



Case Report

Primary Papillary Carcinoma of a Thyroglossal Duct Cyst

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Abstract

The incidence of papillary carcinoma arising in a thyroglossal duct cyst (TDC) is rare and occurs in about 1% of TDCs. Slightly more than 100 cases have been reported in the English literature. In most cases, the diagnosis is only established after excision of a clinically benign TDC. We report a case of papillary carcinoma arising in a TDC, presenting with an anterior neck mass in a 70-year-old man, who underwent a Sistrunk procedure for removal of the mass and an adherent lymph node. A second, more extensive operation was indicated based on pathologic findings after the initial dissection. Based on the histopathologic findings, the patient received postoperative I¹³¹ ablation treatment (120mCi). After 2 years, there was no evidence of disease recurrence. This type of carcinoma usually has a good prognosis, with only rare instances of metastasis, and can be successfully managed with surgery and careful follow-up. (*Tzu Chi Med J* 2008;20(2):150–153)

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1. Introduction

A thyroglossal duct cyst (TDC) is a congenital anomaly, generally appearing as an asymmetric midline neck mass. It is thought to originate from epithelial remnants of the thyroglossal tract (1). Malignant transformation is a rare complication of TDC (1%), and 80% of these cases are papillary carcinomas of the thyroid gland (2). Metastasis to the regional lymph nodes was reported in only 7.7% of these malignant cases, much less frequently than primary papillary carcinomas of the thyroid gland (1). Carcinoma is usually an incidental finding on histologic examination of a seemingly uncomplicated TDC.

To date, just over 100 cases of papillary carcinoma arising in TDCs have been reported in the English literature. We report an additional case and discuss appropriate management.

2. Case report

A 70-year-old man visited our hospital with the chief complaint of an anterior neck mass in the median submental region. He had first noticed the mass approximately 1 year prior to consultation. The mass was painless and gradually increased in size. The patient denied any voice change, dysphagia or pain. He had no



Fig. 1 — Contrast-enhanced computed tomography of the neck at the level of the hyoid, showing a multiloculated 4.0-cm cystic mass with an enhanced solid component and calcification in the interior of the mass.

history of surgery or radiation to the head and neck. Physical examination revealed a 6.0×4.0cm mass over the submental region in the anterior median area of the neck. It was elastic, movable, and nontender, and moved with swallowing. Examination of the ears, nose, nasopharynx, oral cavity, oropharynx and larynx showed no abnormal findings. The thyroid gland was of normal size with no palpable nodules. No cervical lymph nodes were palpable. Hematological and biochemical tests revealed no abnormalities.

Computed tomography (CT) of the neck showed a multiloculated, 4.0 cm cystic mass with an enhanced solid component, and calcification in a portion of the interior of the mass (Fig. 1). No abnormality was displayed in the thyroid gland. Some small internal jugular and submandibular lymph nodes < 1 cm in diameter were identified bilaterally. On the basis of the clinical and imaging findings, a TDC containing malignant or benign tumor tissue was suspected.

The patient underwent a Sistrunk procedure for removal of the mass and an adherent lymph node. Gross examination of the specimen showed a smooth cystic mass measuring 4.5×3×3 cm with a segment of hyoid bone attached. When cut, the mass exuded bloody fluid. The cystic mass was multiloculated with a papillary tumor within the cyst cavity and in the surrounding tissue of the cyst (Fig. 2). Histologic examination showed cystic spaces lined with stratified squamous epithelium. A papillary carcinoma characterized by ground-glass nuclei, with prominent grooving in a papillary pattern and psammoma bodies, was present within the lumen,

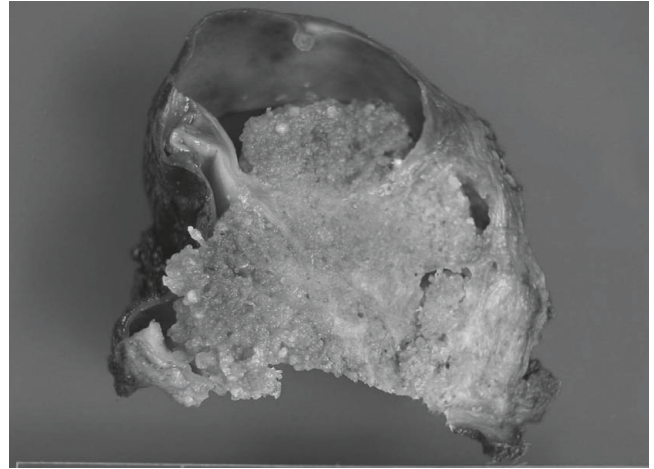


Fig. 2 — Multiloculated cystic mass with a papillary tumor within the cyst cavity and surrounding tissue of the cyst.

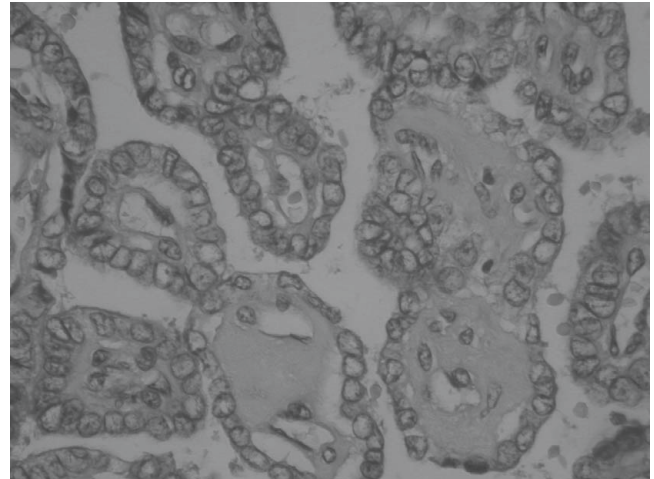


Fig. 3 — Papillae lined by tumor cells with ground glass nuclei, some showing prominent grooving (400×).

in the cystic wall, and in the surrounding tissue of the cyst (Fig. 3). The hyoid bone was also involved by the tumor. The microscopic diagnosis was papillary carcinoma arising in a TDC.

After the initial operation, serial studies of the thyroid gland were done. Thyroid function tests (T3, T4, TSH levels) were normal. Thyroid sonography revealed some isoechoic and hypoechoic nodules over the bilateral thyroid gland. An I¹³¹ thyroid scan showed decreased radioactivity in bilateral thyroid lobes, especially on the left side. Since the carcinoma may have either originated in the TDC or metastasized from the thyroid gland, a total thyroidectomy and central neck dissection were done. The thyroid gland revealed a nodular goiter, without evidence of malignancy. The dissected neck lymph nodes were negative for malignancy. After the

second operation, the patient received I¹³¹ ablation treatment and thyroxine suppression therapy.

The patient continues to receive follow-up in the outpatient department. After 2 years, there is no evidence of disease recurrence.

3. Discussion

TDCs are the most common non-odontogenic cysts in the neck, estimated to occur in 7% of the adult population (3). They are usually located in the midline or, less frequently, in the lateral anterior region of the neck (1). TDCs are most frequently seen between the thyroid gland and the hyoid bone (61%), followed in order of frequency by the suprahyoid (24%), suprasternal (13%) and intralingual (2%) regions (4).

The histopathologic features of TDC include cyst fluid that is mucoid to purulent, often containing cholesterol. The lining may be stratified squamous, cuboidal, or pseudostratified columnar epithelium. In the presence of inflammation, the lining may be replaced by dense fibrous or granulation tissue. Relatively normal thyroid tissue has been found in the walls in as many as 62% of cases (4,5). The cyst observed in the present case shared these features.

Generally, a TDC is benign, but about 1% degenerate into carcinoma (1). Since 1915, just over 100 cases of papillary carcinoma arising in TDCs have been reported in the English literature. Most of these tumors arose from ectopic thyroid tissue within the cyst, and presented the same histologic types as thyroid carcinoma (6,7). Reports 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%) (2,8). The remaining 6% include rare cases of Hürthle cell, follicular and anaplastic carcinoma (8). Medullary carcinomas of the thyroid arising in a TDC have not been reported.

Carcinoma presents more often in females than in males (1.5:1). Usually, carcinoma occurs later in life (mean, 39 years) than benign cysts, which present at an early age (mean, 5.5 years) (1); but both lesions can be found in all age groups.

In most cases, patients are asymptomatic. Development of a carcinoma in the wall of the TDC does not change the presentation (2). Page et al (6) stated that pain, hoarseness, sudden growth of a mass, weight loss, regional lymphadenopathy, and respiratory involvement suggest malignant changes, but these are unusual findings. In our case, there was only one suspicious condition present—that of a sudden enlargement of a neck mass.

CT findings are usually different in carcinomas arising in TDC. Samara et al studied 12 cases of carcinoma arising in the TDC (4). The most common finding was a solid nodule in the cyst (7 cases), and calcification

(2 cases), as was observed in our case. Other findings were an irregular margin (1 case), and a thick wall (1 case). In two of the 12 cases, there was no evidence of malignancy on CT.

Clinical examination, CT findings, and fine-needle aspiration may enhance the preoperative diagnosis, but in most reported cases, the carcinoma remained clinically occult until surgery. The diagnosis of carcinoma is usually made histologically after resection of a seemingly uncomplicated TDC, as evident in this case, with a papillary carcinoma demonstrated within the lumen, in the wall and in the surrounding tissue of the TDC.

Finding a papillary carcinoma in the TDC raises the question of origin. Tumors may arise from thyroid tissue present in the wall of the TDC, or metastasize from a thyroid carcinoma. Papillary carcinoma of the thyroid has been shown to metastasize without any detectable lesion in the thyroid gland itself (5). However, most authors believe that these tumors arise from ectopic thyroid tissue, which is found in the majority of benign TDCs (6,7). Primary carcinoma of the TDC should conform to criteria such as localization of the carcinoma to a clearly demonstrable TDC or tract, and absence of carcinoma after histopathologic examination of the thyroid gland (2). In this case, the histopathologic examination of the resected thyroid was not done after the first operation because intraoperative examination of the thyroid showed no abnormalities on either gross observation or palpation. Upon further histopathologic examination, the finding of tumor cells infiltrating into surrounding tissues suggested that the tumor may have originated from the thyroid gland. A thyroid ultrasound and thyroid scan were done, which showed bilateral thyroid nodules and hypofunctioning thyroid lobes, respectively. Based on these findings, total thyroidectomy was performed, and a nodular goiter was found on pathologic examination. Primary papillary carcinoma of the TDC was diagnosed.

A thyroidectomy is not routinely indicated unless there are palpable abnormalities in the gland or significant scintiscan findings. If there is a question of metastatic spread to the TDC or thyroid gland involvement, if the TDC carcinoma extends through the cyst wall, or if adequate follow-up of the patient is not likely, a total thyroidectomy may be indicated (5,7). In this patient, total thyroidectomy was done due to abnormal findings on thyroid ultrasound and I¹³¹ thyroid scan.

This type of carcinoma usually has a good prognosis with only rare instances of metastasis, and can be successfully managed by surgery and follow-up. Our patient's hyoid bone was invaded by the tumor and the margins were positive for tumor, so the patient received I¹³¹ ablation treatment (120 mCi), thyroxine suppression therapy, and careful follow-up in the outpatient

department. After 2 years, there is no evidence of disease recurrence.

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