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Ectopic Thyroid Tissue Presenting as a Carotid Body Paraganglioma. A Clinical Case Report.

Running title: Ectopic thyroid tissue presenting as a carotid body paraganglioma.

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Abstract

Ectopic thyroid tissue is a rare developmental abnormality, typically presenting with midline ectopia due to incomplete embryological migration via the thyroglossal duct. The presence of ectopic thyroid tissue lateral to the midline is very rare, accounting for 1-3% of all ectopic thyroid tissue. Clinically these lesions are often mistaken for other diagnoses including enlarged lymph nodes or metastatic tumours.

We present a case of lateral ectopic thyroid tissue presenting as a presumed carotid body paraganglioma in a middle-aged woman. The patient presented with a slow-growing lateral neck mass, which was clinically and radiologically consistent with a carotid body paraganglioma. She subsequently underwent tumour excision after which histological analysis identified ectopic thyroid tissue with no evidence of malignancy.

Background

Embryologically, the thyroid gland begins development at the third week of gestation,¹. It is derived from the fusion of a large median anlage and two lateral anlages. The median anlage, which produces the follicular cells (making up the majority of the thyroid parenchyma), arises first as an endodermal growth from the second pharyngeal arch,². This growth is known as the thyroid diverticulum. The thyroid diverticulum then descends caudally down the midline until the eighth gestational week when it reaches its final destination; sitting anterior to the trachea and larynx,³. This line of migration is known as the thyroglossal duct.

The two lateral anlages, which produce the parafollicular C cells, are derived from the fourth pharyngeal pouch. They migrate to fuse with the posterior surface of the median anlage during its descent in the fifth week of gestation,^{2,4}.

In contrast, the carotid body, which sits within the adventitial layer of the carotid bifurcation, is derived from the mesoderm of the third pharyngeal arch and the neural crest ectoderm. Tumours of the carotid body arise from the paraganglioma cells. There is no known relationship between the carotid body and the thyroid gland,⁵.

Ectopic thyroid tissue (ETT), defined as thyroid tissue not located anterolaterally to the second - fourth tracheal cartilages,⁶, was first described by Hickman in 1869, when a new-born suffocated as a result of a lingual thyroid causing upper airway obstruction,⁷. Since then, multiple cases of ETT have been reported. Typically, as with Hickman's case, the ETT is found in the midline due to incomplete descent of the median anlage. The presence of ETT lateral to the midline is rare, likely due to dysembryogenesis in the migration of the lateral anlage,⁴. Due to their rarity these are often clinically misdiagnosed as enlarged lymph nodes, metastatic tumours,⁸, or as in our case, carotid body paragangliomas. Even with modern imaging it can be difficult to differentiate ectopic thyroid tissue from other pathology such as carotid body

paraganglioma and therefore many are only diagnosed after surgical excision and histological analysis,⁹.

Case Presentation

A woman in her fifties presented with a right sided lateral neck mass, which had been present for a few months and was associated with mild discomfort, but no disturbance to speech or swallowing, and no symptoms of thyrotoxicosis. Examination revealed a solitary, firm mass in the right anterior triangle, with no overlying skin changes and no lymphadenopathy, consistent with a provisional clinical diagnosis of carotid body paraganglioma. Ultrasound scan (Figure 1) of the neck identified a “well-defined hyperechoic mass at the right carotid bifurcation”, and a subsequent CT (Figures 2 and 3) revealed a 51mm hyper-enhancing mass, straddling the carotid bifurcation, sitting 17mm inferior to the skull base, described as “consistent with a carotid body tumour”. There was no evidence of any pathological lymphadenopathy and the upper aerodigestive tract appeared normal. The presence of a thyroid gland in the normal orthotopic position was also noted. Based on these findings, the mass was initially diagnosed as a carotid body paraganglioma, and the patient was scheduled for endovascular embolization followed by surgical resection. All pre-procedure haematological and biochemical investigations were normal. Previous thyroid function tests were within normal ranges.

During the endovascular embolization the angiogram showed modest hypervascularity relating to the mass. The 3 largest feeder vessels were embolised and this resulted in a 70-80% reduction in the lesions “blush”.

Intra-operatively the tumour was encountered at the level of the carotid bifurcation, and complete excision was performed. The tumour encased the carotid arteries but was easily resectable, with no invasion of the adventitia. An enlarged jugulodigastric lymph node lying superficial to the tumour was also excised.

Histological analysis showed lobulated thyroid with marked necrosis and haemorrhage in keeping with embolization. There were no features to suggest malignancy from the primary lesion or lymph node. The final diagnosis of ectopic thyroid tissue was made.

During the initial post-operative period the patient developed left arm weakness. This was initially investigated and treated as a potential cerebro-vascular accident, however a CT and MRI showed no evidence of any infarct. There were no further concerns from the vascular team prior to discharge. Total length of stay was 10 days.

Follow-up at four weeks confirmed the patient was recovering well, with no further neurological symptoms and recovery of most of her arm function. The wound had healed well and there were no symptoms of hypothyroidism and thyroid function tests were normal.

Discussion

Ectopic thyroid tissue is a rare embryological abnormality with a prevalence of 1/100,000-300,000,¹. Typically, it occurs along the median path of migration, as a lingual thyroid (80-90% of cases), or thyroglossal cyst (5 to 15%), secondary to incomplete descent or obliteration of the thyroglossal duct,⁴. Ectopic tissue lateral to the midline is rare, accounting for only 1-3% of all ETT. The most accepted hypothesis for its development is failure of the lateral anlage to fuse with the medial anlage and non-migration of the gland,^{4,5,10}. Mutations in a number of transcription factors (NKX2-1, PAX8, FOXE1 and TSHR), associated with definition and migration of the developing thyroid, are recognised causes of thyroid dysgenesis,¹¹. These are often tested for in patients with congenital hypothyroidism, which is associated with thyroid dysgenesis and ectopy. However, previous biochemistry showed our patient to be euthyroid and therefore this was not examined.

Where reported, 70% of lateral ETT are located in the submandibular region or, less commonly, near the carotid sheath⁴. There have been only six previously reported

cases of ETT at the carotid bifurcation,^{5,9,12-15}. Other unusual locations of ETT include mediastinum, chest, and abdomen,¹².

The clinical presentation of our patient was similar to those in previously reported cases; a slow-growing, asymptomatic mass. However, on retrospective review, it was noted our patients pre-operative CT scan identified a thyroid gland located in the normal orthotopic position. In cases of lateral ETT only 41% have also had a thyroid gland in the normal orthotopic position, however it is noteworthy that a further 24% had another site of thyroid tissue located elsewhere such as a lingual thyroid. In comparison, a second site of thyroid tissue was seen in only 25% of those with ectopic lingual thyroids,⁴.

Given this, it is important to consider the functional effects when excising ETT. It has been reported that in 70-80% of cases, ETT may be the only functional tissue,⁵ and therefore excision can lead to irreversible hypothyroidism. This was not the case in this instance. The simultaneous finding of lateral ETT and a normally located functional thyroid gland is extremely rare,^{5,16}.

Even with modern imaging techniques it can be difficult to discriminate lateral ETT from other diagnoses, including carotid body tumours, and therefore management is often via first-line excision biopsy. Consequently, ETT is often only diagnosed after histological analysis,^{4,9}. When ETT is suspected, Technetium-99 or Iodine-131 scintigraphy are not only important diagnostic investigations, but may also demonstrate the presence or absence of normally located or other functional thyroid tissue,^{4,16}. Somatostatin receptor scintigraphy, such as octreotide scans, or 68Ga-DOTATATE PET scans are often used to diagnose neuroendocrine tumours, including paragangliomas, however, uptake of octreotide and 68Ga-DOTATATE uptake within thyroid goitres, and in a case of ETT, has also been reported,^{12,17}.

Albeit rare, ETT is an important differential to consider given the risk of irreversible hypothyroidism. Others have highlighted the importance of combined investigations such as CT, MRI, scintigraphy (somatostatin receptor, technetium-99 or iodine-131),

and fine needle aspiration cytology (FNAC) to give a higher sensitivity and specificity when investigating a lateral anterior triangle neck mass,^{4,12,16}.

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Key Messages

- Ectopic thyroid tissue presenting at the carotid bifurcation is a rare phenomenon and may clinically mimic carotid body tumours.
- Lateral ETT typically arises due to dysembryogenesis of the lateral anlage.
- ETT is an important differential in the presence of anterior triangle neck mass and thorough investigations including scintigraphy or FNAC should be considered.
- ETT may be the only functional thyroid tissue, therefore excision may lead to irreversible hypothyroidism. If diagnosis is made post-operatively it is imperative the patient undergoes follow up thyroid function tests.

Declarations

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Conflict of Interest: The authors declare that they have no conflict of interest.

Patient Perspective

I found the lump on the right-hand side of my neck whilst watching a training course on my laptop, I had rested my chin on my hands. I felt a slight swelling on my neck below my jawline. My GP looked at the lump and thought that it could be a swollen lymph gland as I had recently had Covid and a following chest infection. After waiting another few weeks to see if it reduced, I was referred to ENT. The speed of the initial appointment with ENT surprised me because it was so quick. Several possibilities of the cause of the swelling were discussed and I was sent for an ultrasound scan. I was seen very quickly, and I was told that I had a mass near my carotid artery and would need further investigation,

I was referred to another department -Vascular Surgery because of the position of the mass close to the carotid artery. I was told that it was a Paraganglioma, and it was decided that a surgical procedure to remove it would be the better option for treatment. My consultant described in great detail all of the possible side effects and all of the contingency plans that he would have in place in case of any difficulties during the surgery or in the initial recovery period.

I was surprised when I was told the results of the histology tests, that it was not carotid body tissue but was thyroid tissue. I was relieved that it had been removed. From first finding the lump to having the lump removed was less than 10 months, which really impressed me especially with the constraints on the NHs at the moment. I am grateful to my consultant and his team for the care that I have received during this period.

Figures:

Figure 1: Ultrasound scan of the neck (right) showing a mass at the level of the carotid bifurcation.

Figure 2: CT neck image (axial view) showing a mass at the level of the right carotid bifurcation.

Figure 3: CT neck image (sagittal view) showing a mass at the level of the right carotid bifurcation.



Figure 1: Ultrasound scan of the neck (right) showing a mass at the level of the carotid bifurcation.

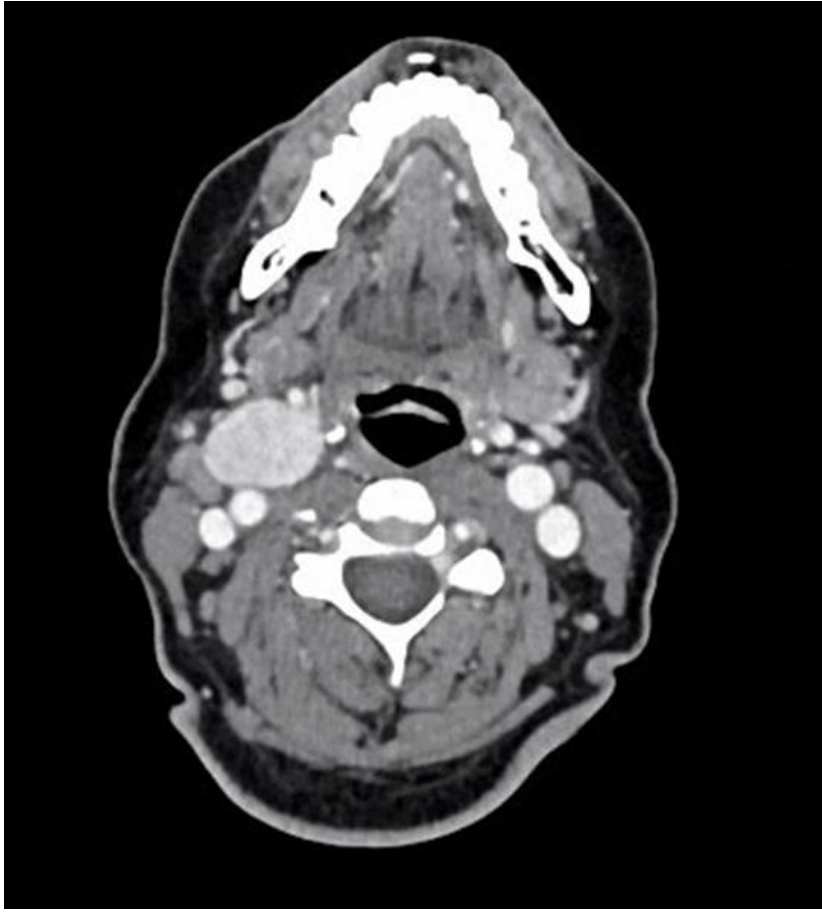


Figure 2: CT neck image (axial view) showing a mass at the level of the right carotid bifurcation.

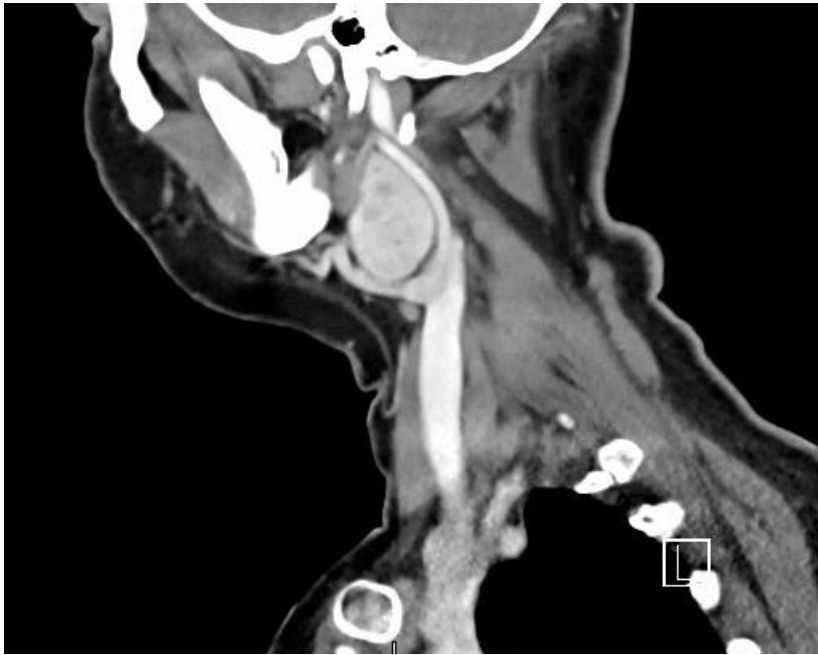


Figure 3: CT neck image (sagittal view) showing a mass at the level of the right carotid bifurcation.