



## Thyroid Paraganglioma, An Uncommon tumor at a Rare Site- A Case Report

Authors

Milap Shah, Palak Jain, Harika Puligolla, Hemanth Vudayraju

### Abstract

*Thyroid paragangliomas are exceedingly rare neoplasms with only few cases reported worldwide. They have a very similar radiology, cytology and histology to other thyroid neoplasms however treatment and prognosis differ significantly due to which accurate diagnosis becomes a sine qua non for their appropriate management. Here we present a case of a 65 year old female who complained of thyroid swelling which on Fine needle aspiration cytology was diagnosed as multinodular goitre under Bethesda category 2. However total thyroidectomy was done and histomorphology and immunohistochemistry confirmed the diagnosis of thyroid paraganglioma.*

**Keywords:** *Thyroid tumor, paraganglioma, thyroid paraganglioma.*

### Introduction

Extra-adrenal Paraganglia both sympathetic and parasympathetic are derived from the primitive neural crest cells of the autonomic nervous system. Parasympathetic paragangliomas are almost exclusively present in the head and neck region along the cranial nerves, the arterial vasculature or rarely in orbit and paranasal sinuses.<sup>[1]</sup> Carotid body tumor is the most common head and neck paraganglioma, followed by glomus jugulare, glomus tympanicum, and the vagal paraganglioma.<sup>[2]</sup> Among the rare anatomic sites of extra-adrenal paraganglioma, thyroid is possibly one of the rarest site constituting <0.1% of all thyroid neoplasms.<sup>[3]</sup>

As paragangliomas are rare in thyroid and resemble closely with primary thyroid neoplasms which are more common, their diagnosis becomes both necessary and challenging. Thyroid paragangliomas may clinically, radiologically,

cytologically and to some extent histologically simulate other thyroid neoplasms but the management differs significantly therefore accurate diagnosis on histology and immunohistochemistry are wand for appropriate patient management.

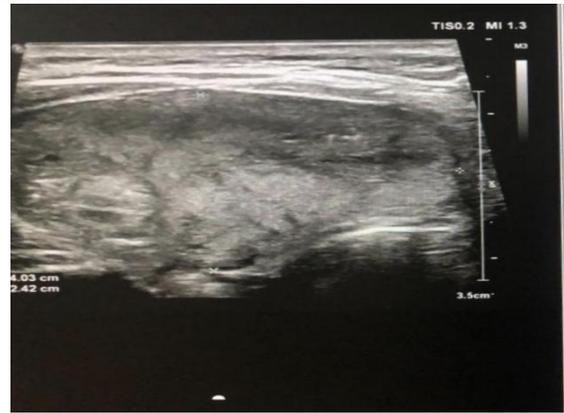
In this paper we describe a case of a 65 year old female who presented with thyroid swelling and euthyroid status with emphasis on the diagnostic challenges.

### Case Report

A 65 year old female presented with a thyroid swelling which was since last two years and was gradually increasing in size. Clinical examination showed a swelling of approximately 6 x 5cm predominantly towards right which moved on deglutition with no palpable cervical lymphadenopathy. Biochemical and serological investigations revealed euthyroid status of patient.

Her family history was negative for any prior thyroid or endocrine disorders. Doppler Ultrasound of neck revealed an enlarged right thyroid lobe with a well delineated, solitary hypoechoic nodule measuring 6.5 x 3.5x3cm with increased intra- and peri-nodular vascularity. Left lobe and isthmus showed no significant finding. FNA was done and showed atypical cells with moderate to abundant pale cytoplasm, large nuclei having speckled chromatin and inconspicuous to prominent nucleoli. The FNA was reported as suspicious of malignancy, Bethesda category V. Total thyroidectomy was done for definitive diagnosis and management. Total thyroidectomy specimen weighed 150g, right lobe measured 9x4x3cm, Isthmus measured 2x1cm and left lobe measured 5x2x1cm. The external surface was encapsulated and showed no capsular breach. Cut section of right lobe showed solid, cystic and haemorrhagic areas while cut section of left lobe and isthmus was homogenous brown.

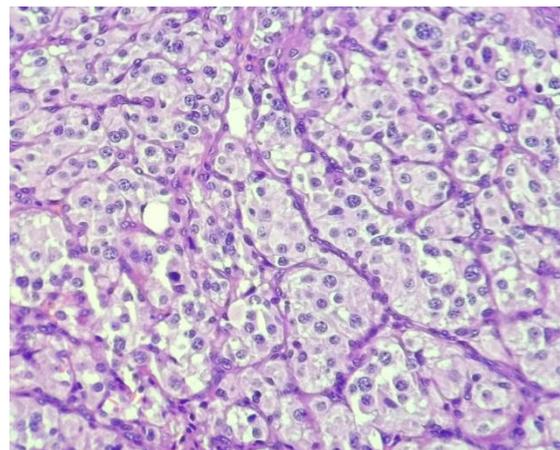
The histopathological examination showed a well encapsulated neoplasm comprising of nests and trabeculae of round to polygonal cells surrounded by slender vascular septae; thus, giving a Zellballen pattern. The tumor cells had round to ovoid nuclei with fine granular chromatin and small nucleoli in some with moderate amount of granular eosinophilic cytoplasm. There were prominent, thin walled vascular spaces and areas of hemorrhage within the neoplasm (fig. 2). Amyloid like material or thyroid follicles were not seen in the right lobe. Immunohistochemistry was performed and the neoplastic cells were diffusely and strongly positive for synaptophysin and chromogranin, S100 highlighted sustentacular cells while PAX8, CEA and CK were negative (fig 3,4,5). Histomorphology and immunohistochemistry were consistent with the diagnosis of Intrathyroid paraganglioma. The patient is under close follow up since last 3 months.



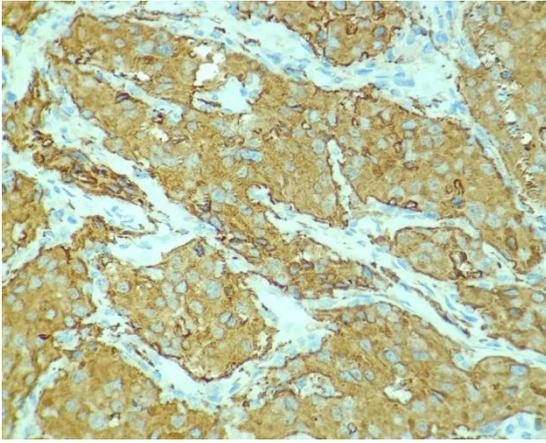
**Figure 1a-** Doppler Ultrasound showing enlarged right thyroid lobe with a well delineated, solitary hypoechoic nodule measuring 6.5 x 3.5x3cm with increased intra- and peri-nodular vascularity.



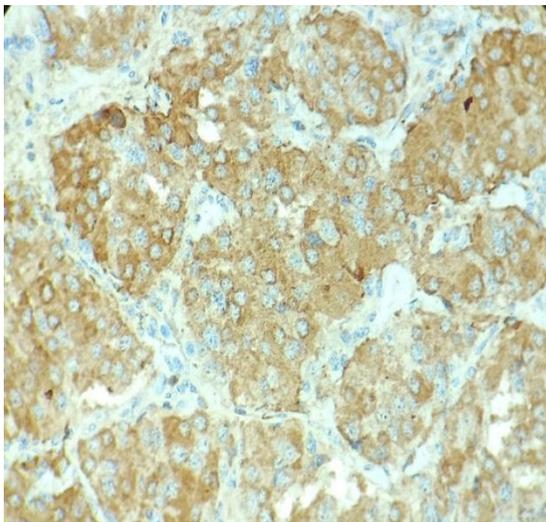
**Figure 1b-** Gross examination of cut surface of right thyroid lobe showing solid, cystic and haemorrhagic areas



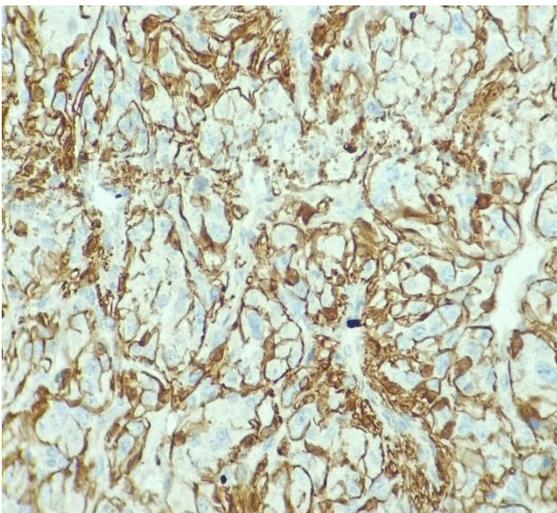
**Figure 2** Hematoxylin and Eosin section showing tumor arranged in Zellballen pattern comprised of cells having abundant eosinophilic cytoplasm, round nuclei with vesicular chromatin



**Figure 3** Immunohistochemical stain revealed Positivity of synaptophysin.



**Figure 4.** Immunohistochemical stain showing chromogranin positive tumor cells



**Figure 5.** Immunohistochemical stain for S100 highlights sustentacular cells

### Discussion

Thyroid paragangliomas are exceptionally uncommon and show a strong female preponderance typically appearing between 40- 60 years of age. They present as asymptomatic, solitary thyroid nodules. The preoperative diagnosis is difficult since the cytologic and histologic features may overlap with other more common primary thyroid neoplasms such as medullary thyroid carcinoma, thyroid neuroendocrine tumor and thyroid trabecular adenoma.<sup>[4]</sup> Preoperative U/S may reveal the presence of a hypervascular thyroid nodule. Surgical removal may be hazardous due to the increased vascularity and friability of the tumor which may also be densely adherent to the surrounding tissues. Histologic diagnosis of a thyroid paraganglioma is important since it denotes a mostly benign behavior with no reported distant metastases, whereas thyroid medullary carcinoma is clearly a malignant tumor with considerable metastatic potential.<sup>[4]</sup> Surgical removal of thyroid paragangliomas is the mainstay of treatment. Its identification and differential diagnosis from other thyroid tumors is extremely important in terms of subsequent management and prognosis.<sup>[5]</sup>

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