

# Pyramidal lobe-dominant papillary thyroid carcinoma - a rare entity with important clinical implications.

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## Abstract

Papillary thyroid carcinoma of the pyramidal lobe is rare. We describe a case of a 77-year-old woman who corroborates current literature findings, namely greater presence of adverse prognostic factors. Upper Neck Papillary Thyroid Cancer is a new entity, proposed to group these tumors with potential clinical and therapeutic implications.

## TITLE PAGE

*Pyramidal lobe-dominant papillary thyroid carcinoma – a rare entity with important clinical implications.*

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Brief running title: Pyramidal lobe-dominant papillary thyroid carcinoma

## Ethical Statement

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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**Abbreviations:** AJCC, American joint committee on cancer; BMI, body mass index; CT, computerized tomography; FNAC, fine needle aspiration cytology; MDT, multidisciplinary head and neck oncology team; PTC, Papillary thyroid carcinoma; T4, thyroxine; TGDC, thyroglossal duct cysts; TSH, thyroid stimulating hormone; UPTC, Upper Neck Papillary Thyroid Cancer.

## ABSTRACT

### Background

Papillary thyroid carcinoma (PTC) primarily located in the pyramidal lobe of the thyroid gland is extremely rare, so the clinical and pathological features are not well understood.

### Case

The authors describe a case of multifocal PTC with larger pyramidal lobe tumour, in a 77-year-old female patient who underwent en bloc total thyroidectomy with pyramidal lobe, hyoid bone and cervical lymph node excision.

### Conclusion

In line with the present case, current literature reports a greater presence of worse prognostic factors, namely extrathyroidal extension, advanced T stage or presence of cervical lymph node metastasis. Recently, a new classification has been suggested – Upper Neck Papillary Thyroid Cancer (UPTC) – which encompasses these carcinomas, Delphi ganglion metastases and thyroglossal duct cyst carcinomas, with potential clinical and therapeutic implications, particularly the need for orthotopic thyroidectomy. Also, the complete excision of the pyramidal lobe during thyroidectomy may influence the success of radioactive iodine therapy and the patient’s follow-up surveillance.

Keywords: Thyroid, Thyroid cancer, Pyramidal lobe of thyroid, *Lalouette* pyramid, Papillary thyroid carcinoma.

## INTRODUCTION

The pyramidal lobe of the thyroid gland is present in 15-75% of the population. [1] On the other hand, papillary thyroid carcinoma (PTC) of the pyramidal lobe, defined as a single pyramidal lobe cancer or multifocal cancer in which pyramidal lobe tumor is the largest, is extremely rare. [2,3] Because of this, the clinical and pathological characteristics of these PTCs are not well understood. [3]

The authors describe a case of PTC of the pyramidal lobe and its clinical implications according to a revision of the current scientific literature.

## CASE PRESENTATION

A 77-year-old female patient was referred to the Head and Neck Surgery department of the Portuguese Institute of Oncology of Lisbon, because of a midline neck mass, present at least for ten years, but with progressive growth in the previous nine months [Figure 1]. The remaining head and neck and general physical examination was unremarkable. She had a personal history of obesity (BMI 38.16 Kg/m<sup>2</sup>), asthma, obstructive sleep apnea, hypertension, dyslipidemia and depressive disorder, and a family history of unspecified breast cancer (sister). She denied smoking, high alcohol intake or other family history of cancer or endocrine disease.

The patient was accompanied with a neck CT scan and ultrasound that had identified two thyroid nodules – the largest one with 25mm located in the pyramidal lobe [Figure 2] and another with 21mm located in the right lobe [Figure 3], both were TI-RADS 5 and suggestive of papillary thyroid cancer (PTC) after fine needle aspiration cytology (FNAC). General and endocrine blood tests, including TSH, T4 hormone and calcitonin, were within normal values range.

In our hospital, the review of the cytology samples confirmed the diagnosis and after the decision of the treatment by the multidisciplinary team (MDT), the patient underwent a total thyroidectomy *en bloc* with

the pyramidal lobe and hyoid bone. [Figure 4,5,6] In addition, intraoperatively, the presence of suspicious lymph nodes in the central compartment of the neck led to a lymph node dissection at this level. It was not possible to preserve the right recurrent laryngeal nerve because of direct invasion of the cancer. Finally, due to the close proximity of the main malignant nodule to the trachea, tracheal shaving was also performed.

The immediate postoperative period was complicated by dyspnea and the need for tracheostomy, which the patient maintained at the time of hospital discharge. The laryngoscopy, performed during hospitalization, identified bilateral paralysis of the vocal cords. There were no other inward complications, including hypocalcemia.

The histologic result reported a multifocal papillary thyroid cancer, with the largest node at the pyramidal lobe (maximal dimension of 25mm), with extrathyroidal extension and lymphovascular invasion, as well as metastasis in two central compartment lymph nodes. [Figure 7,8] The TNM staging was pT3bpN1a. This time, the MDT decided for adjunctive treatment with radioiodine therapy.

Currently, at six months of follow-up, the patient was finally able to remove the tracheostomy after verified mobility of the left vocal cord and permeable glottis on laryngoscopy. No other incidents were reported during follow-up. Also, body scintigraphy and neck CT scan confirmed absence of local or regional disease.

## DISCUSSION

The widespread availability of imaging modalities, such as high resolution ultrasound and ultrasound guided FNAC have contributed to the higher detection of thyroid carcinoma. [4] The most frequent histologic type of thyroid carcinoma is papillary carcinoma, accounting for approx. 80% of differentiated thyroid carcinomas. [5] Although PTCs are generally associated with an indolent clinical course and favorable prognosis – 10 year relative survival rate of 93% – recurrence is significant at 5-20%. [6] This is specially associated with advanced age, male gender, tumour size, multifocality, extrathyroidal extension, extranodal spread and lymph node ratio. [7] The pyramidal lobe of the thyroid gland, usually connected to its isthmus, is located in the inferior portion of the thyroglossal duct, a remnant of the embryological development of the thyroid. [1] It is reported to be present in 15-75% of the population. [1] On the other hand, papillary thyroid carcinoma (PTC) of the pyramidal lobe is extremely rare, thus, the clinical and pathological characteristics of these PTCs are not well understood. [1,2] In comparison with papillary thyroid carcinoma of other locations, pyramidal lobe PTC was found to have a more advanced patient age at diagnosis (mean of 58 vs 43 years old), smaller nodule dimension, but more frequent extrathyroidal extension, lymphatic invasion, advanced AJCC staging and multifocal location. [3] These associations are supported in the present case: our patient exhibited all but one (smaller nodule dimension) of the previous reported adverse features, which correlated with a more aggressive disease, specifically the close relation of the cancer lesion with the trachea and direct invasion of right recurrent laryngeal nerve during thyroidectomy and long period of tracheostomy dependency. Even though, these features also put the patient at increased risk of recurrence [7], there were no signs or symptoms of recurrence at the latest follow-up of 6 months after surgery. Zizic M *et al.* identified a high diagnostic heterogeneity in the scientific literature concerning not only pyramidal lobe papillary carcinoma, but also Delphian node metastasis and thyroglossal duct cysts (TGDC) carcinoma, that may explain the controversy in managing these patients. In order to address the issue, a new terminology to group these entities was created – Upper neck papillary thyroid cancer (UPTC). [Table 1] [8] The clinical distinction between these entities has important implications for the patient's treatment, namely decision for orthotopic thyroid resection, which is generally recommended in the presence of PTC in the pyramidal lobe, metastasis to the Delphian node and indetermined origin, but not in TGDC papillary carcinoma. [8] The reported presence of residual thyroid tissue in patients submitted to radioiodine treatment after total thyroidectomy is approx. 50%, and the rate of occult PTCs in the same location is around 4%. [3,9] These reports highlight the oncological importance of the complete resection of the pyramidal lobe during thyroidectomy in papillary thyroid carcinoma, namely the possibility to improve radioiodine treatment efficacy or facilitating postoperative follow-up surveillance with scintigraphy or blood thyroxine values. [9,10] The characteristic multifocality of PTC and the activation of the residual pyramidal lobe when it is not removed during thyroidectomy, also make the pyramidal lobe a potential place for recurrent PTC.

[7] In conclusion, papillary carcinoma of the pyramidal lobe is a rare diagnosis, but associated with adverse prognostic features. A new classification of Upper Neck Papillary Carcinoma (UPTC) further distinguishes papillary pyramidal lobe carcinomas from PTCs of other thyroid locations. This pathological entity was proposed to facilitate the analysis of the literature, the diagnostic investigation and the decision for the treatment. Complete excision of the pyramidal lobe during total thyroidectomy for papillary carcinoma may have important implications for the effectiveness of adjuvant radioiodine therapy as well as for patient's follow-up surveillance and risk of recurrence. Future studies are needed to better understand the usefulness of the reported new terminology, as well as the clinical behavior and implications of the pyramidal lobe in the surgical and adjuvant treatment of these rare thyroid carcinomas.

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## Data Availability Statement

The authors declare that all data supporting the findings of this study are available within the article and its supplementary information files.

## Conflict of Interest

The authors declare to have no conflicts of interest in connection with this scientific work.

## LEGENDS

[Figure 1] Pre-operative frontal view of midline neck mass.

[Figure 2] Neck CT scan with large midline neck mass, anterior to thyroid cartilage.

[Figure 3] Neck CT scan, with a thyroid nodule at the right thyroid lobe.

[Figure 4] Surgical procedure: Right lobe pulled to the medial side, so that the right common carotid artery can be seen).

**[Figure 5]** Surgical procedure: Right lateral view of the pyramidal lobe nodule between the hyoid bone and the thyroid gland.

**[Figure 6]** Surgical procedure: Final operative specimen with the largest nodule in the pyramidal lobe in the upper center of image and the rest of the thyroid gland centered below with the reference of the right lobe nodule with long silk stitch.

**[Figure 7]** Histological slice at the level of the nodular lesion in the pyramidal lobe showing a classic pattern papillary thyroid carcinoma.

**[Figure 8]** Higher magnification of histological slice (figure 7), with more evident extrathyroidal extension, adjacent muscle invasion and lymphovascular invasion of PTC.

**[Table 1] Upper neck papillary thyroid cancer (UPTC)** clinical entity with key histologic features for final pathologic diagnosis. (adapted from Zizic M *et al* [8]).

**TABLES**

FINAL PATHOLOGIC DIAGNOSIS	KEY HISTOLOGIC FEATURES
<b>Thyroglossal duct cyst (TGDC) cancer</b>	Presence of benign respiratory, cuboidal or squamous epithelial cyst lining in association with PTC and Lack of features of lymph node architecture and Lack of evidence of a primary PTC within thyroid parenchyma
<b>Delphian node metastasis</b>	Histologic features of PTC with lymph node architecture, including lymphoid stroma and subcapsular sinus
<b>Pyramidal lobe primary</b>	PTC surrounded by a background of benign thyroid parenchyma Lack of features of lymph node architecture or TGDC features
<b>Indeterminate origin</b>	Exact diagnosis based on above criteria could not be achieved.

**Table 1 – Upper neck papillary thyroid cancer (UPTC)**clinical entity with key histologic features for final pathologic diagnosis. (adapted from Zizic M *et al* [8]).











