case report

Follicular thyroid carcinoma within a thyroglossal duct cyst in adult

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Thyroglossal duct cyst carcinoma is a rare malignancy, with an incidence of 0.7% to 1.6%. Most cases of thyroglossal duct cyst carcinoma are papillary carcinoma, with follicular carcinoma having been rarely reported. In this study, a 33-year-old man presented with a typical thyroglossal duct cyst and underwent surgical resection of the cyst, which was determined to be follicular carcinoma. We have reported this rare case to increase the awareness of such entities within the general otolaryngology and the community of head and neck surgeons, as well as among endocrine surgeons.

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Thyroglossal duct cyst (TGDC) is a common benign mass of the midline neck, affecting approximately 7% of the adult population. Up to 1.6% of TGDC cases have been reported to host carcinomatous thyroid tissue; there are around 260 reported cases in the literature to date.[1] Papillary carcinoma is the most common carcinoma arising from TGDC, with an incidence of 87%.[2] Other carcinomas that are specific to the thyroid glands, such as follicular, mixed papillary-follicular, and Hurthle cell carcinoma have been infrequently reported.[2] In this study, we report an adult male with a thyroglossal duct cyst, which was discovered to harbor follicular carcinoma. We reported this rare case to increase the awareness of such entities within the general otolaryngology and the community of head and neck surgeons, as well as among endocrine surgeons.

CASE

A 33-year-old healthy male patient was referred to the Otolaryngology–Head and Neck Surgery Clinic at King Abdullah Medical City, Makkah, Saudi Arabia, with a 2-year history of painless swelling of the midline neck. On examination, a midline, infra-hyoid, single, well-defined, non-tender, mobile, and firm swelling was palpated and measured 4×5 cm. The swelling moved with both deglutition, as well as, protrusion of the tongue. There was no palpable cervical lymphadenopathy (**Figure 1A**). All laboratory examination results, including a thyroid function test, were within normal limits.

Ultrasonography (US) and computed tomography (CT) imaging of the neck showed a midline heterogeneous cystic lesion at the hyoid and infrahyoid level, measuring 4×3 cm, with central areas of hypoattenuation and peripheral enhancement. The thyroid gland appeared normal with no cervical lymphadenopathy (**Figure 2**). A technetium-99m pertechnetate thyroid

scan confirmed normal function of the thyroid gland, with a slight uptake in the right lobe (**Figure 3**), while US-guided, fine-needle aspiration cytology (FNAC) did not reveal any conclusive tumor diagnosis (Bethesda category-1 cytopathology).

The patient underwent classic surgical resection using the Sistrunk's procedure (**Figure 1B**). The post-operative pathology report confirmed a 3.5-cm tumor focus of well-differentiated follicular carcinoma arising from a TGDC (**Figure 4**). The patient's case was discussed at our multidisciplinary thyroid meeting. Although the American Thyroid Association (ATA) guidelines have no strong recommendation for any further adjuvant therapy, we thought that the 3.5-cm tumor focus represented the upper limit of T2, and there were no clear recommendations in the literature for this type of TGDC, with a known slight aggressive behavior of thyroid cancer in Saudi Arabia. Therefore, we recommended total thyroidectomy, followed by radioactive iodine therapy as an adjuvant therapy for this case. A thyroidectomy was performed and the histopathology showed no evidence of cancer in the native thyroid gland.

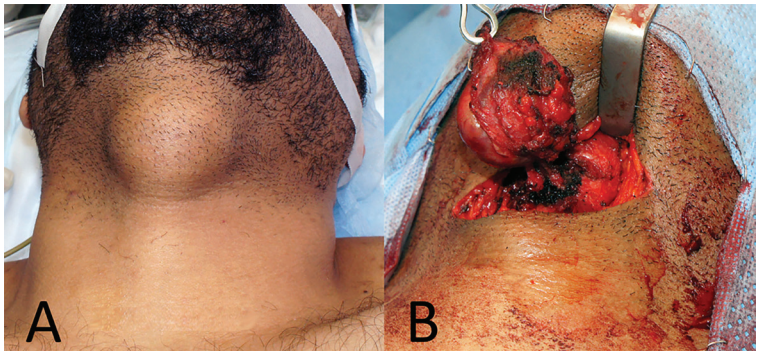

Figure 1. Clinical images: (A) preoperative and (B) intraoperative.

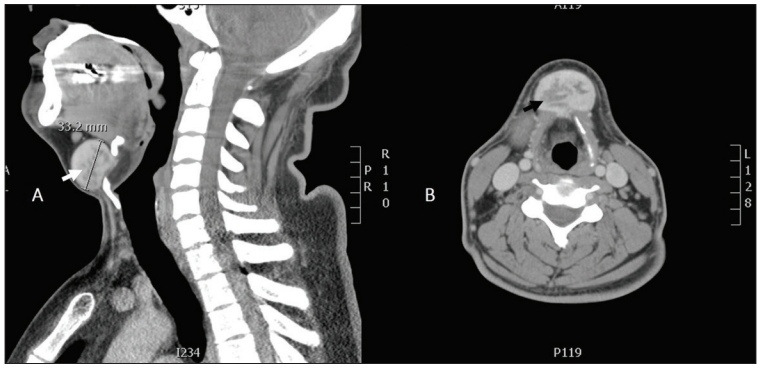

Figure 2. (A) Sagittal and (B) axial views of a computed tomographyscan of a midline neck heterogeneous cystic lesion at the hyoid and infrahyoid level, measuring 4×3 cm, with central areas of hypoattenuation and peripheral enhancement.

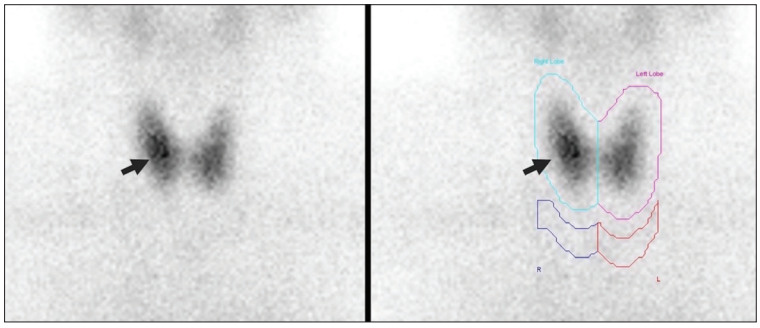

Figure 3. Technetium-99m pertechnetate thyroid scan showing a normal functioning thyroid gland with a slightly higher uptake in the right lobe, with no uptake in the mass.

DISCUSSION

Carcinoma arising from TGDC is very rare.[1] The overall sex and age distribution of TGDC carcinoma is similar to that of thyroid carcinoma, with women affected more than men in a 3:2 ratio.[3] The median age at presentation is 40–50 years, with peak incidence in the third and sixth decade for women and men, respectively.[3]

Even though papillary carcinoma is considered the most common TGDC type, with an incidence of 87%, only 10 cases have been reported to harbor follicular carcinoma.[1] **Table 1**[4-7] lists 4 case series of 61 patients with TGDC carcinoma, with 6 cases of follicular carcinoma having been analyzed. In the previous studies, all patients were women, with 1 unspecified gender, and the median age was 38–72 years. Surgeons led by Patel et al. performed only the Sistrunk's procedure without following the procedure with aggressive management using total thyroidectomy and radioactive iodine ablation. In the other three series, US with FNAC revealed the mass to be highly suspicious of TGDC carcinoma. Therefore, patients underwent treatment with the Sistrunk's procedure and total thyroidectomy, followed by radioactive iodine ablation and suppressive levothyroxine. Malignant involvement of the thyroid gland was found in four cases, with three cases of papillary histotype and one case of follicular histotype. Lymph node metastasis and distant metastasis were not reported in any case. Moreover, all patients were recurrence-free during their follow-up period. Details of the other four cases were not found in the literature.[4] In our case, this patient with follicular carcinoma of a TGDC was male and younger than the median age reported in the other studies. The same management was obtained. During a 9-month follow-up period, there was no evidence of recurrence.

The exact origin of carcinoma of a TGDC is still controversial. Two theories have been suggested to elucidate the origin of TGDC carcinoma: either it is

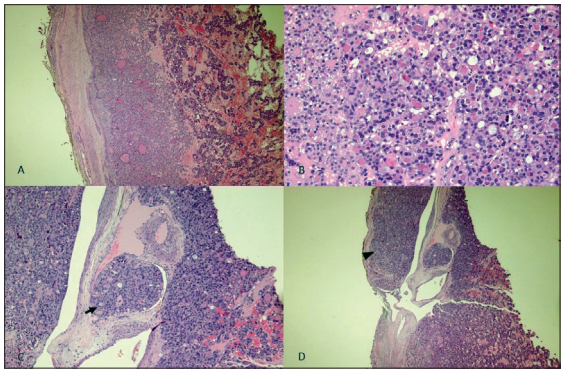

Figure 4. Histopathological hematoxylin-eosin staining, under both low- and high-power magnification, showing: (A) a well-circumscribed nodule, surrounded by a thick capsule, and containing thyroid follicles and colloid tissue; (B) cells exhibiting dense and enlarged pleomorphic nuclei; (C) vascular invasion; and, (D) capsular invasion, as indicated by the arrowhead.

representative of a "metastatic lesion" from an occult primary thyroid gland, or it is "de novo" in origin. The former theory is supported by the fact that most TGDC carcinoma cases present with a concurrent thyroid cancer. In addition, the presence of a fibrous tract that connects the TGDC and the thyroid gland is thought to be a possible path for the thyroid carcinoma to spread into the cyst.[8] While TGDC carcinoma shows a larger tumor size, increased frequency of adjacent soft tissue extension, the presence of agglomerates of thyroid follicles within the cyst wall in 62% of cases, and the absence of reported cases of medullary carcinoma that arise from a different origin tissue of the thyroid other than follicular cells, all are characteristics that favor the "de novo" origin theory.[7,8] Other supporting evidence of the de novo theory is the fact that some of the reported cases of follicular TGDC carcinoma in the literature presented with a different type of carcinoma (papillary carcinoma) in the thyroid gland.

The majority of TGDC carcinomas are detected incidentally in a postoperative histopathology report.[7] Clinical signs that raise the suspicion of carcinoma, such as fixation to the hyoid bone or surrounding structures, hardness, irregularity, a sudden growth of the mass, and lymphadenopathy, are rarely present in patients with TGDC tumors.[8]

Radiologic examinations (US, CT, and/or magnetic resonance imaging) can suggest malignancy when the presence of a mural lesion in the cyst wall, microcalcifications, invasion of the cyst wall, and irregular margins are discovered.[9] The role of scintigraphy is to prove the existence of a functional thyroid gland and exclude the presence of functional ectopic thyroid tissue within the cyst.

Table 1. Summary of follicular carcinoma within thyroglossal duct cyst carcinoma: review of the recent literature.[4-7]

1st author	No. of patients	No. with FC	Age/ sex	U/S with FNAC	Management	Thyroid	Lymph node metastases	Distant metastases	Adjuvant therapy	Follow-up (months)	Recurrence
Miccoli et al[5]	18	1	58 / -	TGDC carcinoma	SP→TT	PTC	No	No	RAI + suppressive L.T4	63.0	No
Patel et al[4]	5	1	38 / F	-	SP	-	No	No	Suppressive L.T4	144.0	No
Dzodic et al[6]	12	1	51 / F	Intra-cystic mass	SP, TT	FC	No	No	RAI + suppressive L.T4	204.0	No
Pellegriti et al[7]	26	3	41 / F	Cyst with follicular lesion	SP, TT	PTC	No	No	RAI + suppressive L.T4	182.9	No
			43 / F		SP, TT	PTC	No	No	RAI + suppressive L.T4	17.0	No
			72 / F		SP, TT	No	No	No	RAI + suppressive L.T4	77.0	No
Our case	1	1	33 / M	Heterogeneous cystic lesion	SP→TT	No	No	No	RAI + suppressive L.T4	9.0	No

FC=Follicular Carcinoma; U/S=Ultrasound; FNAC=Fine Needle Aspiration Cytology; SP=Sistrunk's Procedure; TT=Total Thyroidectomy; PTC=Papillary Thyroid Carcinoma; RAI=Radio-Active Iodine; L.T4=Levothyroxine

FNAC is considered the most reliable diagnostic tool for examining TGDC carcinoma with a positive predictive value of 69% and sensitivity of 56–62%. The false-negative diagnosis by FNAC is mostly due to small neoplasm size, marginal location of the tumor in the cyst, and hypocellularity that results from dilution due to the cystic content.[12] Hence, a negative FNAC cannot rule out a diagnosis of TGDC malignancy, especially if there is high clinical suspicion.[10]

Since there are a limited number of TGDC carcinoma cases in the literature, there is no definite consensus on the optimal management of this malignancy. Low-level evidence from an expert opinion recommends a conservative approach with the Sistrunk's procedure and total thyroidectomy only when there is clinical suspicion of malignancy by "prognostic risk group assessment". High-risk factors include age older than 45 years, history of radiation exposure, radiologic evidence of a concurrent tumor in the thyroid gland, presence of lymphadenopathy, tumor size more than 1.5-4.0 cm, presence of cyst wall invasion, and positive margins on histopathological examination.[10] Other clinicians prefer more aggressive management with total thyroidectomy based on the following conditions: a multi-focality feature of papillary and follicular carcinoma,[13] high incidence of concomitant thyroid malignancy, for optimization the effect of radioactive iodine ablation as an adjuvant therapy, and the use of thyroglobulin as a follow-up oncology marker.[9] In conclusion, a well-differentiated follicular carcinoma within TGDC is a rare clinical entity. Management is dependant on the specifics of the case.

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