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Case report

Papillary carcinoma arising from a thyroglossal duct cyst: A case report and literature review

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ABSTRACT

Introduction: Papillary carcinoma originating from a thyroglossal cyst is rare and peculiar, with majority of cases detected after surgery. Despite an excellent prognosis, its management remains controversial. Herein, we report the case of a 53-year-old woman who underwent Sistrunk procedure for a thyroglossal duct cyst and was subsequently confirmed to have papillary thyroid carcinoma.

Presentation of case: A 53-year-old woman presented with an anterior midline neck mass for 7 years. The patient had no symptoms of hypo-or hyperthyroidism. Additionally, she had no history of compressive symptoms. Neck ultrasound revealed a well-defined 3.5 cm × 2.2 cm × 3 cm-sized cystic lesion inferior to the hyoid bone, with a peripheral solid component. Neck computed tomography revealed a well-defined 3.7 cm × 3.4 cm × 2.7 cm-sized cystic lesion with an enhanced central solid component with focal calcifications, inferior to the hyoid bone, and in contact with the anterior wall of the thyroid cartilage. Sistrunk procedure was performed. The patient was then diagnosed with papillary thyroid carcinoma with TNM stage pT2 and underwent total thyroidectomy as a follow-up procedure.

Discussion: Thyroglossal duct cyst carcinoma is usually detected in the fourth decade of life with a higher prevalence in women. Neck ultrasound is performed during the initial radiological workup to assess the cyst and confirm the presence of the thyroid gland.

Conclusion: The Sistrunk procedure is highly effective in low-risk patients. A more aggressive approach is required for high-risk patients.

1. Introduction

Thyroglossal duct cyst (TGDC) is the most common congenital thyroid gland disease, accounting for >75% of midline neck masses in children [1]. Approximately 1% of TGDCs cases occasionally transform into a TGDC carcinoma (TGDCC). TGDCC presents as a rapidly growing or asymptomatic neck mass. Brentano reported the first case of TGDCC in 1911 [1]. Currently, there are 300 published cases of TGDCCs [2]. Embryologically, thyroid gland development begins in the third week of gestation, and originates from the endodermal proliferation on the primitive pharyngeal median surface between the first and second pharyngeal pouches. This endodermal proliferation starts as an invagination within the tongue at the site of the foramen cecum, thereby creating a thyroid diverticulum. The thyroid diverticulum descends caudally and crosses the hyoid bone and larynx anteriorly to reach its final location in front of the trachea. It maintains the attachment to the tongue via the thyroglossal duct (TGD), which starts to degenerate by the fifth week of gestation, and disappears by the eight week. If the TGD

Abbreviations: CT, computed tomography; FNA, fine needle aspiration; PTC, papillary thyroid carcinoma; SP, Sistrunk procedure; TFT, thyroid function test; TGD, thyroglossal duct; TGDC, thyroglossal duct cyst; TGDCC, thyroglossal duct cyst carcinoma; TT, total thyroidectomy.

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fails to disappear, it persists as an ectopic tissue, a duct, or a cyst as observed in 7% of adults. Genetic factors are involved in this pathology [1]. Since the presentation of TGDCC is similar to that of benign lesions, most TGDCC cases are discovered only after surgery [1]. However, there is no consensus on the ideal management of TGDCCs [3]. In this report, we present a rare case of papillary carcinoma arising from a TGDC along with a literature review focusing on the management of TGDCC. This work has been reported in line with the SCARE 2020 criteria [4].

2. Presentation of case

A 53-year-old woman with hypertension on regular medications presented to our surgical clinic with an anterior midline neck mass persisting for 7 years without any changes in size. Occasionally, she had mild pain. She had no hypothyroidism, hyperthyroidism, or compressive symptoms. Other systemic review findings were unremarkable. She denied having prior radiation exposure or a family history of the same condition. Upon local examination, a 3 cm × 3 cm-sized oval, firm, and slightly tender anterior neck mass was detected, which moved up when swallowing and upon tongue protrusion. No palpable cervical lymph nodes were observed.

Results of all laboratory parameters, including thyroid function test (TFT), were normal. Neck ultrasound (US) revealed a well-defined 3.5 cm × 2.2 cm × 3 cm-sized cystic lesion located subcutaneously slightly inferior to the hyoid bone, with moderate vascularity of the peripheral solid component. Except for a 3.5 mm-sized isoechoic nodule in the isthmus, the thyroid gland was normal in size, shape, and echo pattern. Neck computed tomography (CT) revealed a well-defined 3.7 cm × 3.4 cm × 2.7 cm-sized cystic lesion with an enhanced centrally solid component with focal calcifications. The mass was observed slightly inferior to the hyoid bone, and in contact with the anterior wall of the thyroid cartilage (Fig. 1). Both US and CT revealed no abnormal cervical lymph nodes. Fine needle aspiration (FNA) revealed a benign cystic lesion.

The patient underwent the Sistrunk procedure (SP). A transverse incision was made over the mass. A 3-cm TGDC was located inferior to the hyoid bone. It was completely excised along with the rim of the hyoid bone (Fig. 2A). The patient's postoperative period was uneventful. The final histopathological results revealed a conventional 3-cm-sized papillary thyroid carcinoma (PTC). The surgical margins on the outer surfaces were clear (Fig. 3). Furthermore, there was no microscopic invasion of the adjacent muscle or fatty tissue, and no lymphovascular or perineural invasion. The pathologic TNM staging was determined to be pT2. The hyoid bone was tumor-free.

Considering the patient's age, tumor size, and the presence of isthmus nodule, total thyroidectomy (TT) was decided at a multidisciplinary meeting. The patient underwent TT (Fig. 2B). Post-operatively, the patient was doing well and was discharged from the hospital in good condition. The final histopathological report of the thyroid gland revealed a multifocal papillary thyroid microcarcinoma, with the largest one being 3 mm in the left lobe (Fig. 4). The lesion in the isthmus was a benign fibrotic nodule. Given the size of 3 mm, the presence of free margins, and the absence of lymph node involvement, the patient was considered to be at low risk. Therefore, it was decided at a multidisciplinary meeting that she needed to follow-up with neck US, TFT, and thyroglobulin level measurement. The patient was followed up for six months.

3. Discussion

TGDCC may occur in individuals aged 1–82 years. However, it is most common in the fourth decade of life with an increased prevalence in women than in men (female: male, 3:2) [1,5]. TGDCC manifests as a rapidly growing tender mass in the neck [6], but it may also present as an asymptomatic mass in the neck [5]. Notably, a definitive diagnosis is usually made after excision [3].

Overall, the risk of malignant transformation in TGDC cases is 1%. Histologically, PTC (80%) is the most frequently identified cancer in TGDCs. However, cases of mixed papillary/follicular carcinoma (8%) and squamous cell carcinoma (6%) have been reported. Other rare cancer types (6%) include Hurthle cell, follicular, and anaplastic carcinomas [1]. The patient in our case had PTC.

Synchronous papillary carcinoma in TGDCs and the thyroid gland exhibit multifocal tumors rather than a metastatic spread [1]. Moreover, metastasis to the cervical lymph nodes ranges from 7% to 15%, which is less in comparison to PTCs. Additionally, the mortality rate of TGDCCs is

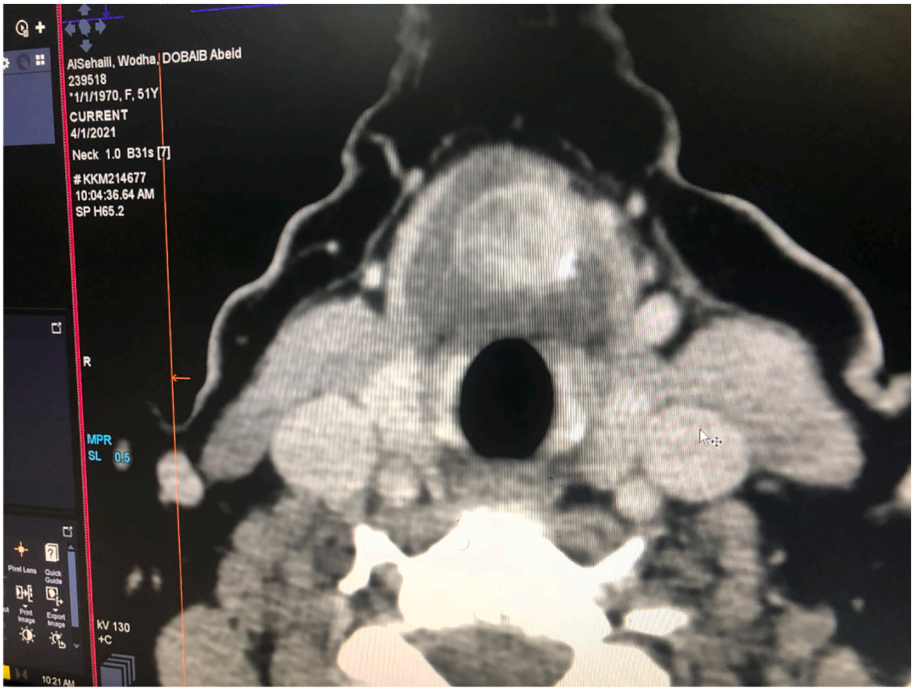

Fig. 1. Neck computed tomography showing a cystic lesion with a contrast-enhanced central solid component with focal calcifications.

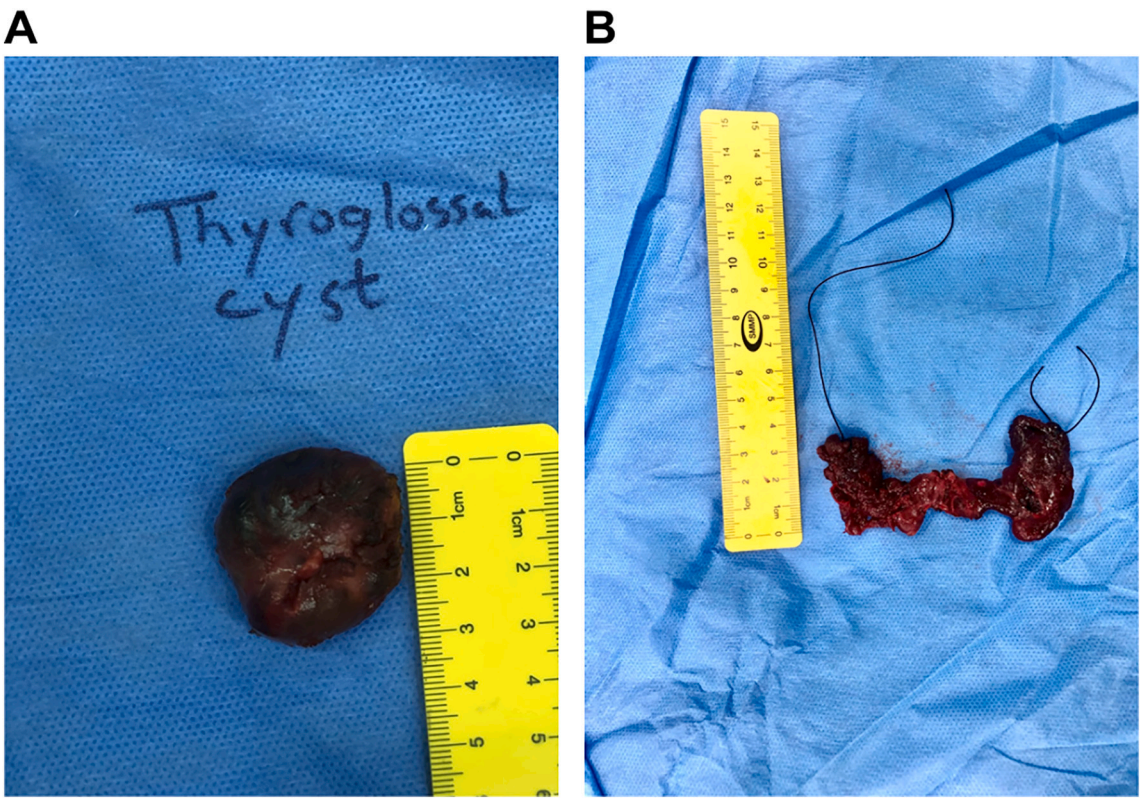

Fig. 2. Operative specimen photographs showing the thyroglossal duct cyst (A) and the thyroid gland (B).

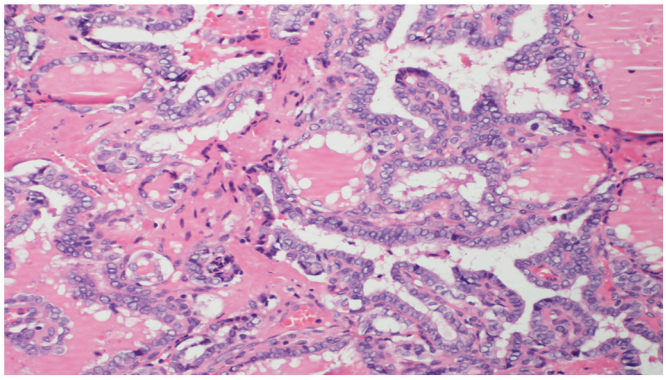

Fig. 3. Microscopic examination of the thyroglossal duct cyst showing a papillary thyroid carcinoma (degree of magnification is 20x).

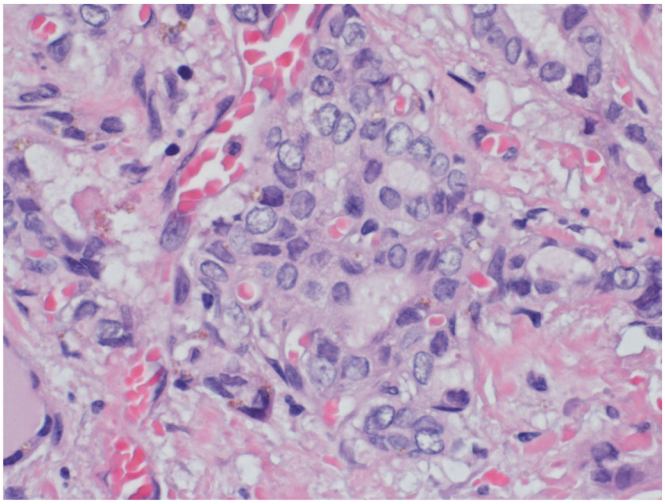

Fig. 4. Microscopic examination of the thyroid gland showing a papillary thyroid microcarcinoma (degree of magnification is 40x).

extremely low [1].

The etiology of TGDCCs remains unknown; however, possible theories include metastasis from an occult primary tumor or a spontaneous development of ectopic thyroid tissue present within the TGDC wall [1,6]. The latter is the most prevailing theory. This finding is supported by the absence of medullary carcinomas in TGDCs [1,5].

In general, neck US is recommended as part of the initial radiological workup for TGDCs to examine the cyst and confirm the existence of the thyroid gland. US features suggestive of TGDCCs include calcification, regional lymphadenopathy, and solid components (mural nodules) [6]. Contrarily, neck magnetic resonance imaging and CT can identify the tumor as a solid nodule within the cyst, combined with thickening of the cyst wall, calcification, or margin irregularity [1]. US and CT of our patient revealed malignant features (such as a solid component, calcifications, and intense vascularity within the mass).

The diagnostic potential of preoperative FNA in TGDCCs remains controversial. However, the diagnostic rate for TGDCC detection is 53%, which could increase if the solid component is sampled [6]. In the present case, FNA cytology results indicated a benign tumor.

Although there is no definitive agreement regarding TGDCC management, SP should be performed in all cases as an initial procedure [5]. SP offers a cure rate of up to 95% in papillary carcinoma of TGDCs, with an excellent prognosis (95–100%) [1]. Patel et al. found that TT did not have a significant effect on the outcome when SP was added. Furthermore, using univariate analysis, the authors discovered that the extent of the initial surgery was the sole significant predictor of overall survival [7]. Additionally, the risk of complications following TT (hypocalcemia and recurrent laryngeal nerve injury) is at odds with the need for further

surgeries [1]. Furthermore, it has been suggested that SP is an adequate operation and that TT should not be considered, particularly in the following circumstances: patients aged <45 years with no prior history of neck radiation, a tumor sized <1–1.5 cm or 4 cm (based on the series), clinically or radiologically normal thyroid gland, presence of negative margins, and absence of metastatic lymph node spread [1,5].

Conversely, other authors advocated TT as a follow-up surgery in patients with a TGDCC as a result of thyroid gland involvement in 33–45% of cases leading to radioactive iodine (RAI) ablation and the use of thyroglobulin level as a follow-up marker [5,8,9]. According to a recent study, SP, TT, neck dissection, and RAI are recommended as the ideal approaches in high-risk patients (patients aged >45 years, patients with tumor invasion to the surrounding soft tissues, tumor size >4 cm, lymph node and distant metastases, male sex, and cold nodules in the thyroid gland on thyroid scan) [10,11]. Moreover, investigators have recommended postoperative RAI ablation, a suppressive dose of levothyroxine therapy, and a whole-body scintigraphy as the best approach for follow-up [1].

As a rare disease, TGDCC management remains controversial. Furthermore, it should be managed on a case-by-case basis with multidisciplinary collaboration. Future case series may address and resolve the challenges with TGDCC treatment. We believe our case enriches the literature on such a rare tumor.

4. Conclusion

TGDCC is almost always diagnosed post-operatively with excellent prognosis. Given the rarity of TGDCCs, there is no definitive consensus on its management. However, SP is sufficient in low-risk patients, whereas more aggressive approaches are required in high-risk patients. Further case series could resolve the controversy around appropriate treatment.

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Ethical Approval

The requirement for ethical approval was waived by our institution as this was a case report.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author Contribution

Author name	Role
Saad M. Alqahtani	Conceptualization, Methodology, Validation, Investigation, Writing- Original draft preparation, Writing- Reviewing and Editing, Visualization, Supervision, Project administration, operation, and follow-up. Guarantor of manuscript.
Musaed Rayzah	Revised manuscript critically for important intellectual content, literature review, and assist in operation.
Ahmed Al Mutairi	Endocrinologist involved in clinical management of the patient and follow up. Revised manuscript critically for important intellectual content.
Mohammed Alturiqy	Acquisition, review, and interpretation of all radiological images, and revised manuscript critically for important intellectual content
Ahmed Hendam	Data collection and clinical management of the patient.
Maraei Bin Makhashen	Acquisition, review, and summary of the slides
All authors	Read and approved the version to be published.

Guarantor

Saad M. Alqahtani.

Declaration of competing interest

None.

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