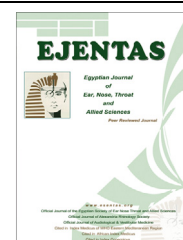




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## CASE REPORT

# Papillary carcinoma in thyroglossal duct cyst: An unusual case

Nitin Gupta \*, Arjun Dass, Mohit Bhutani, S.K. Singhal, Hitesh Verma, Mohit Bhutani, R.P.S. Punia

Department of ENT, Govt. Medical College and Hospital, Sector-32, Chandigarh 160030, India

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### KEYWORDS

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**Abstract** Thyroglossal duct cysts (TDC) are commonly encountered in ENT practice. They are treated by Sistrunk's operation in which the cyst along with the entire tract and part of hyoid bone is excised to prevent any recurrence. Very rarely TDCs may harbour malignancy. In that situation the management protocol is different. We present the case of a 33-year-old female with the unexpected finding of a papillary carcinoma arising in a submental TDC and is described with special regard to the rarity of the localization and the different options in the management strategy when carcinoma is found incidentally following surgery.

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## 1. Introduction

Thyroglossal duct cysts are the most common anomaly in thyroid development. They are twice as frequent as branchial cleft abnormalities and, in children, are second only to enlarged cervical lymph nodes as the cause of neck mass. Generally, duct cysts are benign, but 1% of cases may be malignant.<sup>1</sup>

Most cases of TDC carcinoma are diagnosed during the third and fourth decades of life, and rarely in children less than

14 years of age.<sup>2,3</sup> Papillary carcinomas are most commonly seen and have a favourable prognosis as cervical and distant metastasis is rarely seen. Further management of the patients who are incidentally diagnosed as carcinoma after surgery depends on the spread of tumour. Limited tumours are managed by regular monitoring while the more aggressive ones are managed by total thyroidectomy and radioiodine ablation.<sup>4,6</sup>

## 2. Case report

RK 33 year old female presented to the ENT OPD of our institute with complaint of painless, midline swelling over the upper neck for the last 8 months, insidious in onset and gradually progressive in size. There was no history of dysphagia, dyspnoea or hoarseness. There was no history suggestive of hypo or hyperthyroidism. The patient gave no history of radiation exposure. There was no other significant past or personal history. On examination, there was a single swelling in the

\* Corresponding author. Mobile: +919646121704.

E-mail address: [nitinent123@gmail.com](mailto:nitinent123@gmail.com) (N. Gupta).

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**Figure 1** Swelling in suprahyoid region.

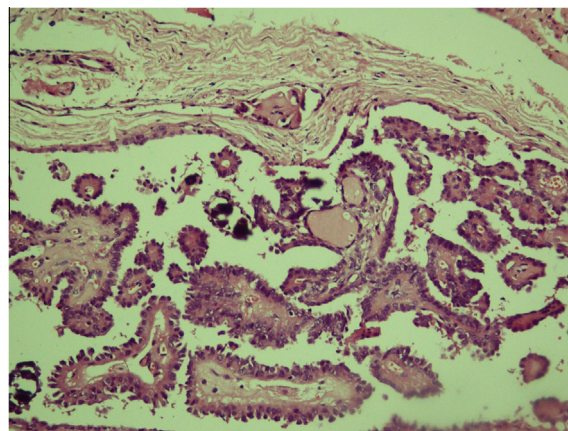
suprahyoid region in the midline, approximately  $4 \times 3$  cm in size, oval shape, well defined, and soft in consistency, overlying skin was normal in appearance and could be pinched easily (Fig. 1). It moved with deglutition and protrusion of the tongue. On palpation the swelling was non tender and soft in consistency. Overlying skin was normal in appearance and could be pinched easily. There was no palpable lymphadenopathy. The thyroid gland was clinically normal in shape and size. Rest of the systemic examination was normal. A provisional diagnosis of thyroglossal cyst was made. Routine blood investigations and thyroid function tests (T3, T4, TSH) were normal. USG neck revealed a  $2.7 \times 1.8$  cm cystic swelling in the suprahyoid region. Both lobes of the thyroid gland were normal. Aspiration cytology of the mass was suggestive of a thyroglossal cyst. Under general anaesthesia, the patient underwent Sistrunk's operation with removal of the complete thyroglossal tract and part of the hyoid bone between the lesser cornu along the tract. A horizontal incision was made over the skin and deepened to deep fascia. There was a sac of about  $3 \times 2$  cm just above the hyoid (Fig. 2). The tract was extending superiorly up to the base of the tongue. The cyst along with the tract and part of the hyoid bone between greater cornu was removed (Fig. 3). Complete specimen was sent for histopathological examination. Negative suction drain was kept which was removed after 72 h. The postoperative period was uneventful. Biopsy report revealed fibrocollagenic wall like tissue along with skeletal muscle bundles showing areas of fibrosis and many foci of papillary tumour lined with cuboidal epithelial cells with numerous Psammoma bodies, indicating papillary carcinoma (Fig. 4). On twelfth post-operative day, thyroid scan was performed. It did not show any uptake and the thyroid gland was normal. The patient has been on a regular follow up and has not shown any evidence of recurrence.



**Figure 2** Intra-operative photograph showing the cyst along with tract.



**Figure 3** Excised specimen showing the cyst, tract and hyoid bone.



**Figure 4** Histopathological picture (H&E X 200x) showing fibrocollagenic wall with cystic papillary tumour lined with cuboidal epithelial cells with Psammoma bodies.

Thyroidectomy was not performed because there was no history of previous irradiation and the carcinoma was limited to the cyst with no cervical metastasis. Post-operative thyroid scan showed normal functioning gland.

### 3. Discussion

The thyroid gland descends from the foramen caecum 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 5–10th gestational weeks. Incomplete atrophy of the thyroglossal tract or retained epithelial cysts, however, create the basis for the origin of a thyroglossal duct cyst (TGDC).<sup>2,7</sup> A thyroglossal remnant may be a cyst, a tract or duct, a fistula, or an ectopic thyroid within a cyst or duct. Failure of this tract to close predisposes to the formation of a thyroglossal cyst.<sup>3,8</sup> 60% of TGDC are located between the hyoid bone and the thyroid cartilage, 13% in the substernal region, 24% above the hyoid bone including the submental site and 2% are intra-lingual.<sup>3,9,10</sup> TGDCs are the most common congenital anomalies in thyroid development, but TGD carcinomas are extremely rare, with 90% of them originating from thyroid remnants.<sup>9,11</sup> Their cause is unknown

and there are no predisposing factors, i.e. neither clinical history nor physical examination can lead to a preoperative diagnosis.<sup>12</sup> Papillary types comprise 94%, and less than 5% are of squamous cell origin.<sup>6,13</sup> Among the various types of neoplasia in TDC, a papillary thyroglossal duct cyst carcinoma has the best favourable prognosis, with occurrence of metastatic lesions in fewer than 2% of cases, while a squamous cell carcinoma has the worst prognosis.<sup>3,7</sup> The incidence of cervical and distant metastasis is low, with, respectively, a rate of about 8% and 1.3%.<sup>11</sup> Generally, there are two theories to explain the thyrogenic origin of TGD adenocarcinomas. Firstly, the de novo theory is based on the fact that in 62% of cases, ectopic thyroid tissue can be identified histopathologically, and this is supported by the absence of a medullary carcinoma in the TGD as it arises from parafollicular cells.<sup>8</sup> The second is the metastatic theory which suggests that thyroglossal duct carcinoma is metastatic from an occult primary thyroid gland, as papillary carcinoma is multifocal in nature.<sup>6</sup> Due to these aetiopathogenetic controversies, strict criteria are required to diagnose a primary TDC carcinoma, as proposed by Joseph and Komorowski.<sup>5</sup> These are a thyroglossal remnant, ectopic thyroid nests within the cyst wall and a clinically normal thyroid gland.

Regarding the pre-operative diagnosis, finding a TDC carcinoma is unusual. As observed in the present case, patients are generally euthyroid, and the mass is often asymptomatic and not distinguishable from other more common, benign, histological types, in terms of growth, location, size and consistency.<sup>3</sup> Regarding the management, some authors believe that well-differentiated incidentally discovered thyroid carcinomas arising in a TDC and limited within their walls, without positive histological margins and cervical metastasis spread, in the presence of a clinically and radiologically normal thyroid gland, can be managed adequately by the Sistrunk's operation, thyroid suppression and strict long-term follow-up.<sup>6,8</sup> On the other hand, other authors have suggested a more aggressive approach characterized by the Sistrunk procedure, total thyroidectomy, post-operative radioactive iodine therapy and thyroid hormone replacement, based upon the observation that papillary thyroid carcinoma may metastasize through the thyroglossal duct remnant without a lesion being clinically detected in the gland itself.<sup>10,11</sup>

Thyroidectomy is recommended in cases where (a) the thyroid gland is found to be nodular, with a cold nodule in a

thyroid iodine uptake scan; (b) enlarged lymph nodes are present, or (c) a history of neck irradiation exists.<sup>4</sup>

#### 4. Conflict of interest

None.

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