PRIMARY PAPILLARY CARCINOMA OF MEDIASTINAL ECTOPIC THYROID TISSUE

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ABSTRACT

Primary carcinoma of ectopic thyroid tissue is extremely rare, with fewer than 50 published case reports. As such, set criteria for the diagnosis and treatment of this entity are yet to be established. We report the unusual presentation of a 56-year-old female with primary papillary carcinoma of ectopic thyroid tissue within the mediastinum. She was also found to have papillary microcarcinoma of the native thyroid. Differentiating between primary versus metastatic transformation of ectopic thyroid tissue is challenging. Features such as tumor size, blood supply, and anatomical separation are crucial to this analysis.

Key words: Ectopic, Thyroid, Papillary, Carcinomas, neoplasms.

INTRODUCTION

The thyroid gland develops from the floor of the primitive pharynx, tracking down the thyroglossal duct to reach its final midline position [1]. Aberrancy of this migration can result in ectopic thyroid tissue anywhere within or adjacent to this descent [2]. Ectopic thyroid tissue is an uncommon entity with a prevalence of 1 in 100,000-300,000 persons. Lingual ectopic thyroid tissue is most commonly observed, with other sites including the larynx, trachea, oesophagus and mediastinum representing approximately 7-10% of presentations [3]. Primary malignant transformation of ectopic thyroid tissue is extremely uncommon with only 43 published reports [4]. We present the unusual case of a 56-year old female with ectopic papillary thyroid carcinoma in the mediastinum and concurrent papillary microcarcinoma of the midline thyroid gland.

CASE REPORT

A 56-year-old women was referred by her general practitioner with a 2-month history of excessive weight loss, lethargy, and hair thinning. Apart from being an active smoker, she had no significant medical history. On examination, she was a thin lady with an obvious goitre that moves with swallowing. Her thyroid gland was palpable and soft. No cervical lymphadenopathy was found on examination.

Laboratory tests showed supressed TSH (0.20 mU/L), normal T4 (17 pmol/L) and negative thyroid antibodies. On USS, multiple thyroid nodules with a large cystic solid nodule in the right upper pole measuring 20 mm with calcifications. FNAC of the large cystic nodule identified a Bethesda system category 2 benign lesion.
Considering this classification and the biochemically euthyroid state, conservative management and 12-month follow-up was arranged.

At this point, the patient presented with a persistent goitre, however biochemical markers of thyroid function were normal (TSH = 0.40 mU/L, T4 = 15 pmol/L, T3 = 4.0 pmol/L). Repeat USS showed a large cystic structure below the left thyroid lobe that wasn’t visualised previously, in addition to the small cystic nodules. FNAC of this structure reported cystic fluid only. CT scan of the thyroid identified two adjacent cysts with an intervening narrow wall in the suprasternal notch (not seen on USS), measuring 32 mm x 25 mm in the axial plane with a height of 37 mm in the coronal reformat (Figure 1). The thyroid gland also showed several small low density cysts in both lobes measuring up to 5 mm with a right lobe lower pole mixed density nodule of 12 mm. No other soft tissue abnormalities were seen. Given the persistent goiter, the patient was booked for a non-urgent total thyroidectomy.

Total thyroidectomy was performed via routine technique. During the procedure, the large mediastinal mass was mobilised and found to be completely distinct from the midline thyroid tissue. Blood supply for the mediastinal mass appeared to derive from thoracic vessels. Postoperative recovery was uneventful, and the patient was put on daily thyroid and calcium supplements.

Histopathologic examination of the right mediastinal mass revealed an encapsulated cystic nodule containing foci of ectopic thyroid papillary carcinoma. The margin of the cyst was free of malignant cells. Examination of the right thyroid lobe also identified a 2 mm non-encapsulated papillary carcinoma confined to the thyroid. Lymph nodes were free of tumour.

As per recommendations from the radiation oncologist, the patient was treated with adjuvant low dose radioactive iodine. At her latest outpatient appointment, no abnormalities were detected on clinical and biochemical examination.

**DISCUSSION**

Carcinoma of ectopic thyroid tissue is extremely rare and represents less than 1% of all thyroid carcinoma. The most common malignant subtype of this aberrant tissue is papillary carcinoma. Even more unusual is primary malignant transformation of ectopic thyroid tissue (3, 4). Our case highlights the identification of ectopic thyroid papillary carcinoma within the mediastinum, and the diagnostic challenge of distinguishing whether this represents a primary carcinoma of ectopic tissue within a cyst, or metastases from a primary papillary carcinoma of the right thyroid lobe.

To our knowledge, three other cases of ectopic thyroid papillary carcinoma within the mediastinum have been reported [5-7]. Only one of these cases also detailed a coexisting carcinoma of the orthotopic thyroid [7]. In a previously published case of ectopic papillary thyroid carcinoma in a lateral neck cyst, suggestions were made to discriminate between primary versus metastatic transformation of the ectopic tissue after a 0.5 mm papillary carcinoma of the native thyroid was subsequently identified on histology. According to Xu et al., absence of lymph node parenchyma in the cyst, negative lymph nodes, no extracapsular invasion and complete anatomical and vascular separation all support a primary ectopic aetiology [8].

Interestingly, papillary thyroid microcarcinoma (lesions smaller than 1 cm) is found in 10-30% of all autopsies (for unrelated death) and thus considered to be benign [9]. Indeed, in the case reported by Xu et al. [8] the solitary papillary microcarcinoma in the native thyroid was considered an incidental finding. The 2 mm lesion of the native thyroid reported in our case is also likely to be unrelated to the ectopic thyroid carcinoma.

Given the infrequency of this pathology, no evidence-based guidelines exist for the treatment of primary ectopic thyroid carcinoma within the mediastinum [2]. Total thyroidectomy been performed in several other...
cases of ectopic carcinoma as it aids to exclude native thyroid tissue as a source of primary malignancy [4-8]. In our case, the thyroid was removed to exclusively to treat a persistent goitre rather than the carcinoma which was only identified later on histopathology. As such, more research is required to determine the optimal management for this phenomenon from both a medical and surgical perspective.

CONCLUSION
Ectopic thyroid within the mediastinum is a rare entity. Even less common is primary malignant transformation of this tissue. Difficulty arises in demarcating primary versus metastatic transformation of ectopic thyroid, however several criteria have been put forth such as anatomical separation and thoracic blood supply. As evidenced by this case report, microcarcinoma of the native thyroid does not preclude the diagnosis of true malignancy of ectopic tissue.

STATEMENT OF HUMAN AND ANIMAL RIGHTS
All procedures performed in human participants were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. This article does not contain any studies with animals performed by any of the authors.

CONFLICT OF INTEREST
No financial or other conflicts of interest need to be declared for this case report.

ACKNOWLEDGEMENTS
None

REFERENCES

Cite this article:

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