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SCIENTIFIC LETTER

Papillary thyroid carcinoma on thyroglossal duct cyst: A series of 7 cases

Carcinoma papilar de tiroides sobre quiste del conducto tirogloso: una serie de siete casos

Thyroglossal duct cyst (TGDC) is the most common cervical cystic lesion. The occurrence of a carcinoma on a TGDC is rare, with a prevalence of <1%. Papillary carcinoma (PC) accounts for more than 90% of malignant cases, followed by mixed papillary and follicular carcinoma.[1,2]

The aim of this study is to present a descriptive retrospective analysis of a series of cases with a diagnosis of PC on TGDC from a tertiary referral center from 2006 through 2023. PC on TGDC was diagnosed in 7 (1.58%) out of 442 patients with PC. The objective of the study is to analyze the clinical characteristics, treatments administered, and final outcomes in this cohort (Table 1).

Diagnosis is usually established in adulthood, being predominant in women[3]; however, in our study, 42.9% were women, with a mean age at diagnosis of 54 years. The diagnosis of PC on TGDC is histological following cyst removal. The incidence rate of malignancy varies from <1% up to nearly 20% according to different series. Clinical presentation is similar to that of benign TGDC cases, usually a midline cervical mass. In 71.4% of the study cases, the reason for consultation was the evaluation of TGDC, and in 28.6%, thyroid nodularity.

It is also recommended to rule out malignant lesions in the thyroid gland, as some studies suggest that although 11% up to 33% of cases exhibit thyroid carcinoma, it is difficult to differentiate if these are synchronous neoplasms or independent primary carcinomas.[4] Five of our patients underwent preoperative fine-needle aspiration (FNA) of the TGDC: 4 showed cytological criteria of malignancy and 1 benignity. In the remaining 2 patients, the finding was incidental in the pathological study after TGDC surgery.

The treatment of TGDC with suspected malignancy is surgical. The Sistrunk procedure (SP) is the gold standard surgical therapy. The current trend in managing papillary thyroid carcinomas (PTC) on TGDC is to reserve total thyroidectomy (TT), cervical lymphadenectomy, and radioiodine therapy for high-risk patients (age > 55 years, tumor size > 4 cm, squamous cell carcinoma, extracapsular or vascular invasion, positive margins, lymph node or distant metastasis) or for concurrent malignant thyroid lesions.[5,6] However, this was not always the case, and in many instances, surgical treatment of PTC on TGDC included TT even without radiological evidence of nodularity. The choice of a more conservative approach is currently based on the use of preoperative diagnostic tests that help rule out local or distant disease, knowledge of the lack of influence of microscopic lymph node invasion on long-term survival, the high cure rate with SP, and the possibility of long-term follow-up.

As shown in Table 1, in our series, 71.4% of patients underwent SP and TT. Although in one of the cases, TT and lymphadenectomy were performed later (case #3), no preoperative FNA was ever conducted. In the remaining more recent cases (cases #6 and #7), the procedure was only SP as the histopathological characteristics were of low risk (lesions < 1 cm, encapsulated, without capsule rupture or vascular or perineural invasion) and the thyroid gland did not show suspicious malignant features on the ultrasound. Case #2 presented thyroid nodules with Bethesda category III FNA and another nodule with cytological suspicion of PC (which was not subsequently confirmed). In all cases from our series, the PC was localized to the TGDC with no evidence of malignant thyroid involvement. In case #3, prophylactic central neck dissection was performed (negative), and in case #2, a perihyoid lymph node was removed, which turned out positive for PC metastasis.

The prognosis of PC on TGDC is excellent, with 10-year survival rates of 95%, and the rare presence of distant metastases.[7] However, risk staging is necessary to guide therapeutic decisions. Except for the 2 patients in the study who only underwent SP, the rest were categorized as stage I (AJCC 8th Edition)[8] with a low risk of recurrence (ATA 2015).[6,9] In 71.4% of our patients, a BRAF (B-Raf proto-oncogene) V600E mutation was detected. BRAF positivity in thyroid neoplasms appears to be a predictive factor for locally advanced disease and a higher likelihood of recurrence. For some authors, the presence of this mutation in the FNA of TGDC may serve as a preoperative indicator for performing TT along with SP.[10] However, its positivity has not been associated with worsened disease-free survival or worse final outcomes.[11] A total of 57.1% of our patients received a single dose of radioiodine, with a mean of 82 mCi (SD ±54 mCi). Treatment response was excellent, with no recurrences and no evidence of disease at the time of data collection, considering that 3 patients have undergone a 12-month follow-up.

Table 1 Clinical characteristics, treatments, and final outcomes of patients with papillary carcinoma on thyroglossal duct cyst.

	Case #1	Case #2	Case #3	Case #4	Case #5	Case #6	Case #7
Year of diagnosis	2006	2007	2010	2014	2023	2023	2023
Sex	F	F	M	M	M	F	F
Age at diagnosis	72	44	55	70	41	52	48
Reason for diagnosis	Thyroid nodule study	Thyroid nodule study	Compressive symptoms from TGDC	Incidental finding on PE	Incidental finding on cervical US	Compressive symptoms from TGDC	Compressive symptoms from TGDC
FNA of cyst	Malignant	Malignant	Not performed	Malignant	Malignant	Not performed	Benign
PC size (cm)	U	0.4	ND	2.6	0.9	1.2	1.2
Metastatic lymph nodes	No	Yes (unilateral)	No	No	No	No	No
Surgical Treatment	SP + TT	SP + TT + LD (Ipsilateral CND)	SP + TT + LD	SP + TT	SP + TT	SP	SP
Molecular study (BRAF)	V600E	V600E	Wild type	V600E	V600E	Not performed	V600E
I-131 treatment	Yes	Yes	Yes	Yes	No	No	No
Risk categorization (ATA 2015)	Low risk	Intermediate risk	Low risk	Low risk	Low risk	Low risk	Low risk
Dynamic risk stratification	ER	ER	ER	ER	ER	ER	ER

F: female. M: male. PE: physical exam; US: ultrasound; TGDC: thyroglossal duct cyst. cm: centimeters. ND: not disclosed. SP: Sistrunk procedure. TT: total thyroidectomy. LD: lymphadenectomy. CNS: central neck dissection. I: Iodine. BRAF: B-Raf proto-oncogene. ATA: American Thyroid Association. ER: excellent response; U: unknown.

In conclusion, PC on TGDC is a rare form of PC with an excellent prognosis. Proper preoperative staging is essential to determine the best approach. Given the favorable prognosis of these tumors, surgical management of the TGDC alone may be sufficient as a therapeutic option, unless there is suspicion of thyroid gland involvement.

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Conflicts of interest

None declared.

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