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CASE REPORT

Papillary carcinoma in a thyroglossal duct remnant: A case report and discussion on management

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KEYWORDS

Thyroglossal duct carcinoma;
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Abstract *Objective:* Thyroglossal duct carcinoma is a malignant tumor arising within a thyroglossal remnant. It is a rare entity, seen generally in adults, and characterized by relatively non-aggressive behavior. This case is presented because of its rarity and the presence of cervical lymph node metastasis.

Case report: A 37-year-old female patient presented with a slowly and progressively growing mid-line neck mass. Neck ultrasound and computed tomography scanning revealed a mid-line sub-mental pre-hyoid highly enhancing neck mass, bilateral cervical lymphadenopathy, and tiny nodules in both lobes of the thyroid gland. Fine needle aspiration cytology of the neck mass revealed papillary thyroid carcinoma. The patient underwent total thyroidectomy, Sistrunk procedure, bilateral level I–IV neck dissection, and postoperative I131 ablation and hormonal suppression with thyroxine. The patient has been disease free for 1 year after the operation.

Conclusion: Computed tomography scanning and fine-needle aspiration cytology enhance the preoperative diagnosis. Sistrunk procedure is the standard treatment with a clinically and radiologically normal thyroid gland, while the more aggressive treatment is necessary in advanced cases. The concept of prognostic risk groups should be used to identify patients who would need a more aggressive approach. The prognosis is generally excellent with adequate treatment.

1. Introduction

A thyroglossal duct cyst (TDC) is the most common anomaly in the development of the thyroid gland. This is the most common form of congenital malformations in the neck as 70% of mid-line masses diagnosed during childhood and 7% in adults are TDCs. It can be located in an area from the base of the tongue to the pyramid of Lalouette and is present characteristically as a mid-line cervical mass at the level of the thyrohyoid

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membrane.[1–3] Thyroglossal duct carcinoma (TDCa) is a malignant tumor arising within a thyroglossal remnant or a thyroglossal duct cyst. This is a rare condition as 1% of TDCs can harbor malignancy.[4] A review of the literature showed that 260 cases of malignant thyroglossal cysts have been reported since the first description by Bretano in 1911. The majority of the cases were discovered during pathological examination of TDC specimens.[3,5]

In this article, we present a case of papillary thyroid carcinoma of a thyroglossal duct remnant because of its rarity, unusual manifestation, and the lymphatic spread, and discuss the current opinions concerning the management.

2. Case presentation

A 37-year-old female patient presented with mid-line neck mass that grew slowly and progressively over 2 years. The mass was intermittently painful. The patient had no history of hoarseness, breathing difficulty or dysphagia. She reported no history of radiation exposure. Physical examination revealed a hard and well circumscribed mass that was mobile with deglutition and protrusion of the tongue. It was fixed to the deep planes of the neck. However, it was not attached to the skin.

Thyroid function tests [serum-free thyroxine (T4), free triiodothyronine (T3), and thyroid-stimulating hormone (TSH)] were within normal limits. Neck ultrasound revealed an echogenic mass of 3.7×2.6 cm in size in the sub-mental region in the mid-line with no cystic changes and bilateral cervical lymphadenopathy, predominantly at level II and III, and more on the right side. The largest lymph node was on the right side and measured 1.3×0.6 cm in size. The thyroid gland was of normal size, shape, and position and few tiny nodules less than 5 mm were seen in both lobes. A computed tomography (CT) scanning of the soft tissue of the neck with and without contrast revealed a mid-line sub-mental pre-hyoid neck mass of 4×2.5 cm in size, highly enhancing, deep to the platysma muscle, and attached to the infra-hyoid muscles (Figs. 1–3). Some heterogeneity was seen in the thyroid gland, while there was no significant lymph node enlargement. Fine needle aspiration cytology of the neck mass revealed papillary thyroid carcinoma.

After multidisciplinary discussion, a fully informed written consent was obtained from the patient, and she underwent total thyroidectomy, Sistrunk procedure, and bilateral level I–IV neck dissection. The strap muscles of the neck were only pushed by the sub-mental mass and were not involved by it. Therefore, they were completely dissected of the sub-mental mass and preserved.

The postoperative histopathology report revealed $3.5 \times 2.6 \times 3$ cm papillary thyroid carcinoma of the thyroglossal duct remnant with negative margins and uninvolved hyoid bone and another focus of papillary micro-carcinoma (0.5 cm) confined to the left lobe of the thyroid gland. Non-keratinizing squamous epithelium and normal thyroid follicles were identified and a focus of vascular invasion was seen in the main Sistrunk specimen. The tumor was partially encapsulated and no invasion into the muscles was seen.

Metastatic papillary thyroid carcinoma was identified in 12 out of 103 lymph nodes within the levels left IIA and III and right IIA and III. The largest metastatic lymph node was at the level right III, and was 13 mm in size.

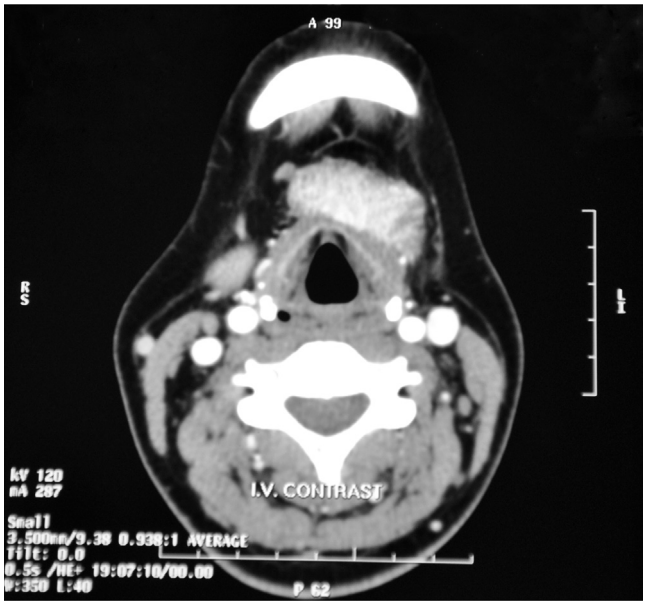

Figure 1 (Axial view) Preoperative post contrast computed tomography scanning of the soft tissue of the neck showing a mid-line pre-hyoid neck mass, highly enhancing, deep to the platysma muscle, and attached to the infra-hyoid muscles.

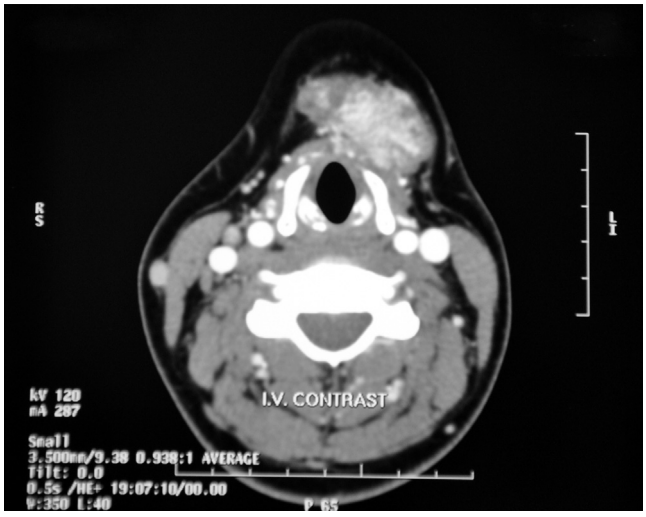

Figure 2 (Axial view) Preoperative post contrast computed tomography scanning of the soft tissue of the neck showing a mid-line neck mass, highly enhancing, deep to the platysma muscle, and attached to the infra-hyoid muscles.

Postoperatively, the patient had good wound healing, normal voice and breathing, transient hypocalcemia, and uneventful recovery. One month postoperatively, the patient received I131 (120 mCi) followed by hormonal suppression with thyroxine. Ten months postoperatively, I131 whole-body scanning showed no active iodine-avid tissue in the neck region, and revealed no metastasis. Neck ultrasound revealed no residual or recurrent mass. Thyroxine withdrawal stimulated thyroglobulin level was 0.4 ng/ml and thyroglobulin antibody level was negative about 45.5 U/ml. The patient has been a disease free for 1 year after the operation. The Institutional Reviewer

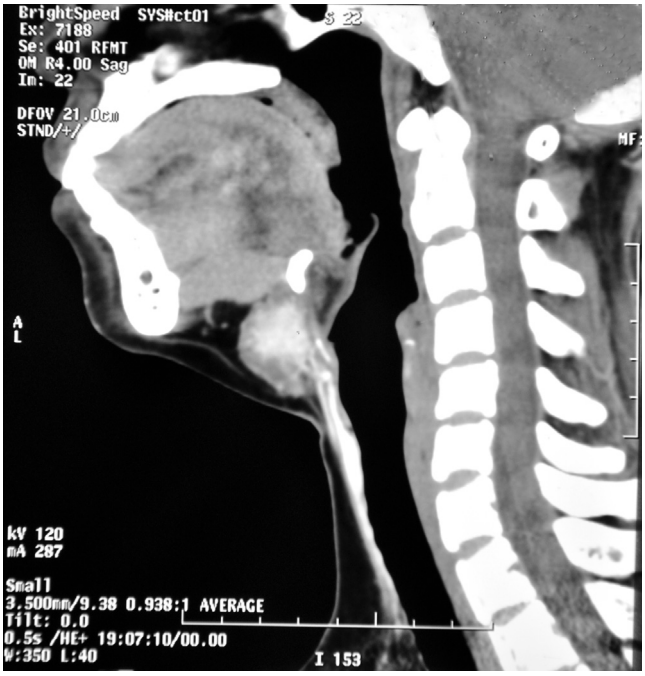

Figure 3 (Sagital view) Preoperative post contrast computed tomography scanning of the soft tissue of the neck showing a mid-line pre-hyoid neck mass, highly enhancing, deep to the platysma muscle, and attached to the infra-hyoid muscles.

Board (IRB) of Saad Specialist Hospital has approved the publication.

3. Discussion

The thyroid gland descends from the foramen cecum to its location at the point below the thyroid cartilage. It leaves behind an epithelial tract known as the thyroglossal tract. This tract usually disappears during the 5th–10th gestational weeks. However, incomplete atrophy of the thyroglossal tract or retained epithelial cysts creates the basis for the origin of a thyroglossal duct cyst. A thyroglossal remnant may be a cyst, a tract, a duct, a fistula, or an ectopic thyroid within a cyst or duct.[6]

TDCs most often present with a palpable asymptomatic mid-line neck mass at the level of or below the hyoid bone. Suprahyoid thyroglossal duct cysts are located in the mid-line of the neck. The more common infrahyoid thyroglossal duct cysts often have both mid-line and off-mid-line components, with the latter embedded in the strap muscles. The presence of a solid mass along it should raise the suspicion of ectopic thyroid tissue, in which occult malignancy is more likely.[7]

Malignant transformation is a rare complication of untreated TDCs. Most cases of TDCa are diagnosed during the third and fourth decades of life, and rarely in children under the age of 14 years. The sex ratio, female/male is 3:2.[5,8] Our case is a 37-year-old female patient.

The clinical presentation of TDCa is often similar to that of TDC. Therefore, in most cases, the diagnosis of malignancy is not made until surgery is done. Usually, asymptomatic anterior mid-line neck mass is the chief complaint. A hard, fixed remnant, rapid increase in size, the occurrence of pain, and the presence of enlarged lymph nodes may suggest malignancy.[9] Our patient had preoperatively signs suggestive of malignancy.

Two representative types of thyroglossal duct carcinoma exist, thyrogenic carcinoma and squamous-cell carcinoma. The former most likely arises from thyroembryonic remnants in the duct or cyst and the latter from metaplastic cuboidal cells.[10] The thyrogenic TDCa has two theoretical origins. The first is the de novo origin, based on the fact that ectopic thyroid tissue can be identified histopathologically in 62% of cases. This theory is further supported by the absence of a medullary carcinoma in the TDC as it arises from parafollicular cells, which are absent in the thyroglossal remnant.[11] The second is the metastatic origin which suggests that TDCa is a metastasis from an occult primary thyroid gland, as papillary carcinoma is multifocal in nature. Some physicians have suggested that a patent thyroglossal duct could act as a natural conduit for the spread of thyroid carcinoma. However, the metastatic theory seems less likely.[12,13]

The reported TDCa cases are distributed as the following: papillary thyroid carcinoma in 81.7% of the cases, mixed papillary–follicular carcinoma in 6.9%, squamous-cell carcinoma in 5.2%, follicular and adenocarcinoma in 1.7% each, and malignant struma, epidermoid carcinoma and anaplastic carcinoma in 0.9% each.[14] The type of the tumor in our case was a papillary thyroid carcinoma.

Metastasis to the regional lymph nodes has been reported in 7.7–12.9% of reported cases, much less frequent than in primary papillary carcinomas of the thyroid gland and local invasion rarely occurs.[15] Our patient has bilateral cervical lymph node metastasis and has no local invasion.

The differential diagnosis for these mid-line malignant lesions includes a primary thyroid carcinoma in the pyramidal lobe and metastasis to the Delphian lymph node. It is important to distinguish between these possibilities for selection of the appropriate surgical and adjuvant treatment as well as for prognostic reasons.[16]

The recognition of carcinoma arising from a thyroglossal remnant has both clinical and practical importance. On neck ultrasound, the carcinoma may appear as a mural lesion in the cyst, sometimes with microcalcification, or as a tumor invading the cyst wall.[17] A CT scanning or magnetic resonance imaging (MRI) is performed to confirm the diagnosis and to exclude other nodal masses. On CT scanning or MRI, carcinoma may be seen as a small peripherally based mass in relation to a cyst, a solid mass throughout the thyroglossal duct, or a complex invasive mass in the mid-line of the neck. CT scanning may reveal calcifications in cases of carcinoma, within either the primary carcinoma mass or the metastatic lymph node.[18] The presence of calcifications on ultrasound or CT is quite specific for carcinoma. Fine-needle aspiration is considered a safe, well-tolerated and cost-effective procedure, and it should be performed preoperatively for suspicious lesions.[19] In our case, fine-needle aspiration was positive for papillary carcinoma and oriented our management plan.

In managing patients with TDCa, it is important preoperatively to identify whether the normally functioning thyroid tissue is in its usual location or not. Thyroid scans and thyroid function studies have been suggested preoperatively.[9] Our patient had normally functioning thyroid tissue.

The definitive surgical management of TDCa is still debated. The first step should always be a wide complete Sistrunk

procedure which includes en-bloc resection of the thyroglossal duct remnant with the tract, the middle part of the hyoid bone (the body), and the soft tissues along the course of the thyroglossal duct to the level of the foramen cecum.[20] However, there is still controversy regarding the need to remove the thyroid gland in cases of TDCa.[21]

Total thyroidectomy permits the excision of those unsuspected thyroid carcinomas, enables a good follow-up, increases the sensitivity of radio-iodine scans, and makes it possible to use radioactive iodine treatment. Furthermore, thyroglobulin levels may then be used as a sensitive marker of tumor status.[13,19] Therefore, some authors recommend total thyroidectomy in patients with TDCa, even without a clinically evident thyroid mass due to concern about multifocal disease[16,20], such as in our case.

However, without an obvious lesion of the thyroid gland, most authors deny any benefit of removal of the total thyroid gland as the incidence of metastasis is very low in TDCa. Furthermore, even experienced surgeons cannot completely rule out the risk of vocal cord paralysis or hypoparathyroidism.[22]

Therefore, in the presence of a clinically and radiologically normal thyroid gland, the complete excision of TDCa via Sistrunk procedure without thyroidectomy is valid for patients younger than 45 years and with no history of low-dose neck irradiation in childhood, small carcinoma lesion of less than 1.5 cm in size, no invasion of the wall of the cyst, low-grade tumor, tumor-free margins, and no evidence of nodal or distant metastases.[5,16,20]

Total thyroidectomy is recommended for any suspicious lesions found in the thyroid gland during preoperative examination or intraoperative exploration, cases where the thyroid gland found to be nodular with a cold nodule in a thyroid iodine uptake scan, very close lesion to the thyroid gland, cyst wall invasion, large TDCa of more than 1.5 cm in size, enlarged lymph nodes, patients older than 45 years and a history of neck irradiation.[9,20]

Some authors advocated postoperative radioactive iodine ablation and/or hormone suppression to prevent the thyroid tissue stimulation, whether tumoral or normal, by L-thyroxin following total thyroidectomy.[23]

The incidence of cervical lymph node metastasis is lower than that of papillary carcinoma of the thyroid. Therefore, most authors agree that a neck dissection should be performed only in the presence of positive lymph nodes.[24]

Our patient underwent total thyroidectomy, Sistrunk procedure, bilateral level I–IV neck dissection, and postoperative I131 ablation and hormonal suppression with thyroxine based on the radiological and pathological evidence of cervical lymph node involvement.

Among the various types of neoplasia in TDC, a papillary thyroglossal duct cyst carcinoma has the best favorable prognosis. It is identical to that of papillary carcinoma of thyroid, with a rate of curability of 95%, while squamous-cell carcinoma has the worst prognosis.[7,25] In cases where thyroidectomy was not carried out, long-term monitoring is necessary to detect a latent thyroid cancer.[3] Thyroid stimulating hormone levels should be checked, and thyroid ultrasound should be included in the postoperative follow-up.[16] In our case, there were no clinical or ultrasonographic signs of recurrence or metastasis during the follow-up period, and the patient has been a disease free for 1 year after the operation.

4. Conclusion

Malignant lesions of thyroglossal remnant are rare. Its diagnosis can be easily missed. The majority is papillary thyroid carcinomas. For rapidly growing mid-line neck masses, imaging of the neck and fine needle aspiration cytology are required. Frozen section examination on TDC should be performed when there are any suspicious findings for malignancy. This type of carcinoma implies good prognosis, and lymphatic spread is rare.

Sistrunk procedure is adequate for most patients presenting with a clinically and radiologically normal thyroid gland. However, the more aggressive treatment, including total thyroidectomy, radioactive iodine ablation therapy, and thyroid-stimulating hormone suppression, is necessary in advanced lesions, clinically or radiologically suspicious synchronous neoplastic lesions in the thyroid gland, and cases of suspected metastasis. In case of lymphadenopathy discovered preoperatively or during the complementary examinations, dissection of the involved lymphatic chain is indicated.

Conflict of interest

Dr. Omar Sabra, Dr. Waleed M. Basha, and Dr. Moutaz Osman declare that they do not have any conflict of interest, financial or otherwise. The authors do not have any sponsor or funding source. This manuscript is original and it, or any part of it, has not been previously published; nor is under consideration for publication elsewhere. This manuscript has not been presented in any meeting.

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