hyperplasia without evidence of malignancy. On the other hand, the dissected neck lymph nodes revealed metastatic papillary carcinoma. Taken together, these findings suggested the tumor was a primary papillary carcinoma arising in the thyroglossal duct cyst.

**Key Words:** Thyroglossal duct cyst, papillary carcinoma

## INTRODUCTION

Persistent thyroglossal duct is reported in 7% of adults,<sup>1</sup> and the thyroglossal duct cyst is the most common disease among all congenital masses of the neck.<sup>2</sup> About 1% of thyroglossal duct cysts are histologically malignant, and 80% of these cases are papillary carcinomas of the thyroid gland type.<sup>3</sup> Metastasis to the regional lymph nodes has been reported in 7.7% to 12.9% of these malignant cases,<sup>4-6</sup> much less frequently than primary papillary carcinomas of the thyroid gland. The prognosis is good except for the anaplastic type.<sup>3</sup> In

this report, a 31-year-old female with papillary carcinoma arising in a thyroglossal duct cyst is described with special regard to the histopathologic findings.

## CASE REPORT

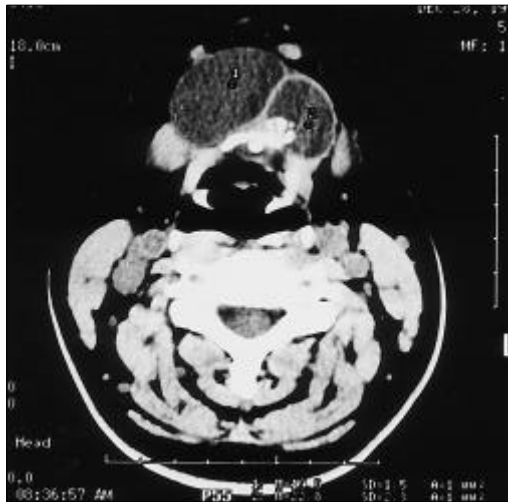
A 31-year-old woman visited our hospital with the chief complaint of an anterior neck mass in the median region of the neck. She had first noticed the mass one and a half year prior to the consultation. The mass was painless, had gradually been increasing in size, was located immediately above the hyoid bone, was 6.0 × 6.0 cm in size, elastic, soft, and had a smooth surface. No abnormalities were noted by palpation of the thyroid, nor were any cervical lymph nodes palpated. Computed tomography (CT) examination displayed no abnormality in the thyroid or cervical lymph nodes. Hematological, biochemical, and tumor marker tests revealed no abnormalities.

CT findings showing a multiseptated, large cystic mass with calcification and enhancing solid areas in part of the interior of the mass (Fig. 1), suggested the possibility of a thyroglossal duct cyst containing malignant or benign tumor tissue. Sistrunk operation was performed. The mass was a multilocular cystic mass, measuring 6.0 × 5.5 cm with a segment of hyoid bone. Sectioning exposed a multiloculated cyst which was filled with yellow mucinous material. The inner surface showed a few papillary projections, the largest one of which measured 0.8 cm in diameter. The remaining surface was grayish-brown, glistening, trabeculated and sclerotic. Histologic examination confirmed that the mass was a thyroglossal duct cyst

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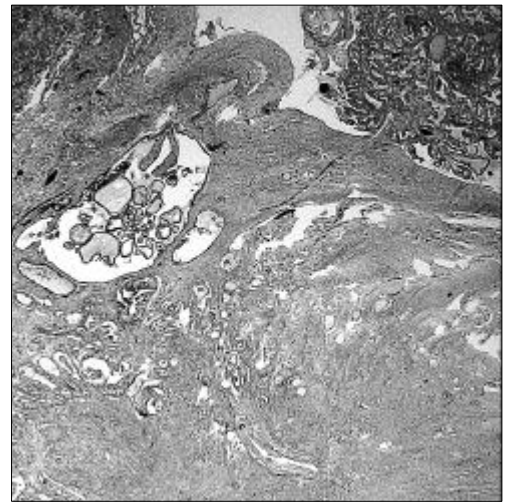
**Fig. 1.** CT scan of the neck at the level of hyoid showing a multiseptated large cystic mass with amorphous calcification in the interior and a solid component in the upper posterior portion.

and that a papillary carcinoma was present within the lumen, in the wall, and in the surrounding tissue of the cyst, and that the tumor was associated with marked sclerosis (Fig. 2 and 3).

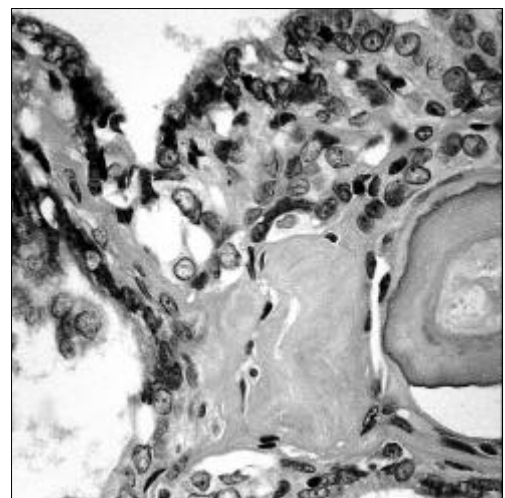
One year later, a small nodular mass in the left thyroid gland and lymph node enlargement of the right cervical lymph node were noted by follow-up CT examination. Since the carcinoma might have either originated in the thyroglossal duct cyst or have metastasized from the thyroid gland, total thyroidectomy, right modified radical neck dissection and central neck dissection were carried out. The thyroid gland revealed nodular hyperplasia without evidence of malignancy. On the other hand, the dissected neck lymph nodes revealed metastatic papillary carcinoma of one lymph node among the level III cervical lymph nodes. Taken together, these findings suggested the tumor was a primary papillary carcinoma of the thyroid arising in the thyroglossal duct cyst.

## DISCUSSION

Malignant transformation is a rare complication of untreated thyroglossal duct cysts. The diagnosis is, however, often made postoperatively on histologic examination of the resected specimen. In one study the age at presentation ranged from 6



**Fig. 2.** Microscopic findings of the resected specimen around the cystic wall. Intracystic papillary projections and cuboidal epithelial cell lining of the cyst are observed. Tumor cells, accompanied by colloid formation, partly infiltrate into the surrounding tissues. Around the tumor, scar formation is observed (H&E,  $\times 12.5$ ).



**Fig. 3.** Microscopic findings of the tumor showing ductal structures with papillary proliferation and psammoma bodies. Tumor cells show optically clear, large, and irregular nuclei with prominent grooving and intranuclear inclusions. (H&E,  $\times 400$ ).

to 81 years, with an average of 39 years.<sup>4</sup> Women are affected more often than men.<sup>4,7</sup>

To confirm a diagnosis of thyroglossal duct cyst, the following criteria should be fulfilled: the cyst must be located in the median region of the neck; the cyst wall must be composed of cuboidal epithelial cells; and lymphatic tissues and normal

thyroid follicles must be present in the cystic wall.<sup>8</sup> The cyst observed in the present case fulfilled all of these criteria.

In our case, a papillary carcinoma was demonstrated within the lumen, in the wall and in the surrounding tissue of the cyst. Concerning the origin of the tumor, it may either have arisen from thyroid tissue present in the wall of the thyroglossal duct cyst, or have metastasized from the thyroid carcinoma. The findings of scar formation in the center of the papillary carcinoma, of part of the tumor infiltrated into the surrounding tissues, and of an absence of clinically observed tumors in the thyroid, collectively favored the former explanation for the tumor origin.

Approximately 215 cases of malignant tumors in thyroglossal duct cysts have been reported to date in the world literature since the first report in 1911. Most of these tumors arose from ectopic thyroid tissue within the cyst. There are two representative types of thyroglossal duct carcinomas, thyrogenic carcinoma and squamous cell carcinoma.<sup>9</sup> The former most likely arises from thyroembryonic remnants in the duct or cyst, and the latter from metaplastic cuboidal cells. The results in the literature show that papillary carcinoma is the most common type (80%), followed by "mixed" papillary-follicular carcinoma (8%) and squamous cell carcinoma (6%).<sup>3,5,9,10</sup> The remaining 6% include rare cases of Hurthle cell, follicular, and anaplastic carcinomas.<sup>11-14</sup> To our knowledge, there have been no reports of medullary carcinomas of the thyroid arising in the thyroglossal duct cyst. In patients with thyrogenic carcinomas, the possibility of metastasis from an occult primary thyroid tumor exists, and some physicians have suggested that a patent thyroglossal duct could be the metastatic route from the thyroid gland.<sup>15</sup> Nevertheless, if a papillary carcinoma is found during the histopathological examination of the thyroid gland, primary carcinoma of the thyroglossal duct should not be excluded. McConahey et al.<sup>16</sup> reported a 21% incidence of multiple foci for malignant tumors in patients with papillary carcinoma of the thyroid gland, while others have reported 18% to 75% incidences of multicentricity.<sup>8,17,18</sup> Thyroglossal duct carcinoma should also be distinguished from papillary carcinoma arising from the tip of the

pyramidal lobe; a distinction that must be based on both gross pathologic features and surgical findings.

Widstrom et al.<sup>19</sup> suggested two, histopathologic, diagnostic criteria for primary carcinoma of the thyroglossal cyst: the localization of the carcinoma to a clearly demonstrable thyroglossal duct and the absence of carcinoma on histopathologic examination of the thyroid gland. In our case the histopathologic examination of the resected thyroid was not conducted at the time of initial surgery for the following three reasons. Firstly, intraoperative examination of the thyroid showed no abnormalities either by gross observation or palpation. Secondly, the tumor was strongly suggested to have originated in the cyst by the findings of scar formation in the center of the tumor and tumor cells partly infiltrated into the surrounding tissues. These histopathologic findings are characteristic of primary papillary carcinoma of the thyroid arising in the thyroglossal duct cyst.<sup>7</sup> Lastly, this type of carcinoma implies good prognosis with only rare instances of metastasis that can be successfully managed by careful follow-up. However, in conjunction with the pathologic findings of the thyroid from the subsequent total thyroidectomy which revealed the absence of tumor, the features of our case exactly fulfilled the diagnostic criteria proposed by Widstrom et al.<sup>19</sup>

Weiss and Orlich<sup>5</sup> reviewed 115 cases up until 1991 and demonstrated that 11.4% of the patients with thyroglossal duct carcinoma had microscopic foci of carcinoma of the thyroid gland. On the basis of their case by case observations, they performed total thyroidectomy as an additional operation when the presence of papillary carcinoma had been pathologically demonstrated in the thyroglossal duct cyst, even when no abnormalities had been noted in the thyroid at the initial operation. They regarded the microscopic lesion in the thyroid as metastatic. However, Kojima et al.<sup>7</sup> insisted that if papillary carcinoma of the thyroid is located in the thyroglossal duct cyst, and if no abnormalities are noted in the thyroid either clinically or by intraoperative examination, then total thyroidectomy for the exclusion of metastasis from the thyroid is unnecessary, both diagnostically and clinically. They considered the pathologic confirmation that the tumor

has originated in the thyroglossal duct cyst to be sufficient. In conclusion, this type of carcinoma implies good prognosis and our opinion concurs with that of Kojima et al. in regarding total thyroidectomy to be unnecessary.

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