Ectopic mediastinal thyroid presenting as metastatic cancer

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Introduction

Thyroid cancer, the commonest endocrine cancer, is rare in comparison to other cancers (<1%). It is still rarer in ectopic thyroid tissue. Its rarity makes it one of the least suspected primary culprits in individuals presenting with metastatic disease and an unknown primary. In contrast to other cancers, thyroid cancer is almost always curable. Prognosis is generally very good with mortality rate of around 1.5 %.

We present a case of a lady with a history of treated thyroid cancer who presented with metastatic follicular thyroid cancer probably arising from ectopic thyroid tissue. Apart from its rarity, this case also serves to highlight the importance of suspecting a primary thyroid malignancy in metastatic disease and the significance of regular long term monitoring for disease recurrence and use of thyrotropin (TSH) suppressive doses of thyroxine in those individuals with a history of thyroid cancer.

Case presentation

A 73 year old lady presented with symptoms of progressively worsening shortness of breath and unintentional weight loss of two stones over the preceding six months. She had a history of well controlled chronic obstructive airways disease and a left thyroid lobectomy 28 years previously for localised follicular thyroid carcinoma (FTC).There was no previous history of upper body irradiation. There was no family history of thyroid cancer.

She was found to have ascites on examination and pleural effusion on a chest x-ray. Given the history of weight loss, a CT scan of the chest, abdomen and pelvis was performed. This revealed a heterogeneous cystic-nodular mass (Figure 1) measuring 20 X 14 mm between the brachiocephalic and left subclavian artery, significant amount of ascites, multiple metastatic liver lesions and omental thickening. No primary malignant lesion was identified. The ascites was drained and the omentum biopsied under ultrasonic guidance. In view of extensive metastases, her treatment was deemed to be palliative. When symptomatically better patient was discharged home according to her wishes. Two weeks after discharge, it was learnt that she had passed away peacefully at home.

Around the same time, we received the result of a serum thyroglobulin level which was 127 µg/L (normal<40µg/L). Histology from the omental biopsy showed metastatic adenocarcinoma with diffuse staining for immunocytochemical markers such as cytokeratin 19 & galectin 3, HBME 1 suggestive of thyroid follicular epithelial primary tumour.

Looking through her old notes, we noted that a CT scan of the chest that was arranged by a chest physician two years ago to investigate breathlessness had shown a similar lesion in the superior mediastinum. A thyroid uptake scan that followed had shown normal uptake in the right thyroid remnant and a high uptake in this structure (Figure 2), suggestive of ectopic thyroid tissue with increased activity. No further action was taken at the time.

Twenty eight years ago, after she had undergone left thyroid lobectomy, she was followed up by the ENT specialists for four years and then discharged to the care of her GP. Thyrotropin (TSH) suppression was not undertaken as she was deemed to be low-risk. Eight years later, she was started on levothyroxine therapy as she was found to become progressively hypothyroid over this period. The TSH was never suppressed and in the last 3 years prior to her presentation, it was not consistently maintained within the normal reference range of 0.30 to 6.00 mU/L ((8.9 mU/L, 4.5 mU/L & 30 mU/L respectively),

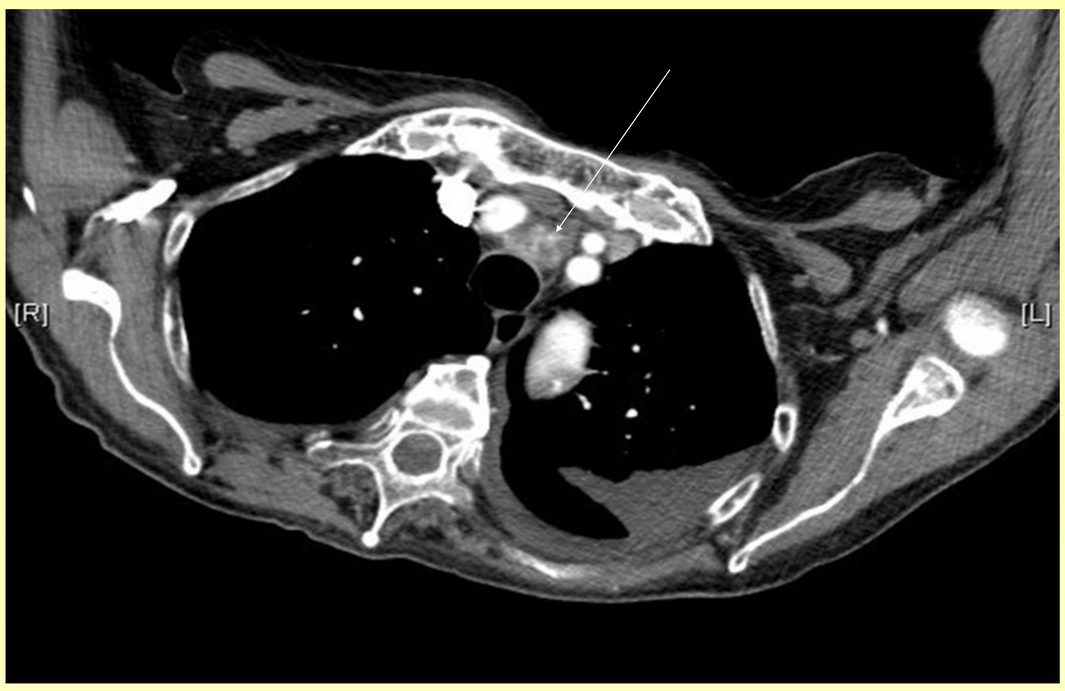

Figure 1. A heterogeneous cystic-nodular mass (white arrow) measuring 20 x 14mm between brachiocephalic artery and left subclavian artery

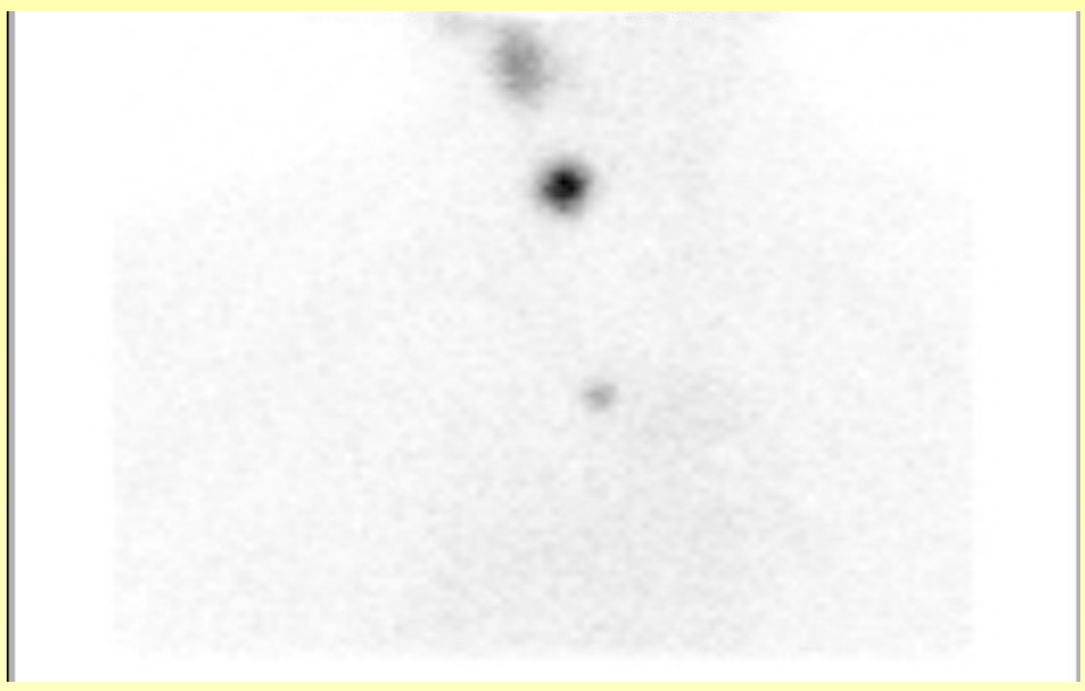

Figure 2. Radioiodine uptake scan showing normal uptake in the right thyroid remnant and increased uptake (hot spot) in the mediastinal tissue.

Discussion

Ectopic thyroid tissue is an uncommon developmental abnormality, with a prevalence of 1 per 100000 – 300000 people (1 per 4000 – 8000 patients with thyroid disease).[1] The mediastinum is the 5th most common site of ectopic thyroid tissue after lingual (90%), submandibular, thyroglossal cyst and intratracheal. Intrathoracic ectopic thyroid accounts for only 1% of mediastinal tumours. Primary thyroid carcinomas arising from ectopic thyroid tissue are extremely rare.[2] Most of these are of papillary origin, but follicular carcinoma has been reported in one case.[3]

This lady was found to have metastatic follicular thyroid carcinoma. The two possible sources of metastases could have been either the thyroid remnant, or a new cancer arising de novo in the ectopic thyroid tissue. The latter is a definite possibility in this case due to the following reasons: 1) No reported change in the size and structure of the remnant by the patient over the last 28 years, 2) The uptake in the thyroid gland remnant on the ^{131}I uptake scan was normal, whereas it was high in the ectopic thyroid. Unfortunately, the diagnosis cannot be confirmed due to lack of histological evidence. If this were to be confirmed, this would have been an extremely rare case - especially given the fact that she has already had a malignant tumour in the orthotopic thyroid gland in the past.

We would also like to highlight another aspect of this case. Since this lady had undergone only a lobectomy for thyroid malignancy with no subsequent radioiodine ablative therapy or external beam radiotherapy, we could speculate that she was probably a ‘low risk’ patient for recurrence of the malignancy (age < 45 years, female and tumour size < 2 cm according to current guidelines). Even so, the current recommendation is that these patients should have their TSH levels suppressed to 0.1 – 0.5 mU/l.[4] All individuals with a history of thyroid malignancy should have life-long surveillance for recurrence by means of annual clinical examination, annual TSH and thyroglobulin measurement, and diagnostic imaging and FNAC when indicated.[4] This is because late recurrences are not rare, and can be treated successfully. They should also be regularly monitored for the long term side effects of thyroid suppressive therapy, hypocalcaemia, and late effects of radioiodine ablative if they have received this. For these reasons, follow up of these patients is best supervised by an endocrinologist. This lady was only followed up for four years, by ENT surgeons.

When the ectopic thyroid lesion was discovered two years ago, a timely referral to an endocrinologist would have been beneficial, given her history. Further investigations such as an excision biopsy or FNAC of the ectopic lesion would have triggered whole body ^{131}I or ^{18}FDG-PET scanning to highlight the extent of the disease. Total ablation with I^{131} may have potentially destroyed remnant thyroid tissue, reduced the disease burden & improved survival.

References

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