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# CASE REPORT Unusual Presentation of Occult Medullary Carcinoma of Thyroid Gland: Case Report

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## ABSTRACT

**Introduction:** Medullary thyroid carcinoma (MTC) is a rare tumour with varied presentation. They commonly manifest as neck swellings, however, may occasionally present as change in voice or an elevated calcitonin level, in an otherwise occult medullary carcinoma.

**Case Report:** We present a case of medullary carcinoma of thyroid gland with an unusual presentation of hoarseness of voice, without an apparent neck swelling.

**Conclusion:** The presentation of medullary carcinoma of thyroid gland, in a 55 year old male, with sole complaints of hoarseness of voice, highlights the need for a high index of suspicion in such cases.

**Keywords:** Adult male, hoarseness, vocal cord palsy, recurrent laryngeal nerve, thyroid.

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## INTRODUCTION

Medullary carcinoma of thyroid gland is more common in females and presents at a mean age of 50 years.<sup>1</sup> Sporadic tumours are usually single and present as midline neck swellings. It is an uncommon tumour accounting for five to eight percent of all thyroid cancers.<sup>2</sup> It may occur sporadically or against a background of an inherited autosomal dominant trait related to a germ line mutation of the RET protooncogene, which cause MEN 2A, MEN 2B or familial

## MTC (FMTC).

### **CASE REPORT**

A 55 year old male, came to our outpatient department with the sole complaint of hoarseness of voice since one year. A detailed history failed to identify any specific cause for hoarseness. On indirect laryngoscopy and 70 degree rigid laryngoscopy, a left vocal cord palsy was noted. On examination of the neck there was no palpable swelling. CT neck (from skull base to upper thorax) revealed a heterogeneously enhancing nodule in lower pole of left thyroid gland extending inferiorly into the tracheo-oesophageal groove with suspicious compression of the left recurrent laryngeal nerve causing left vocal cord paralysis. Serum calcitonin level was then checked, which was raised and vanillyl mandelic acid (VMA) level was normal. Thyroid function tests, showed high TSH and normal T3 and T4 levels. An ultrasound examination of neck showed a well-defined heterogeneously hypo-echoic lesion with multiple small calcifications within, arising from the upper pole of left lobe of thyroid gland. Another similar exophytic lesion was noted arising from the lower pole measuring 1.5 x 1.2 cm, inferiorly extending to tracheo-esophageal groove. Findings were suggestive of a neoplastic aetiology. A metastatic lymph node was noted in left cervical region measuring 1.5 x 0.8 cm, medially abutting the proximal left common carotid artery and distal third of internal jugular vein. Laterally, it was abutting the middle third of left sternocleidomastoid muscle. A PET CT of whole body revealed two hypo dense nodules in left lobe of thyroid, a metastatic left level IV lymph node and no active disease elsewhere in the body. Fine needle aspiration cytology from the left thyroid lower pole and adjacent left cervical lymph node was suggestive of medullary carcinoma of thyroid with metastasis to left cervical lymph node. A total thyroidectomy was planned with ipsilateral central compartment clearance and contralateral anterior compartment clearance. Histopathology report confirmed the diagnosis and 2/13 lymph nodes were involved by the tumour. Post-operative period was uneventful. Patient is followed up regularly. At three months post operative period a repeat CECT neck and thorax is





Figure-1: CT Scan Neck showing the vocal cords.



Figure-2: PET Scan showing the left thyroid lobe.

advised and radiotherapy will then be planned.

## DISCUSSION

Hazard et al.3 segregated medullary carcinoma of thyroid as a distinct entity in 1959. Medullary thyroid carcinoma is a malignant tumour displaying para follicular C-cell differentiation,<sup>4</sup> which secretes calcitonin and neuropeptides. The C cells originate from the embryonic neural crest; as a result, medullary carcinomas often have the clinical and histologic features of other neuroendocrine tumours such as carcinoid and islet cell tumours. Calcitonin is the most specific marker for para follicular cells. Distant metastasis in medullary thyroid carcinoma are frequently present at initial presentation of sporadic MTC patients.<sup>5,6</sup> Organs most commonly include the lungs, bones and liver, and infrequently the brain. Distant metastasis are frequently associated with persistent loco regional disease.<sup>7</sup> The discovery of metastasis is often preceded by detectable or increasing serum levels of calcitonin or carcino embryonic antigen (CEA). Calcitonin and CEA are rather considered risk factors for the presence and development of metastasis and a poor prognosis.<sup>7,8</sup> Patients who have elevated and not fluctuating tumour markers without demonstrable tumour may have a good prognosis and quality of life.9-11 Those, patients are being followed up with serial tumours marker and regular imaging, interval depending on the serum marker level and doubling time.<sup>5,7,8</sup> Approximately 50 percent of patients have clinically detectable cervical lymph node involvement, up to 15 percent have symptoms of upper aero digestive tract compression or invasion such as dysphagia or hoarseness and about 5 percent have distant metastatic disease.<sup>2</sup> Patient was under treatment and evaluation for hoarseness of voice for a year before he presented in our outpatient department and a CT scan neck was advised to determine the cause. As medullary carcinoma is an aggressive tumor, the patient underwent a PET scan to rule out distant metastasis. The calcitonin levels helped us determine the development of metastasis and support the diagnosis. The normal levels of VMA ruled out MEN 2A and MEN 2B, revealing the sporadic origin of the tumour. As literature suggests, in a clinically apparent medullary thyroid cancer of tumour less than 1 cm and no suspicious lymph nodes on untrasound of neck, a total thyroidectomy with bilateral central neck dissection is done. For a tumor more than 1 cm in size with suspicious lateral neck lymph nodes, a total thyroidectomy with bilateral central neck dissection and ipsilateral lateral neck dissection is advised. Contralateral neck dissection is considered for positive contralateral lymph nodes or bilateral tumors of more than 1 cm.<sup>12</sup> Treatment goals in advanced MTC are ultimately related to promoting the quality of life. It is important to explicitly discuss with the patient the expectations and goals with respect to quality of life in the individual context and to agree on a personalized treatment plan.

## CONCLUSION

Medullary carcinoma of thyroid gland is a rare and aggressive tumour. The involvement of recurrent laryngeal nerve causing hoarseness of voice, as presented by this patient, highlights the importance of evaluating the thyroid gland in all patients presenting with vocal cord palsy.

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