

3 Cases of Papillary Thyroid Carcinoma within Thyroglossal Duct Cyst – 1 Pediatric and 2 Adult Patient's

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ABSTRACT

Papillary carcinoma within thyroglossal duct cyst (TGDC) is rare. We present 3 cases of papillary thyroid carcinoma out of 75 cases (4%) of thyroglossal cyst operated in our hospital in last 13 years. All three cases were female. They belonged to age 73, 16 and 30. First case preoperatively found irregular hard mass, heterogeneity with calcification on Ultrasound and solid enhancing component, invading right neck strap muscle and prominent submental lymph nodes on CT scan. Second and third case had no preoperative distinguishing features of malignancy. All were managed with Sistrunk's operation and total thyroidectomy with or without lymph node dissection. For total thyroidectomy histopathology report, first case showed different tumour morphology likely due to second primaries. Second case showed papillary carcinoma. Third case showed lymphocytic thyroiditis. Cervical lymph node metastasis was noted in pathology report in first and second case. Hypocalcemia was noted in second and third case and managed with calcium supplement. All of them underwent radioactive iodine and subsequently thyroid hormone suppression therapy. They were being followed up for 4 years in first case and 10 years in second and third case at the time of review. There is no recurrence and 5 years survival is 100%. Thyroid carcinoma found in thyroglossal cyst is well documented. This should be discussed with patient in management of thyroglossal cyst. Prognosis is good especially with papillary thyroid carcinoma.

Keywords

Papillary Thyroid Carcinoma, Thyroglossal Duct cyst, Pediatrics, Adults.

Introduction

Thyroglossal duct cyst carcinoma is a rare malignant tumor. Its incidence is less than 1% in all age groups [1]. Since its first description in 1911, around more than 200 cases had been reported worldwide. It is even rarer in paediatric populations. There had been around 60 reported paediatric cases of thyroglossal cyst carcinoma [2]. Concomitant papillary carcinoma in the thyroid gland was extremely rare in paediatric patients, being reported in four patients in the literature [2].

During the fourth week of gestation, a ventral diverticulum of the foramen cecum of tongue is formed. It descends in the midline of neck as thyroglossal tract to the position of the normal thyroid in the base of the neck. Portions of the tract and remnants of thyroid tissue associated with it may persist. A thyroglossal duct cyst arises

as a cystic expansion of this remnant. There is debate to whether TGDC thyroid carcinoma arises as primary tumour from thyroid tissue in TGDC or secondary to metastasis from an occult thyroid gland carcinoma.

The prognosis is generally excellent. Because of its rarity, there are controversies in its pathogenesis, diagnosis and treatment. Accumulating knowledge of this rare disease via case reports may guide best management of this disease in the future.

Methods

75 cases of thyroglossal cysts managed in a regional hospital in Hong Kong in the last 13 years (2004 to 2017) were reviewed retrospectively, among which 3 cases of thyroglossal duct carcinoma were diagnosed.

Case Report 1

A 73 year old woman was referred to our centre for neck mass for 8 months. Clinically a 2.9x2.3x3.5cm Irregular hard mass

was noted. Ultrasound scan showed heterogeneity with some calcification. CT scan showed solid enhancing component in posterior aspect of the lesion. Posteriorly it abutted and involved the right neck strap muscle (Figure 1). Prominent lymph nodes were noted in submental region close to the cystic neck lesion. Tiny (2mm) hypodense nodule in left thyroid lobe might represent hyperplastic nodule.

FNAC was done twice (inadequate for diagnosis and cyst content). Incisional biopsy was performed with result showing carcinoma tissue present. Thyroid function test was normal. The patient underwent Sistrunk operation and complete thyroidectomy with level I and VI lymph node dissection in the same operation. Histopathology of the thyroglossal cyst showed papillary thyroid carcinoma. The tumour border was infiltrative. Invasion of the surrounding skeletal muscle and fatty tissue is noted. There was no definite lymphovascular permeation. 2 out of 4 cervical lymph nodes (around thyroglossal cyst) showed metastatic papillary thyroid carcinoma. There was also a small papillary thyroid carcinoma 3.5 x 3mm (papillary microcarcinoma) in right lobe of thyroid. There was no lymphovascular permeation.

Although both the right lobe thyroid nodule and the thyroglossal duct mass represent papillary thyroid carcinomas (PTC), they show different tumour morphology. We believe that they represent different tumours (double primaries, i.e. the larger, upper neck PTC arises within the thyroglossal duct primarily). The patient was treated with radioactive iodine and subsequently commenced thyroid hormone suppression therapy. She was being followed up for more than 4 years with no recurrence.



Figure 1: Solid enhancing component in posterior aspect of cyst invades the right neck strap muscle (Patient 1).

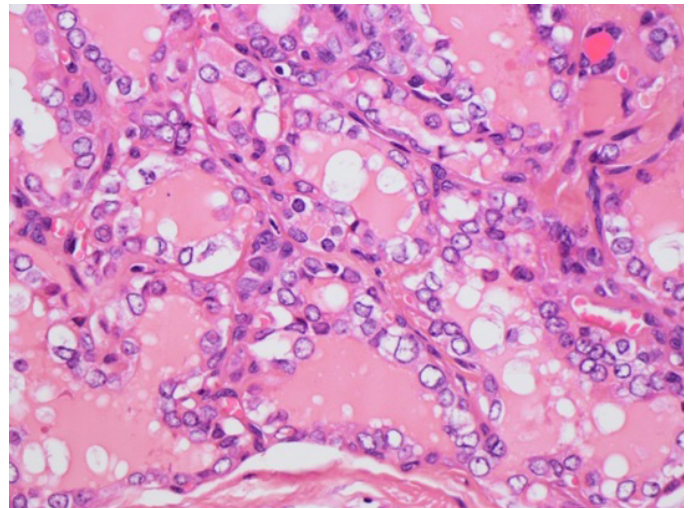


Figure 2: Thyroid PTC (Patient 1), enlarged pleomorphic crowded overlapping nuclei, irregular nuclear contour, nuclear grooves, nuclear pseudo-inclusion, pale to clear chromatin.

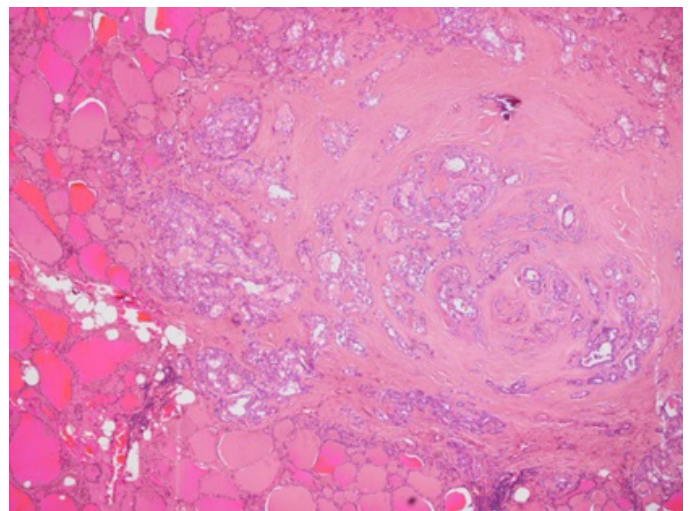


Figure 3: Thyroid PTC (Patient 1), infiltrative tumor proliferation with follicular pattern.

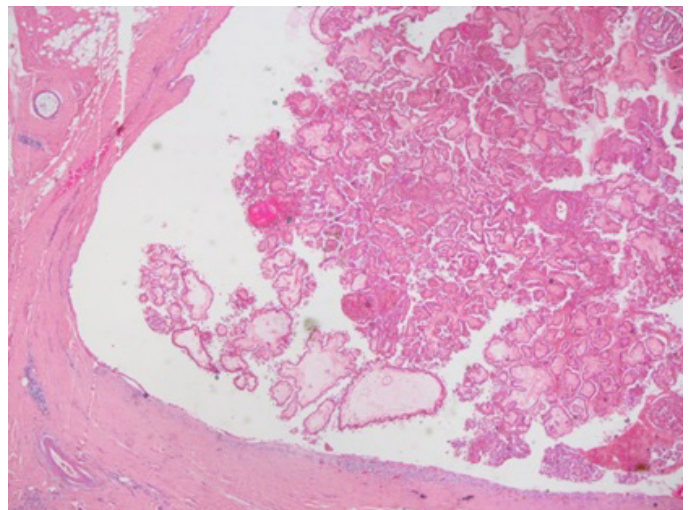


Figure 4: TGDC PTC (Patient 1), papillary proliferation within a cystic space.

Case Report 2

A 16 years old girl was referred to our centre for neck mass for 1 year. Thyroid function test, which was normal, was ordered as the only preoperative investigation. Sistrunk operation was performed. A 2cm thyroglossal cyst was removed. Histopathology of the thyroglossal cyst showed papillary carcinoma of thyroid. The resection margin was focally involved. Ultrasound scan showed multiple enlarged lymph nodes are seen in bilateral cervical region. Total thyroidectomy with level II, III, IV and VI lymph node dissection done. Histopathology report showed papillary microcarcinoma in the thyroid gland. One of the level III lymph showed metastasis. Postoperatively she was noted to have hypocalcemia managed with calcium supplement. The patient was treated with radioactive iodine. She had poor compliance to thyroid hormone suppression therapy due to poor tolerance to hyperthyroid symptoms. She was being followed up for around 10 years with no recurrence.

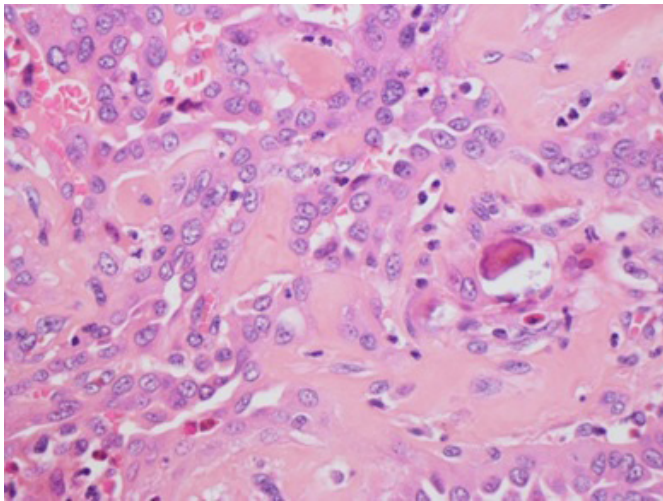


Figure 5: Psammoma body calcification (Patient 2).

Case Report 3

A 30 years old lady was referred to our centre for neck mass for 1 year. She also reported history of neck abscess in childhood with incision and drainage done. Preoperative ultrasound showed normal thyroid gland. Thyroid function test was normal. Sistrunk operation performed with 2cm thyroglossal cyst removed. Histopathology of the thyroglossal cyst showed papillary thyroid carcinoma.

All of the resection margins were unremarkable. Repeated Ultrasound postoperatively showed two 7mm and 5mm thyroid nodules at right middle and lower pole and two nodules 5mm in sizes at left lower pole and non-significant visible lymph nodes. Total thyroidectomy was performed. Histopathology report showed lymphocytic thyroiditis. No evidence of papillary thyroid carcinoma. Postoperatively she developed transient hypocalcaemia given supplement. The patient was treated with radioactive iodine and subsequently commenced thyroid hormone suppression therapy. She was being followed up for around 10 years with no recurrence.

Discussion

The true incidence of thyroglossal duct cyst carcinoma is difficult to define precisely. The reported incidence of carcinoma occurring in TGDCs ranges from 1 to 19.6% [3]. According to our experience, 4% of thyroglossal duct cysts were found to contain thyroglossal cyst carcinoma. TGDCCa affect women more frequently than men with a 2.3:1 female to male ratio observed in the literature [3]. All our three cases were female. Median age for development of papillary carcinoma in TGDC was reported to be 40 years [4], which is consistent with that in our series (range 16-73).

Most patients present with a mobile midline neck mass, clinically indistinguishable from its benign counterpart. The finding of a fixed or hard mass is a possible indicator of malignancy, which is the case in one of our patient. Otherwise thyroglossal cyst carcinoma is difficult to identify preoperatively. Majority are diagnosed from histopathology.

Preoperative FNAC were often used to aid diagnosis. However it had reported true positive rate of 53 % and a false negative rate of 47 % [3]. This is probably due to the cystic nature of thyroglossal cyst with poor cellular yield. Under ultrasound guidance, FNAC permits appropriate sampling of the solid part of the lesion, decreasing the false negative rate. Only one of our patient received FNAC as investigation which was not useful. As carcinomas are extremely rare in children, routine FNAC of all thyroglossal duct cysts may not be cost effective in them.

Imaging e.g. USG, CT or MRI occasionally showed suspicious features of carcinoma. The presence of solid components, calcifications, irregular/thick wall or invasions had been suggested as features of a carcinoma within a TGDC [3].

Histologically, the vast majority of TGDC associated malignancies are PTCs. Follicular carcinoma, squamous cell carcinoma, hürthle cell carcinoma and anaplastic carcinoma, etc. had been reported but was extremely rare [4]. Although seen in only about 5% of cases, squamous cell carcinoma of TGDC is considered as the only primary TGDC tumor by some authors, because other malignancy usually develop in ectopic thyroid tissue [5]. In our series, all three cases were PTCs.

There is debate to whether TGDC thyroid carcinoma arises as primary tumour from thyroid tissue in TGDC or secondary to metastasis from an occult thyroid gland carcinoma. We share the theory of those favoring a primary origin with two of our cases demonstrating primary thyroglossal cyst carcinoma.

Since TGDC malignancies are rarely recognized preoperatively, most TGDCCa are surgically managed initially as benign TGDCs with Sistrunk operation. The univariate analysis of Patel et al. [6], revealed that the only significant predictor of overall survival among patients with TGDC carcinoma was the extent of surgery for the TGDC. Patients treated with simple excision of the cyst had worse prognosis than those who had the Sistrunk procedure.

No clear consensus exists regarding further management after adequate excision of the cyst. There are controversies regarding whether total thyroidectomy and extensive lymph nodes dissection should be routinely performed. Proponents of total thyroidectomy suggested a potential synchronous thyroid gland involvement by thyroid carcinoma. Thyroidectomy also allows radio ablative iodine therapy and the use of serum thyroglobulin levels as surveillance. In our series, all three patients underwent total thyroidectomy. Concurrent papillary thyroid gland carcinoma was found in 2 patients with overall incidence 66.7%.

For paediatric patients, concomitant carcinoma in thyroid gland was considered extremely rare with 4 reported cases found in literatures [2]. This discrepancy between adult and paediatric patient may be accounted by the decreased radiation exposure in children [7]. Our series added one case of concomitant thyroid gland carcinoma in paediatric thyroglossal duct cyst carcinoma. Potential complications of total thyroidectomy should be considered in managing individual patients who included hypocalcaemia, injury to the recurrent laryngeal nerve as well as the need for life-long thyroid hormone supplements.

Regional lymph node metastases from TGDCCa have been described in 13-67 % of cases in literatures, with 2 out of 3 patients in our series showing regional lymph node metastases. Prophylactic lymph node dissection was not recommended on a routine basis, being reserved only for patients with clinically positive nodes [4].

Some authors advocated risk group stratification to guide further management after Sistrunk operation. Patients with <45 years of age with small tumors (cutoffs ranging from 1.0 up to 4.0 cm), classical histology, no extra capsular spread, no vascular invasion, negative margins, no nodal or distant metastases, and a normal thyroid gland by imaging studies, without previous history of neck irradiation are defined as low risk and adequately treated by Sistrunk procedure alone. Patients not meeting these criteria are categorized as high risk and treated more aggressively with the addition of total thyroidectomy with or without lymph node dissection, and consideration of RAI therapy.

The prognosis of TGD carcinoma seems to be excellent. In the review by Patel et al. [6], with a median follow-up of 71 months, the 5 and 10 year Kaplan Meier overall survival was 100 and 95.6%, respectively. Considering the multifocal nature of papillary carcinoma, regular follow-up is essential to detect early recurrence. Tumor related mortality have been reported only in few cases [8-10].

Conclusion

Thyroid carcinoma found in thyroglossal cyst is well documented. This should be discussed with patient in management of

thyroglossal cyst. Concomitant carcinoma in thyroid gland was considered to be extremely rare in paediatric patient but had been reported. Thyroglossal cyst carcinoma can be effectively treated by Sistrunk's operation +/- total thyroidectomy +/- lymph node dissection. There is no consensus to the optimal treatment after Sistrunk's operation and should be individualized in each patient.

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