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Incidental thyroid papillary carcinoma in a thyroglossal duct cyst – management dilemmas

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ABSTRACT

INTRODUCTION: Benign thyroglossal duct tract remnants typically thyroglossal duct cysts, (TDCs) are one of the commonest congenital childhood midline neck masses. Clinical presentation of persistent TDCs in adults is uncommon and the occurrence of incidental primary thyroid papillary carcinoma (TPC) in these cysts is rare.
PRESENTATION OF CASE: We report the case of a 32-year-old female with an asymptomatic midline neck mass compatible with a TDC that was excised by Sistrunk's procedure. Histopathological examination revealed an incidental primary intraluminal TPC arising from the wall of the TDC.
DISCUSSION: Management dilemmas regarding the roles for total thyroidectomy, regional lymph node dissection, radioactive iodine, and suppressive thyroxine therapy are reviewed in the context of relevant evidence based literature.
CONCLUSION: The occurrence of incidental TPC in a TDC is rare. Though Sistrunk's procedure is adequate treatment for TDC, based on low, moderate, and high risk stratification, recommendations for further management of incidental TPC in TDC is discussed.

1. Introduction

TDCs are one of the most common (75%) asymptomatic midline congenital neck masses in childhood.[1–3] Persistent TDCs occur rarely (7%) in adults.[1,3,4] Amongst all cancers, thyroid carcinoma is relatively uncommon (2%).[5] Reported cases of thyroid carcinomas occurring in TDC are extremely uncommon (1–2%).[1–3,6,7]

Though described in 1911 by Brentano, the first reported case in the English literature of a primary TPC in a TDC is by Owen and Ingelby in 1927.[1,3,7] Controversies exist even today regarding: (a) exact origin and (b) optimal management for incidental TPC in TDC.[1–3,7–9]

Majority of TPCs in TDCs are discovered as an incidental finding on pathological examination of the excised cyst specimen treated by Sistrunk's procedure based on the embryological development of the thyroid gland.[9–13] Simple local excision of TDC is no longer advised due to recurrences related to incomplete removal of the thyroglossal tract.[1,9] The rare discovery of an incidental TPC in a TDC raises questions regarding best practice guidelines for further management which still remains controversial and debatable.[1,9,13,14]

We report the case of an incidental primary intraluminal well-differentiated TPC arising in a TDC and discuss the potential roles for total thyroidectomy, regional lymph-node dissection, radioactive iodine, and suppressive thyroxine therapy as adjuvant therapy in the context of relevant evidence-based literature.

2. Presentation of case

2.1. Case report

A 32-year-old female who presented with a painless 3 cm swelling in the anterior aspect of the neck, clinically compatible with a TDC was treated by a standard Sistrunk's procedure.

Histopathological examination showed the presence of 0.8 cm papillary neoplasm arising from the cyst wall which contained entrapped thyroid follicles in keeping with a TDC (Fig. 1a). Microscopic examination of the papillary neoplasm showed the presence of complex fibrovascular cores lined by epithelium with characteristic nuclear features of well-differentiated low grade TPC (Fig. 1b). No additional histological subtypes were identified. Immunohistochemical staining with antibodies to low molecular weight keratin, thyroid transcription factor-1 (TTF-1), and thyroglobulin were strongly positive both in the papillary neoplasm and the residual trapped non-neoplastic thyroid follicles (Fig. 1c). The TPC arising in the TDC was completely excised with no evidence of invasion of the underlying cyst wall. No further surgical treatment was undertaken as this was considered an incidental well-differentiated classical thyroid papillary microcarcinoma. 1-year follow up was unremarkable.

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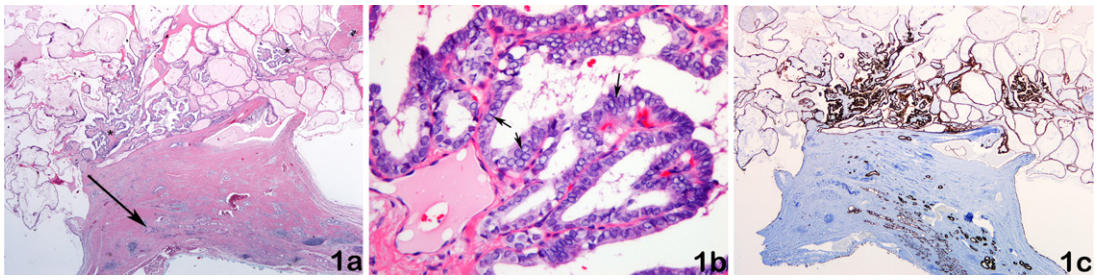

Fig. 1. (a–c) Photomicrographs of TPC arising in the wall of the TDC. (a) Haematoxylin and eosin stained slides at low magnification (lens objective 2×) show the presence of a papillary neoplasm arising from the cyst wall which contained entrapped thyroid follicles (black arrow→) in keeping with a thyroglossal duct cyst. (b) Haematoxylin and eosin stained slides at high magnification (lens objective 10×) confirms the presence of papillae with complex fibrovascular cores lined by the typical nuclei (ground glass nuclei with grooves and pseudo-inclusions [black arrow→]) of thyroid papillary carcinoma. (c) Immunohistochemical stained slides at low magnification (lens objective 2×) shows strong positive staining with antibodies to low molecular weight keratin, TTF-1, and thyroglobulin in the residual trapped thyroid follicles in the cyst wall and the adjacent papillary neoplasm.

2.2. Review of Management Dilemmas of TPC in TDC

A Literature search using PubMed, Medline, Scopus, Embase, and Google/Google Scholar limited to the English language using the search terms 'thyroid papillary carcinoma' AND 'thyroglossal duct cyst' AND thyroglossal duct remnants' AND 'management' was conducted.

3. Discussion

The thyroid gland is one of the earliest endocrine glands to develop in the human embryo.[10] It descends from the foramen cecum at the base of the tongue to the front of the neck in close relation to the hyoid bone via the thyroglossal duct. Thyroglossal duct remnants, most commonly a cyst, develops due to incomplete atrophy of the duct which usually occurs by the 7th foetal week of gestation.[13] TDC is the most common non-odontogenic cyst that presents as a neck mass at any point along the tract representing more than 75% of childhood midline neck masses. It is rare in the adult population (7%).[1,2,4,7,15] TDC clinically presents as a soft/firm/hard/fluctuant mass, that is generally movable in the midline of the neck. TDCs occur along the thyroglossal tract: thyrohyoidal (61%), suprahyoidal (24%), suprasternal (13%), and intra-lingual region (2%).[1]

The development of malignancy in TDC is uncommon and approximately 200 cases are reported in the English literature since its first description in 1911.[1,3,6,13]

In up to two-thirds of TDCs, thyroid follicles that persist in the cyst wall can rarely undergo neoplastic transformation.[3] TPC is the most common histological type of (80%) malignancy that occurs in TDCs. Infrequently squamous, follicular, mixed papillary/follicular, Hürthle cell and anaplastic carcinoma have been reported.[1,2]

The exact origin of TPC in a TDC is still debated as to whether it represents a 'metastatic lesion' from an occult primary TPC in the thyroid gland versus its 'de novo origin'.[1,2,7,8,12,14] The presence of entrapped thyroid follicles in the cyst wall, as seen in our case (Fig. 1a), and the persistent absence of medullary carcinoma occurring in a TDC in keeping with its embryology, strongly favours the 'de novo theory'.[1,2,16]

The diagnosis of a carcinoma in a TDC is usually incidental with no additional clues to suspect this preoperatively at the initial clinical presentation.[1] The diagnosis of primary TPC in a TDC is one of exclusion using the Widstrom criteria which include: (1) carcinoma should be in the wall of the thyroglossal duct remnant, (2) carcinoma must be differentiated from a cystic lymph node metastasis by histological demonstration of a squamous or columnar epithelium lining and normal thyroid follicles in the wall of the thyroglossal duct remnant, and (3) there should be no malignancy in the thyroid gland or any other possible primary site.[1,7] This latter criterion is highly debated as 11–45% of all thyroglossal duct remnant carcinoma cases have a synchronous thyroid carcinoma.[1,3]

Pre-operative thyroid scan, ultrasound guided fine needle aspiration cytology (US-FNAC) and CT/MRI imaging of the neck are gaining more popularity to enhance the accuracy of preoperative diagnosis.[2,17] Although, preoperative thyroid scans confirm the presence of ectopic thyroid tissue in 33% of TDCs, such scans have not been beneficial in the preoperative diagnoses of carcinoma in TDCs.[9] US-FNAC though simple, rapid, inexpensive with minimal risk complications is more reliable in diagnosing solid tumours rather than cystic lesions. Additionally, FNAC is not cost-effective due to the rarity of this malignancy and remains an inappropriate tool for routine use in children.[1,2,18] CT/MRI imaging of the neck are rarely indicated preoperatively unless there is a high clinical/FNAC suspicion of malignancy.[1,2,7,17] Imaging features such as a solid nodule with calcification/irregular margin/thick wall are suggestive of malignancy.[2,7] Ogawa et al. have suggested a possible utilization of three-dimensional computed tomography in providing accurate pre-operative diagnosis of TPC in a TDC.[19]

The dilemma involved with the further management of the discovery of incidental TPC arising in a TDC, is largely related to whether additional management protocols such as total thyroidectomy, regional lymph node dissection, radioactive iodine, and suppressive thyroxine therapy are warranted; as involvement of the thyroid gland has been reported in up to 11–45% of cases wherein a completion thyroidectomy was performed.[1,12] Such findings also raise the possibility of the presence of multifocal disease arising from synchronous TPC in the TDC and the native thyroid gland.[1,3,20]

In Patel et al's analysis of 57 cases of well-differentiated carcinomas in TDC, the only significant predictor for overall survival was the completeness of excision of TDC.[8] Patients treated with simple excision (10-yr survival 75%) had a worse prognosis than those with Sistrunk's procedure (100%).[1] It is hard to justify total thyroidectomy as the overall cure rate of a carcinoma arising in a TDC treated by Sistrunk's procedure is over 95%.[9]

Recently, prognostic risk group assessments are used to identify patients who would benefit from additional total thyroidectomy.[1,2,7,8] Plaza et al. identified high risk factors: (a) age >45 years, (b) past radiation exposure, (c) presence of tumour in the thyroid gland on radiological evaluation, (d) presence of clinical/radiological nodes, (e) tumour >1.5 cm in diameter, (f) cyst-wall invasion, and (g) positive margins on histopathological examination as indicators for additional total thyroidectomy with radioiodine and suppressive hormone therapy.[1] Additional factors that guide therapy/risk assessment include gender, and tumour characteristics such as histological grade/type, tumour focality, and lymphovascular invasion.[21]