Invasive papillary thyroid carcinoma appearing in a thyroglossal duct cyst: A rare case report

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Abstract

Thyroglossal duct cyst (TGDC) is the most frequent congenital anomaly of the thyroid gland. The existence of primary malignancy originating from this cyst is uncommon, accounting for less than 1% of all cases. Because of its rarity, there is no universal consensus regarding optimal treatment option. We present a case of a 59-year-old woman with a 25x20 mm mass in the anterior area of the neck that gradually increased in size over six months. A total thyroidectomy and Sistrunk procedure (SP) were performed. Postsurgery histologic evaluation confirmed papillary carcinoma of the TGDC invading adjacent muscle tissue.

Keywords*: thyroglossal duct cyst, papillary carcinoma, Sistrunk procedure.*

1. INTRODUCTION

Thyroglossal duct cysts (TGDCs) are the most common abnormalities in the formation of thyroid gland. They are found more than 75% of midline neck mass in children and around 7% of adults [1], [2]. During development, the thyroid gland descends from the foramen cecum of the tongue to its final position in the inferior neck, forming thyroglossal duct that maintains connected to its original position. The thyroglossal duct typically completely involutes between weeks 7 and 10 gestation [3], [4]. Remnants of the thyroglossal duct can give rise to the development of TGDCs [5]. Carcinomas arising from thyroglossal duct remnant cysts are extremely rare, occurring in fewer than 1% of cysts. The mainly histological type is papillary-type thyroid carcinoma, followed by mixed papillary-follicular carcinoma and other variants have also been described [6]. Given the rarity of this diagnosis, we present a case of TGDC carcinoma in a female patient, with normal thyroid gland.

2. CASE PRESENTATION

A 59-year-old female patient visited our hospital with a midline neck mass that was increasing gradually in size for six months. In recent weeks, she began to experience significant feeling of pressure associated with this mass. Physical examination revealed a 25x20mm mass that was hard, fixed and well-demarcated mass on the anterior neck. Clinically, a diagnosis of the TGDC (with a possible malignancy) was considered. Lab tests were within normal limits, including complete blood count and thyroid funtional tests. A neck and thyroid gland ultrasound were performed which showed a well-defined and heterogeneous lesion of 24x13 mm with solid component occupying 50% in the midline of the anterior neck area, between the hyoid bone and thyroid cartilage (Figure 1). There were no clinical and radiographic findings of thyroid gland disease and associated cervical lymph nodes.

Fine-needle aspiration (FNA) demonstrated classic features characteristic of papillary carcinoma (including papillary formations, nuclear grooves and nuclear pseudoinclusions) (Figure 2).

This patient underwent total thyroidectomy along with SP. Intra-operatively, a 2.5x1.5 cm cystic tumor with solid components was found. Intraoperative biopsy indicated a papillary carcinoma that infiltrates surrounding muscle tissue. Therefore, the tumor, the thyroid gland, the hyoid bone and the bilateral cervical lymph node were removed and sent for further evaluation. The postoperative pathologic report showed the papillary neoplasm appearing in the TGDC with the presence of normal thyroid follicles within wall of the cyst (Figure 3). Microscopic examination of the papillary neoplasm described the presence of true papillae with fibrovascular cores and the lining cells with nuclear features of papillary carcinoma (Figure 4B). The tumor cells infiltrated into the cystic wall and adjacent soft tissue (Figure 4A). Additionally, entire remaining thyroid parenchyma was confirmed that there were no abnormal findings. Consequently, a diagnosis of papillary carcinoma evolving from the TGDC was made.

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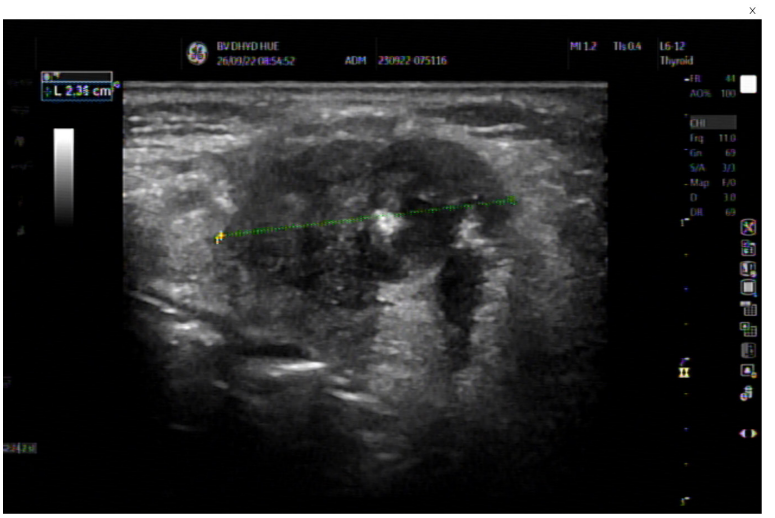

Figure 1. Ultrasound imaging of the neck detected a mass 24mm in diameter.

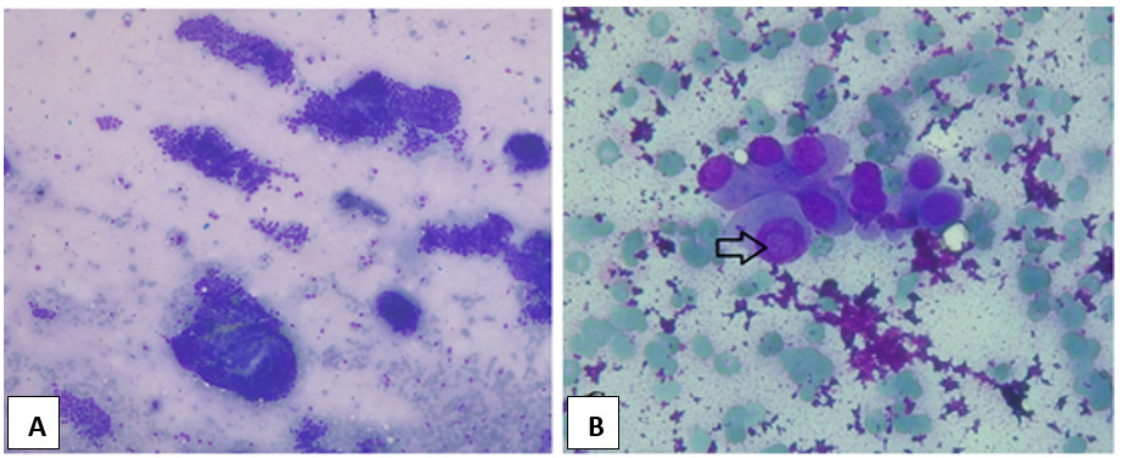

Figure 2. Fine needle aspiration biopsy findings showed papillary formations with nuclear pseudoinclusions (arrowhead) (Giemsa stain, x100, x400 for A and B, respectively).

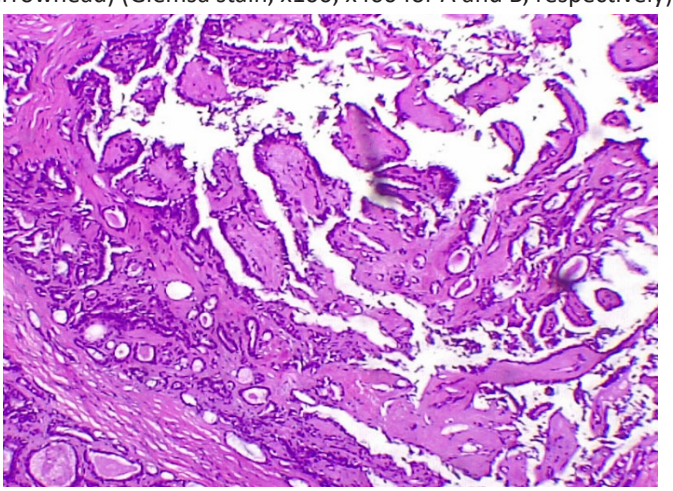

Figure 3. Microscopic image of the histopathology of the papillary carcinoma arising from the TGDC with the presence of normal thyroid follicles (red arrow) within wall of the cyst. (Hematoxylin and eosin stained slides at low magnification)

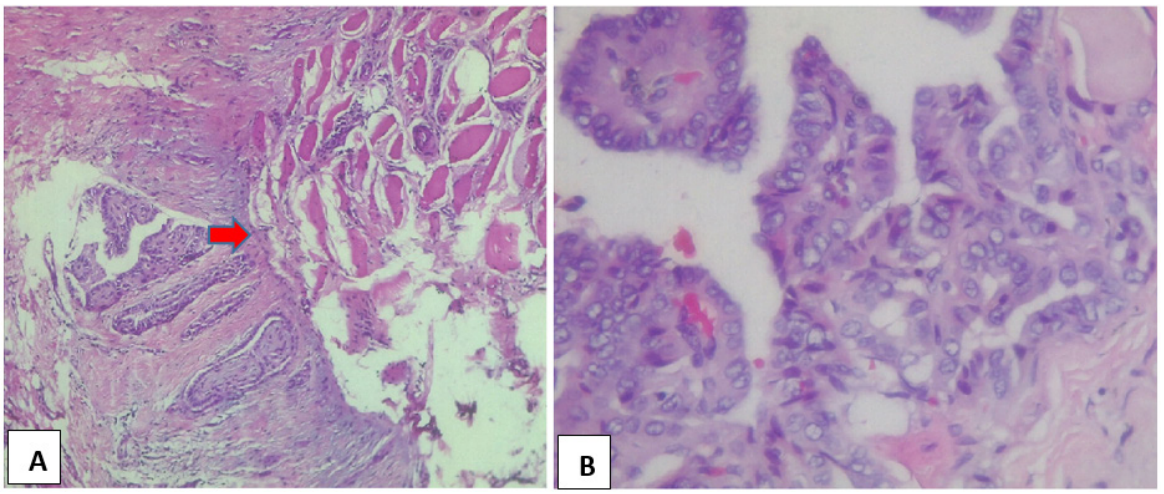

Figure 4. Hematoxylin and eosin staining showed the invasion of tumor cells into adjacent soft tissue (A, x100) and papillary architecture with central fibrovascular cores lined by typical cells of papillary carcinoma (B, x400).

3. DISCUSSION

TGDCs are seen in approximately 7% of the population. Normally, TGDC present as a palpable mass that can be in anywhere from the floor of the tongue to the lower part of the neck. Although most of these TGDCs are benign, the occurrence of malignancy was reported to affect about 1% of all cases [7]. The mean age of cases is in the fourth decade of life [8].

When it comes to the origin of carcinoma arising from TGDC, there are two hypotheses to explain this phenomenon. Most authors agreed that the malignancy arises *de novo* within the remnant of the thyroglossal duct wall whereas others advocated that carcinoma should be considered as a metastasis from a primary thyroid gland cancer. However, the former now is widely accepted because medullary carcinomas occurring in a TGDC has never been reported. (the cells are thought to derive from the ectopic thyroid tissue) [9], [10].

In general, it is tough to differentiate between a benign and a malignant lesion because these masses seem to manifest similar characteristics, as an indistinguishable asymptomatic tumor in the anterior neck area. In some cases, patients may present to the clinic because of pain, weight loss, or significant increase in size, which are symptoms of cancer. In particular, if the lesion is hard, fixed and irregular, clinician should suspect malignancy [7], [9].

The non-invasive imaging diagnosis techniques play an essential role in initial pre-operative diagnosis and further evaluation of adjacent structures. Some key features of carcinoma within the cyst can be detected by ultrasonography, which is mural nodule with micro-calcifications and/or abnormal regional lymph nodes. Moreover, the presence of a dense or enhancing mural nodule in cyst or the penetration of the mass to the surrounding tissues in CT scans is clearly recommended to malignancy. FNA is considered as a safe, reliable and cost-effective approach to assess midline neck masses before surgical intervention [7]. Nevertheless, Fine-needle aspiration can only make a precise diagnosis in about two third of all cases [11]. It is strongly believed that the ultrasound-guided FNA may noticeably reduce false-negative result because this procedure can improve the accuracy in cytological sampling [10].

Histologically, thyroid papillary carcinoma is the most frequent form (80%), followed by mixed papillar-follicular carcinoma and squamous cell carcinoma, about 8% and 6%, in turn. Other malignant types including oncocytic cell, follicular, and anaplastic carcinoma have been described, accounting for around 6% [6].

The criteria for the diagnosis of primary TGDC carcinoma were given by Widström et al., including: (1) thyroglossal duct carcinoma on histopathological examination (2) microscopic identification of normal epithelium lining and normal thyroid follicles in the wall of the thyroglossal duct remnant, and (3) no findings of malignancy of the remaining thyroid gland [12], all of which were met in our case.

Besides the diagnostic difficulties mentioned above, another main difficulty that clinicians have to handle is the choice of treatment method. Because of the rarity of this entity, there have been

no specific guidelines in terms of optimal treatment strategy [9]. Currently, SP, which consists of removal of the TGDC, the body of hyoid bone, and the soft tissues around the tract, is known to be adequate and curative for most patients [13].

According to Yun Mi Choi et al., patients who underwent a SP operation had significantly better outcome than those treated with simple cystectomy regarding 10 years survival rate, about 100% and 75%, respectively [7].

Some experts suggested a definitive surgical management depending on risk group stratification. Tharmabala and colleagues classified risk stratification into three categories: low-risk, moderate-risk and high-risk. Accordingly, low-risk patients should be observed while those who are classified into moderate-risk category have to be considered for total thyroidectomy, followed by hormonal suppressing treatment and radioactive iodine. Particularly, patients with high-risk characteristics should undergo regional lymph nodal dissection [12]. Plaza et al. proposed a definitive algorithm for the diagnosis, treatment, and follow-up of this lesion. They mentioned that SP is carried out as a stand-alone procedure without total thyroidectomy in low-risk patients. The low-risk factors identified include patients above 45 years of age, tumor size < 1.5cm, no cystic wall invasion, no history of neck radiation exposure, normal thyroid gland and the absence of cervical lymph nodes [6].

In this present case, considering that the patient grouped into high risk category (59 years old, the presence of infiltration of cystic wall and adjacent tissue), we performed total thyroidectomy along with SP.

The overall prognosis for papillary carcinomas originating within thyroglossal tract is the same as well-differentiated carcinomas of the thyroid gland, with a distant metastasis rate of less than 2% of cases while squamous cell carcinomas have the poorer prognosis [10], [11].

Although the prognosis for this entity is excellent, long-term follow up consisting of physical examination, ultrasound of thyroid gland and the surgical region, and whole body scintigraphy is compulsory [8], [10] .

4. CONCLUSION

TGDC carcinoma is uncommon. Because of the clinical indistinguishability between benign and malignant lesions, most cases are diagnosed incidentally on postoperative histopathological examination. All anterior midline neck masses should be carefully evaluated by physical examination, imaging techniques, and other advanced procedures to accurately assess the nature and select an optimal surgical management.

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