Case Report

Malignant thyroglossal duct cyst with synchronous occult thyroid gland papillary carcinoma

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ABSTRACT

A 52-year-old male was referred to our department with complaints of a painless midline neck swelling. Clinico-radiological evaluation suggested a 6 × 5 cm thyroglossal cyst with non-palpable nodules in isthmus and right lobe of thyroid gland. FNAC of the thyroglossal cyst was suggestive of papillary carcinoma. He underwent Sistrunk's operation, total thyroidectomy, and central compartment neck dissection. Co-existence of papillary carcinoma of thyroid gland and thyroglossal cyst is a rare presentation and in this report, we describe our management and propose an evidence-based algorithm to assist decision-making in the management of these patients in future.

Key words: Papillary carcinoma, Sistrunk's operation, thyroglossal duct cyst carcinoma, thyroglossal duct cyst, thyroid cancer

Introduction

Thyroglossal duct cyst (TGDC) is the most common anomaly in thyroid development. Thyroid gland descends from foramen caecum to a point below the thyroid cartilage. It leaves an epithelial tract known as thyroglossal tract. The tract disappears during 5th to 10th gestational week. Incomplete atrophy of tract forms the basis of origin of the cyst. In this report, we present an adult male with papillary carcinoma of TGDC and occult papillary carcinoma of thyroid gland.

Case Report

A 52-year-old male presented to our department with a painless swelling in front of neck progressing slowly over the past 6 years. He complained of a rapid increase in size over the past 6 months. No history of irradiation. Clinical examination and CT evaluation suggested the presence of an irregularly-shaped complex TGDC measuring 6 × 5 cm with solid and cystic components. There was also evidence of fine calcification within the solid areas [Figure 1]. There was also a mildly enhancing nodule found in right lobe and isthmus of thyroid with fine calcification within the nodules. There was no cervical lymphadenopathy. Ultrasound-guided FNAC from the solid area of the TGDC suggested papillary carcinoma. Sistrunk operation for thyroglossal cyst done using a single transverse neck incision, and per-operative thyroid exploration revealed a hard nodule in the right lobe of thyroid with few enlarged paratracheal nodes. Since there was a strong suspicion of malignant thyroid nodule as well, total thyroidectomy and central compartment dissection was completed in the same sitting. Gross examination revealed TGDC, which was multiloculated and had solid and cystic areas (about 2 cm) [Figure 2]. There was a 0.5 cm nodule in the isthmus and a 1 cm nodule in the right lobe of thyroid gland. Microscopic diagnosis [Figures 3-5] was multifocal papillary carcinoma of thyroid and thyroglossal cyst with no metastasis in level VI nodes. Post op I^{131} scan showed 0.3% uptake in the thyroid bed and no evidence of functioning distant metastasis. Patient is on suppressive thyroxin therapy.

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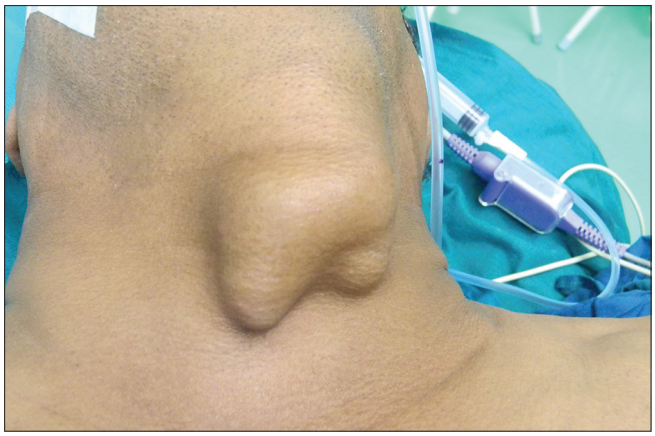

Figure 1: Clinical photograph showing the midline irregular-shaped neck swelling

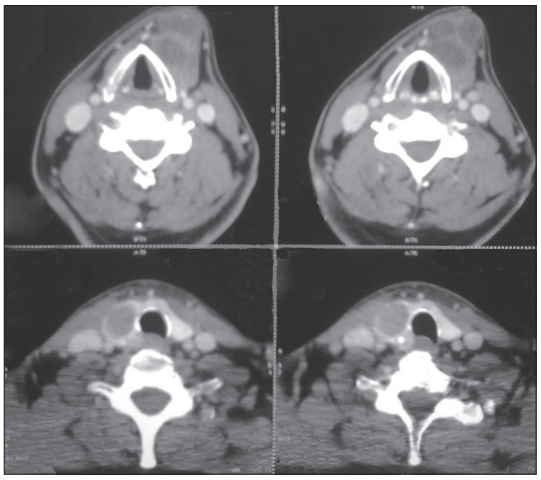

Figure 2: Contrast CT images showing the thyroglossal cyst with thick septae and a nodule in the right lobe of thyroid gland

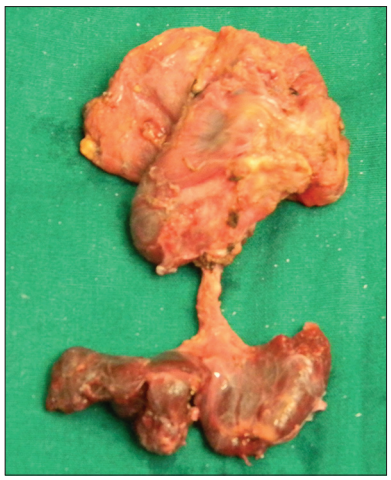

Figure 3: Photograph of the gross specimen showing the thyroglossal cyst attached to the thyroid gland through the thyroglossal tract

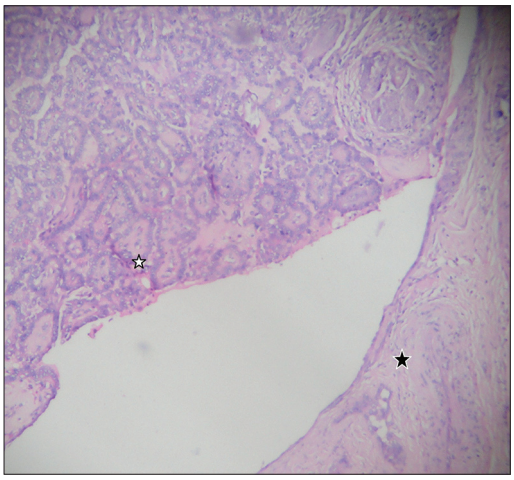

Figure 4: Light microscope appearance with the "black star" denoting normal cyst wall lining and the "white star" indicating papillary carcinoma of the thyroglossal cyst

Discussion

TGDC occurs in the midline neck and is diagnosed clinically. Majority is benign, but 1% may be malignant.[1,2] To date, around 215 cases of malignant TGDC have been reported, 80% of these being papillary carcinoma, 7% mixed papillary and follicular carcinoma, 5% squamous cell carcinoma, 1.7% follicular and adenocarcinoma and 0.9% anaplastic carcinoma.[3]

Since malignant TGDC is rare, diagnosis is made only if there is a strong clinical suspicion. Features that should arouse such suspicion include large or increasing size, hard, fixity, irregular shape, previous exposure to ionizing radiation. There is no role for routine FNAC of TGDC in the absence of suspicious features.

When malignancy is strongly suspected, pre-operative cytological and radiological evaluation aid in planning the extent of surgery. Usually, 2-3 aspirations are required for an adequate sample by FNAC, but yield is positive only in 66% of lesions since dilution by cyst contents can lead to hypocellular smears.[4,5] Scintigraphy may help differentiating a partially descended thyroid from a TGDC and identifying cold nodules in the thyroid gland. USG and CT help characterize the extent, presence of solid components, calcification, co-existing thyroid nodules, and cervical lymphadenopathy.[6]

In the vast majority of patients, diagnosis of malignant TGDC is made post-operatively after a Sistrunk operation. If histopathology of cyst is negative or shows low risk malignancy features like microscopic focus less than 1 cm without cyst wall invasion, follow up alone is adequate.[6,7] If there are high risk features like a neoplastic focus more than 1 cm or cyst wall invasion, then a total thyroidectomy is recommended as these lesions are aggressive. Further, total thyroidectomy allows long-term monitoring of thyroglobulin and I^{131} studies.[6] It has been shown that as many of 11% of patients with malignant TGDC may harbor foci of papillary carcinoma in the thyroid gland.[3]

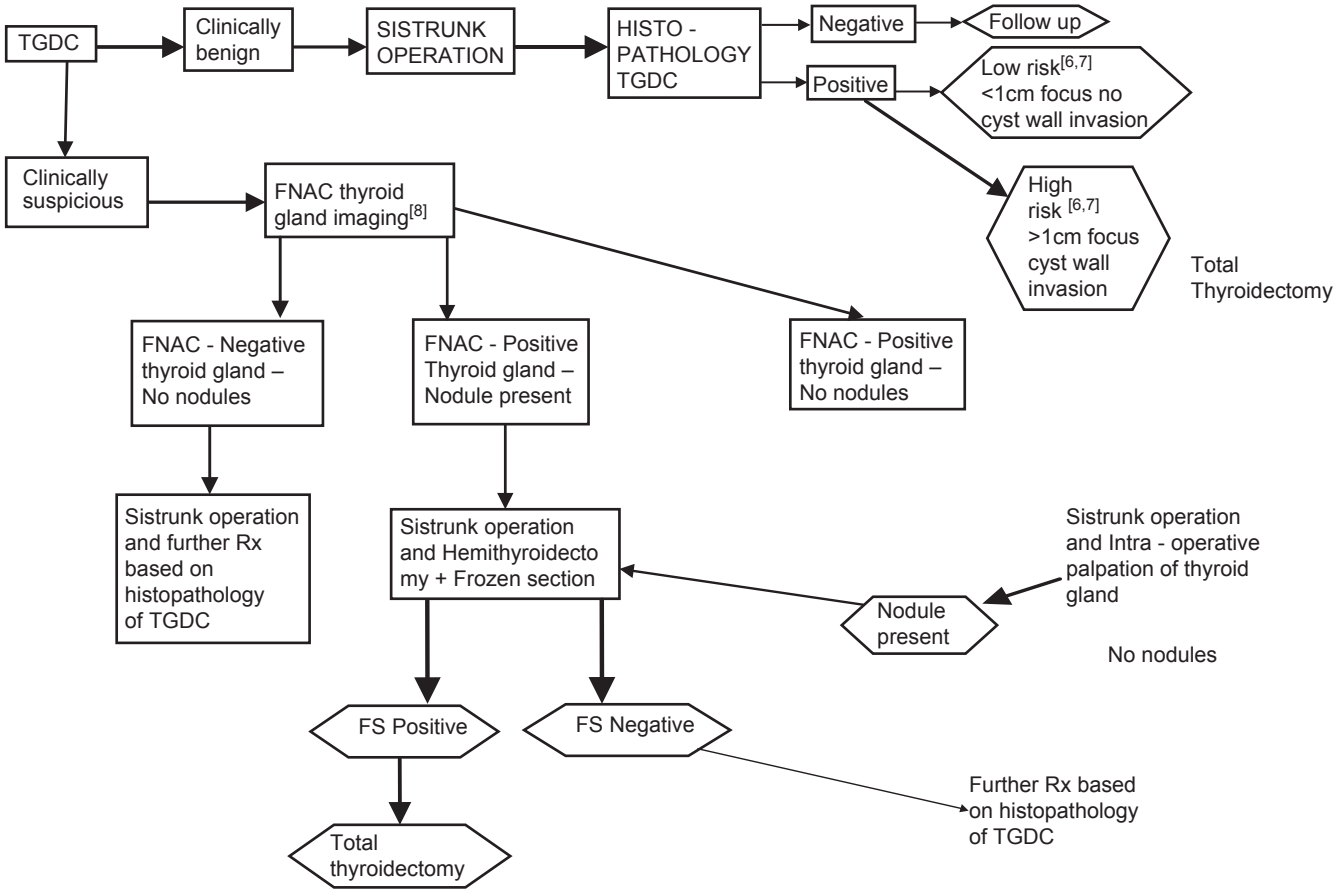

Figure 5: Management algorithm for malignant thyroglossal cyst

When both are present, it remains controversial whether the primary is the malignant TGDC or the thyroid gland.[3,8]

Whenever malignant TGDC is preoperatively diagnosed, pre-operative imaging and careful intra-operative evaluation of thyroid gland is performed to rule out clinically occult neoplastic nodules.[8] Presence of synchronous thyroid nodules warrants hemithyroidectomy and frozen section in addition to the Sistrunk operation. Further surgery is planned based on the frozen section report as one would manage a solitary thyroid nodule. There is no role for prophylactic lateral neck dissection.[8] Postoperative management of synchronous malignant TGDC and thyroid carcinoma is in line with the guidelines adopted for the management if differentiated thyroid cancer.[8]

To conclude, we present an evidence-based algorithm, constructed based on literature review, to aid decision-making in management of malignant TGDC.

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