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CASE REPORT

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# Papillary adenocarcinoma in submandibular region

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**ABSTRACT**

Lateral ectopic thyroid is a rare event and even more uncommon is a primary malignancy in lateral ectopic thyroid. We present a case of papillary adenocarcinoma in lateral ectopic thyroid in submandibular space in a 55-year old male. To our knowledge this is the third case documented in the world and the first one in Europe. Lateral ectopic thyroid in this region is easily masqueraded as a submandibular gland swelling and so was our patients' tumor preliminarily diagnosed as a submandibular gland tumor. Furthermore, in the preoperative computed tomography (CT) scan the tissue was misinterpreted being adjacent to the submandibular gland. The diagnosis was revealed during the surgery and confirmed by the histology. This report demonstrates the difficulty in the differential diagnosis of neck masses. Although rare, ectopic tissue should be remembered as a possible diagnosis of all neck masses and the relevant preoperative examination should be performed by skilled professionals.

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Papillary adenocarcinoma; ectopic thyroid tissue; submandibular mass

## Introduction

Ectopic thyroid tissue (ETT) is defined as the presence of thyroid tissue outside its normal pretracheal location. The clinical diagnosis is rare: 1 per 100,000–300,000 in overall population, and up to 1 per 4000–8000 in patients diagnosed with thyroid disease.[1,2] The actual incidence of ectopy, however, is thought to be greater. In an autopsy study of 200 asymptomatic patients in North America, ETT in the lingual region was found in 10% of the population.[1] Aberrant thyroid tissue is most commonly manifested in females.[2] It may be diagnosed at any age.[2,3] The location is lingual in 90% of the diagnosed cases, but also other locations in various sites in the neck and all the way to the areas below the diaphragm have been described.[1,4] It is generally thought to be an embryogenic developmental abnormality, but hypotheses of surgery or trauma induced ectopy has been presented.[3,5] ETT may coexist with an orthotopic thyroid gland or it can be the sole recognizable thyroid tissue.

Any disease known to affect orthotopic thyroid can also be found in ETT.[5] The tissue carries a low risk of malignancy and is most often diagnosed with inflammation or hyperplasia.[6] Of malignancies papillary carcinoma is the most common type.[2,6] Cases of follicular, mixed follicular and papillary, Hürthle cell and medullary carcinomas have also been reported.[1,2,6]

We present a rare case of papillary carcinoma in lateral ectopic thyroid gland masquerading as a submandibular gland tumor. To our knowledge this is the third documented case with the presented diagnosis in this region.

## Case presentation

A 55-year-old male presented with a two-month history of a painless neck mass on the left. He had no other symptoms related to eating, swallowing or speaking. He had history of COPD, asthma and pulmonary emphysema. The patient had a long history of smoking but had given up smoking one year earlier.

Clinical examination identified a firm, mobile, nontender mass in the region of the left submandibular gland. The neck ultrasound examination revealed a $5 \times 3$ cm heterogenic tumor with microcalcifications

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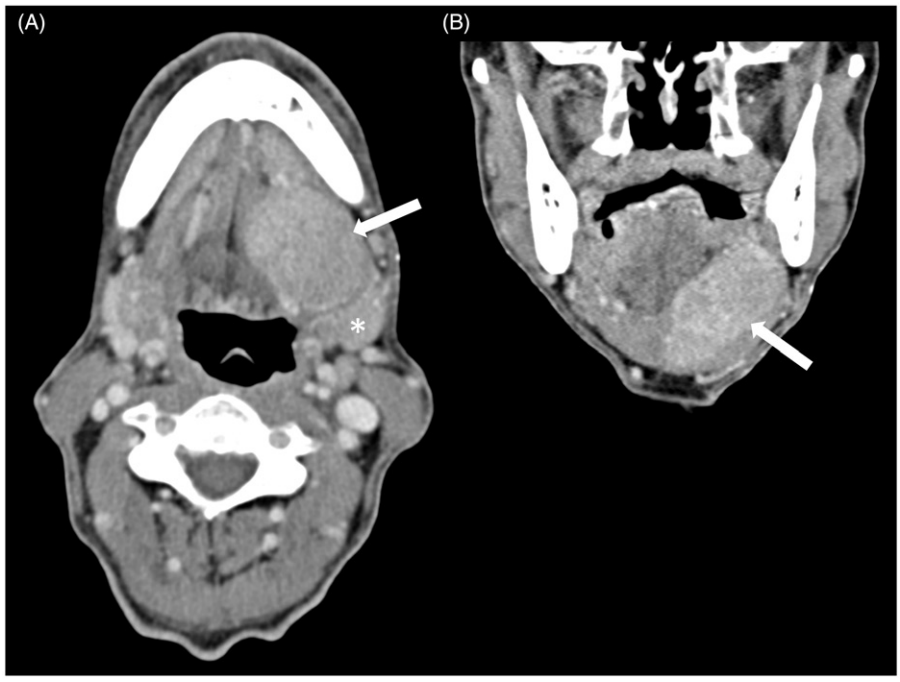


**Figure 1.** Transaxial (A) and coronal (B) contrast-enhanced computed tomography (CT) shows a large enhancing tumor (arrows) in the left sublingual space, close to the adjacent left submandibular gland (asterisk).

and abundant vascularity. Floor of mouth, lymph nodes and thyroid gland were normal. Fiberoptic endoscopy was also performed and was unremarkable.

With the preliminary diagnosis of a submandibular gland tumor the patient was addressed to university hospital for further treatment. A contrast enhanced computed tomography (CT) scan demonstrated a 4 cm tumor which was unfortunately misinterpreted by a radiology resident as a submandibular space lesion associated with the submandibular gland (Figure 1). The tumor was revealed to locate in the sublingual space, separate from the submandibular gland only in the retrospective analyses. Fine-needle aspiration biopsy (FNAB) indicated class IV cytology (Papanicolaou classification – cytology strongly suggestive of malignancy). The first tissue staining's suggested adenocystic carcinoma although basalioma and adenoma were also thought to be possible. The submandibular gland was regarded as the most probable source. However, malignancy of the thyroid could not be completely excluded in the FNAB sample.

The patient was admitted to surgery. Considering both the highly suspected malignancy of the tumor and the deteriorated lung functionality a neck dissection level I–III was planned in the same session even without an exact diagnosis. The surgery revealed that the tumor was located in the floor of the mouth, superior to the mylohyoid muscle, separate from the adjacent submandibular gland. The encapsulated tumor with a diameter of 7 cm was removed, and neck dissection level I–III was performed on the same side.

The final staining of the FNAB was completed three days postoperatively and showed papillary thyroid carcinoma. The diagnosis was confirmed by the immunohistochemical analysis of the tissue. The positive TTF-1 and thyroglobulin along with negative S-100 and Ki-67 suggested thyroid origin: either a metastasis in a lymph node or primary carcinoma in an ectopic thyroid gland. According to the analyses after operation no malignancy was found in the left submandibular gland. Considering the location of the tumor in the floor of the mouth and the intraoperative findings, metastasis was not considered likely and the diagnosis was interpreted as primary malignancy in an ectopic thyroid gland. Small metastasis was found in one of the dissected lymph nodes. The orthotopic thyroid gland appeared normal on both CT and ultrasound.

The patient was admitted to a district hospital for total thyroidectomy and further treatment. Preoperatively, there was no evidence of thyroid dysfunction. No additional aberrant thyroid tissue was found during the surgery. The removed orthotopic thyroid gland was revealed healthy in the histological examination. The patient recovered uneventfully after the operation and did not have problems with breathing, eating or drinking. The treatment continued with radio ablation/iodine therapy followed by thyroxin replacement therapy.

Sixteen months postoperatively the patient was referred back to the university hospital with a probable pathological collum fracture in the left femur bone. A CT guided biopsy was done and the cytology showed metastasis of papillary thyroid carcinoma. No other metastasis was detected in the full-body CT performed. The patient agreed to treatment with an exarticulation from which he recovered uneventfully. Six months after the exarticulation the patient died from lung problems not related to the papillary carcinoma.

## Discussion

The first ectopic thyroid gland was described in 1869 by Hickman: a lingual thyroid obstructing airways and causing death by suffocation in a one-day-old girl.[7] The existence of lateral thyroid tissue, however, remained controversial until recent decades. Known as lateral aberrant thyroid all thyroid tissue found outside the thyroid gland was thought to represent metastasis from thyroid carcinoma.[1,2] Patients presenting benign lateral ETT or a malignant lateral aberrant thyroid coexisting with a benign orthotopic thyroid goiter showed that also a lateral ectopic gland could exist and even carry a primary carcinoma.[8] The first ETT in the submandibular region was described by Helidonis in 1980.[9] It is, on the other hand, largely thought that in some cases of a metastasis of highly differentiated thyroid carcinoma can completely replace the lymph node tissue. The majority of preceding cases of lateral aberrant thyroid are thought to represent a metastasis like this.[10]

The thyroid gland develops during the third to fourth week of gestation. On the seventh week of fetal life, the gland anlage (precursor) descends to its final location anterior to the pretrachea and larynx. During the migration, the gland is attached to the foramen caecum by a narrow tube, the thyroglossal duct. This duct usually atrophies after thyroid formation.[1,2,6]

There are several hypotheses on the origin of ETT. At the embryogenesis, a defect in the descent of the gland may lead to ETT situated along the descending route.[2] The other hypothesized mechanism, concerning lateral ETT, involves two lateral anlagen, one for each lobe, that are thought to be engaged in the morphogenesis.[1] The arrest of the migration of one of the lateral thyroid anlagen could also explain the possibility of non-midline ETT in the neck.[11] When not able to fuse with the median thyroid anlage the anlagen would be presented as ectopic thyroid mass with a co-existing orthotopic thyroid. Also spread of tissue of an orthotopic gland during surgery or as a result of a trauma has been hypothesized as a possible origin for this ectopy.[3,5]

ETT can be found at various sites. About 90% of the reported cases are lingual, located at the base of the tongue,[1] but ectopy can be found all the way along the migration path of the thyroid [12] as well as laterally in the neck, even more seldom in the mediastinum [4,13–15] or in the areas below the diaphragm.[1]

As the autopsy rates reveal, the majority of patients has little or no symptoms of thyroid ectopy, and never become aware of this condition. Patients with submandibular thyroid usually present with a palpable, mobile, painless mass. In most cases of a submandibular thyroid an orthotopic thyroid gland does exist.[5,10] These patients are usually euthyroic. In only one reported case the patient was presented with hyperthyreosis.[16] Cases where the lateral ectopic thyroid is the only functioning tissue have also been documented.[11,17–19]

The differential diagnosis between a primary malign ectopy and a metastasis from the thyroid gland can be difficult. When establishing the correct diagnosis a good histological sample and therefore neck surgery is needed. ETT being confused with lymph node metastasis the patient may undergo an unnecessary radical neck dissection. Differential diagnoses of submandibular space masses include salivary gland tumors, inflammatory and malignant lymphadenopathies, lipomas, lymphatic malformations, epidermal cysts, branchial cleft cyst, sebaceous cysts and dermoid cysts.[6] If a patient presents with an asymptomatic swelling in the neck for more than four weeks, a good clinical procedure is to perform an ultrasound guided FNAB by a skilled professional. The neck should also be examined for a normally located thyroid gland. ETT is most often benign and may even present as the only functioning thyroid tissue. Removal of a benign ETT may thereby cause unwanted hypothyroidism.

Our case is a rarity amidst rarities. The literature review revealed two preceding cases with the same diagnosis: papillary adenocarcinoma in a lateral ectopic thyroid in the submandibular region.[18,19] Both presented with a coexisting orthotopic gland: one with a nodule of papillary thyroid carcinoma without capsular invasion [19] (as per the pathologic diagnosis), as the other's diagnosis remained unclear.[18] The present case is the third documented case and the first one in Europe.

Our patient presented with no thyroid-related symptoms and was euthyroid with a normal orthotopic thyroid gland. Although the ectopic thyroid was

situated in the floor of the mouth, both ultrasound and CT scans as well as clinical examination appeared to suggest a submandibular location. No remnants of the thyroglossal duct were revealed by imaging or by surgery. When the FNAB suggested malignant tissue a salivary gland origin was primarily suspected since this location is rare for thyroid tissue even though there was a suspicion of thyroid origin in the pathology report. As there is an aim to minimize the waiting time of the cancer patients, the operation was performed before the final immunohistochemical analyses of the FNAB which can be discussed if this was unfavorable in this case. If the diagnoses would have been known, the neck dissection could perhaps be avoided, though in the final report there was a small metastasis in one of the evacuated lymph nodes in the neck.

## Conclusions

ETT should be remembered as a differential diagnosis for neck masses. Most commonly found at lingual space it is possible to be found at various locations. It is of remarkable clinical importance when diagnosed with malign disease. As the treatment of choice is surgical it can lead to unnecessary radical neck dissection if considered as an unknown primary to the FNAB.

## Disclosure statement

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of this article.

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