Papillary Carcinoma in a Thyroglossal Duct Cyst

Avraham Ishay MD[1], Irit Elmalah MD[2] and Rafael Luboshitzky MD[1,3]

[1]Endocrine Institute and [2]Department of Pathology, HaEmek Medical Center, Afula, and [3]Rappaport Faculty of Medicine, Technion-Israel Institute of Technology, Haifa, Israel

Key words: papillary thyroid carcinoma, thyroglossal duct cysts, Sistrunk procedure

IMAJ 2008;10:312–313

Thyroglossal duct cysts are the most common form of congenital malformations in the neck. They are cysts of the epithelial remnants of the thyroglossal tract and present characteristically as a midline cervical mass at the level of the thyrohyoid membrane. Carcinomas arising in these remnants constitute a very uncommon clinicopathological entity (approximately 1% of TDCs). Papillary thyroid carcinoma is the most common cancer of the thyroid gland, comprising about 90% of all cases and representing also the most frequently encountered carcinoma in TDC. We present a patient with occult papillary thyroid carcinoma diagnosed after surgical resection of a thyroglossal cyst.

Patient Description

A 61 year old man presented with several years history of a slowly enlarging infrahyoid midline neck mass. Palpation revealed a 3 x 2 cm soft non-tender cystic mass that moved upward with protrusion of the tongue. There was no cervical lymphadenopathy. Thyroid function tests were normal. Preoperatively, the lesion was diagnosed as TDC and the mass was resected by the Sistrunk procedure, which consists of removing the cyst, the mid-portion of the hyoid bone and the entire thyroglossal tract [1]. Gross examination disclosed a 3 x 2.5 x 1.5 cm greyish-tan cystic tumor. Microscopic examination revealed a fibrous cystic structure with benign squamous epithelium focally lining the cyst. A focus of papillary carcinoma was seen within the cystic space. Areas of normal thyroid follicules were seen in the cyst wall [Figure].

Postoperatively, the patient's thyroid sonography was completely normal and no further therapy was given. There was no evidence of recurrence at 2 years follow-up.

Comment

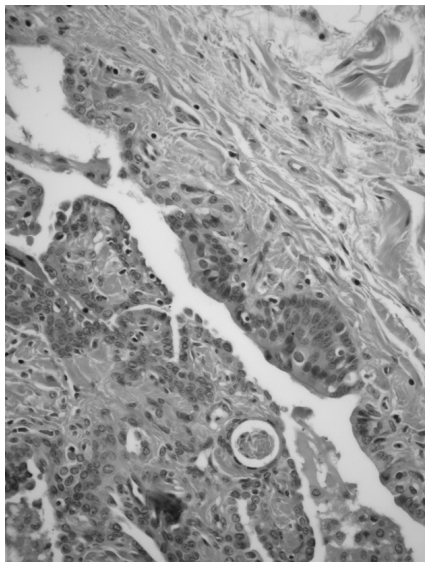
During the fourth week of embryogenesis, the anlage of the thyroid gland forms at the foramen cecum of the tongue. This diverticulum migrates downward to its adult location below the hyoid bone and anterior to the trachea and thyroid cartilage by the seventh week. During the descent, the thyroid gland remains connected to the base of the tongue by means of the thyroglossal duct. If this duct subsequently fails to atrophy it may give rise to a thyroglossal duct cyst. Such cysts are the most common causes of midline neck mass in children. Because of this embryologic development, a portion of the tract may persist anywhere along the course of the thyroglossal duct from the base of the tongue to the anterior neck. While most patients with a TDC are children and adolescents, up to one-third are aged 20 years or older. Thyroglossal duct cysts present usually as a slow growing asymptomatic midline neck mass that rarely causes symptoms when enlarged or infected.

When a clinical diagnosis of TDC is made, the workup should be completed in order to plan the appropriate surgery. Thyroid scanning is generally unnecessary and is justified only in patients with abnormal thyroid function tests and those in whom the normal location of the thyroid gland cannot be detected by ultrasonography [2]. Preoperative ultrasound is useful mainly for guiding fine needle aspiration biopsy. On ultrasound, TDC will appear as anechoic, hypoechoid or heterogeneous lesions, whereas TDC carcinoma may present as a mural lesion of the cyst with irregular margins or as a solid nodule in the cyst, sometimes with microcalcifications. Preoperative FNAB has been considered of questionable value for diagnosis of TDC carcinoma because of its relatively low accuracy. Nevertheless, some authors consider that in experienced hands and under ultrasound guidance FNAB should be performed in all adult patients with TDC [3]

About 1% of TDCs are found ultimately to harbor a thyroid carcinoma. These TDC carcinomas are generally not suspected preoperatively and are clinically indistinguishable from benign TDC. Eighty percent of them are papillary carcinomas. Other histological subtypes are mixed papillary-follicular carcinoma (8%) and squamous cell carcinoma (6%). Finding papillary carcinoma in thyroglossal remnants raises the possibility of an occult primary carcinoma in the thyroid. This is the source of the controversy regarding the optimal surgical management of TDC carcinoma. Papillary thyroid carcinoma frequently carries gene mutations and rearrangements that lead to activation of mitogen-activated protein kinase that promotes the development of the malignant phenotype. In adults, about 40% or sporadic papillary carcinomas have rearrangements involving RET and NTRK1 genes that code for the receptor of thyroid kinases. Somatic point mutations in the BRAF gene that codes for a serine/threonine kinase occur in 29–69% of papillary thyroid carcinomas and may confer a worse clinical prognosis than for this carcinoma without BRAF muta-

TDC = thyroglossal duct cysts

FNAB = fine needle aspiration biopsy



Papillary carcinoma within a thyroglossal duct cyst. On the left is the cyst wall lined by respiratory epithelium (hematoxylin & eosin. X 400).

tions. Furthermore, papillary carcinoma is frequently multifocal. Most of the non-contiguous tumor foci are microscopic (< 1 cm in diameter) and are often found at autopsy. The origin of these foci is unknown. They may be intraglandular metastases of a single dominant tumor, or they may arise from unrelated neoplastic clones as demonstrated by molecular analysis of DNA extracted from distinct foci of multicentric papillary carcinomas [4]. This distinction may have implications for the pathogenesis and treatment of the disease. Some authors consider that patent TDC can be a route for metastatic involvement from the thyroid gland and that such a finding always indicates that carcinoma is present in the thyroid itself or that the cyst represents a cystic area of a papillary carcinoma in the entopic thyroid gland. Consequently, they recommend total thyroidectomy as treatment. Others believe that papillary carcinoma found in a cystic lesion of the midline neck may actually arise in ectopic thyroid tissue and that total thyroidectomy might not be necessary as a routine procedure. Although the surgical management of TDC carcinoma is still controversial, most authors believe that incidentally discovered TDC papillary carcinoma can be adequately resected by the Sistrunk procedure alone, provided there is no clinical or sonographic suspicion of thyroid lesion or cervical adenopathies. This procedure is associated with a cure rate of 95% of TDC in all series reported. Patel and al. [5] showed that the addition of total thyroidectomy to the Sistrunk procedure had no significant impact on outcome. In accordance with their recommendation, we decided not to suggest a total thyroidectomy to our patient. Post-surgical treatment with suppressive doses of thyroid hormones is debated when dealing with differentiated carcinoma without extracapsular invasion and/or lymph node metastases and with normal thyroid [3]. In patients with low risk disease treated with the Sistrunk procedure, there are no data supporting the role of thyroid suppression therapy. Authors who advise total thyroidectomy in all TDC carcinomas recommend also postoperative radioactive iodine ablation and thyroxine suppressive therapy.

The prognosis of papillary carcinoma arising in TDC is excellent, with an overall survival rate of 95.6% at 10 years [5]. The post-surgical follow-up of patients is limited to an annual clinical and sonographic cervical examination and measurement of thyroid stimulation hormone level.

In summary, we present a patient with papillary carcinoma identified after resection of a TDC. We emphasize the importance of determining the nature and origin of papillary carcinoma found in thyroid remnants and suggest that the Sistrunk operation is an adequate procedure for most patients with papillary carcinoma incidentally discovered in TDC.

References

1. Sistrunk WE. Technique of removal of cysts and sinuses of the thyroglossal duct. *Surg Gynecol Obstet* 1928;46:109–12.
2. Kessler A, Eviatar E, Lapinsky J, Horne T, Shlamkovitch N, Segal S. Thyroglossal duct cyst: is thyroid scanning necessary in the preoperative evaluation? *IMAJ* 2001; 3:409–10.
3. Plaza CP, Lopez ME, Carrasco CE, et al. Management of well differentiated thyroglossal remnant thyroid carcinoma: time to close the debate? Report of five new cases and proposal of a definitive algorithm for treatment. *Ann Surg Oncol* 2006;13:745–52.
4. Giannini R, Ugolini C, Lupi C, et al. The heterogeneous distribution of BRAF mutation supports the independent clonal origin of distinct tumor foci in multifocal papillary thyroid carcinoma. *J Clin Endocrinol Metab* 2007;92:3511–16.
5. Patel SG, Escrig M, Shaha AR, et al. Management of well-differentiated thyroid carcinoma presenting within a thyroglossal duct cyst. *J Surg Oncol* 2002;79:134–9.

Correspondence: Dr. A. Ishay, Endocrine Institute, HaEmek Medical Center, Afula 18101, Israel.
Phone: (972-4) 649-5556
Fax: (972-4) 634-2019
email: ishay_av@clalit.org.il