

# HUGE MEDULLARY THYROID CARCINOMA EXTENDING FROM NECK TO MID CHEST

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## ABSTRACT

*Medullary thyroid carcinoma (MTC) is a distinct thyroid carcinoma that originates in the calcitonin producing parafollicular C cells of the thyroid gland. We present a case of an elderly male who presented with breathing difficulty and large right antero-lateral neck swelling. Fine needle aspiration (FNA) revealed a medullary thyroid carcinoma and he was managed by total thyroidectomy with central neck dissection and removal of mediastinal extension as well. Reported case is a rare as to its extension from right upper neck to mid chest.*

*Key words* Medullary thyroid carcinoma, Calcitonin, Parafollicular cells, Multiple endocrine neoplasia.

## INTRODUCTION:

Medullary thyroid cancer (MTC) is a form of thyroid carcinoma that originates from the parafollicular cells (C cells), which produces the hormone calcitonin. Approximately 20% of medullary thyroid cancers are due to underlying genetic abnormality, which is caused by a mutation in the RET proto-oncogene.<sup>1</sup> This form is classified as familial MTC. When MTC occurs by itself it is termed sporadic MTC and if it coexists with tumors of the parathyroid gland and medullary component of the adrenal glands (pheochromocytoma), it is called multiple endocrine neoplasia type 2 (MEN 2). It was first characterized in 1959.<sup>2</sup> After assessment and treatment of associated endocrine conditions, a total thyroidectomy and central neck dissection are recommended. In cases with nodal metastasis to other levels, modified radical neck dissection is mandatory. In addition to the usual cancer follow up, patients should be followed with yearly chest x-ray as well as calcitonin levels. Serum calcitonin is very useful in follow up of medullary thyroid cancer because no other cells of the body make this hormone.<sup>3</sup> Herein we report an unusual case of MTC.

## CASE REPORT:

A 60 year old male presented with a large right neck

swelling for 2 years. It gradually increased in size and became static after one year. Six months later he started difficulty in breathing especially on exertion. On examination there was huge neck swelling predominantly on right side with engorged blood vessels without any visible pulsation. (Fig I) There was no dysphagia, hoarseness or stridor and signs of thyrotoxicosis were also absent. Fiberoptic laryngoscopy showed bilateral mobile vocal cords. His past medical and surgical histories were insignificant. His family history was also insignificant for such disease.

CT scan revealed a large lobulated mass extending from neck on right side going retrosternal (Fig II) and up to right main pulmonary artery, pushing the larynx and trachea on left. Mass was predominantly in anterior mediastinum abutting and compressing left brachiocephalic vein over arch of aorta. This mass



**Figure I:** Patient with neck lumps

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was 9cm x 11.3cm transversely and 19cm craniocaudally. FNA was suggestive of medullary thyroid carcinoma. His multiple endocrine neoplasia (MEN) syndrome workup was negative.

Finally the patient underwent total thyroidectomy with central neck dissection and removal of mediastinal mass via median sternotomy. Postoperative course was uneventful. He was regularly followed for 8 months without any evidence of recurrence after which he was lost to follow up.

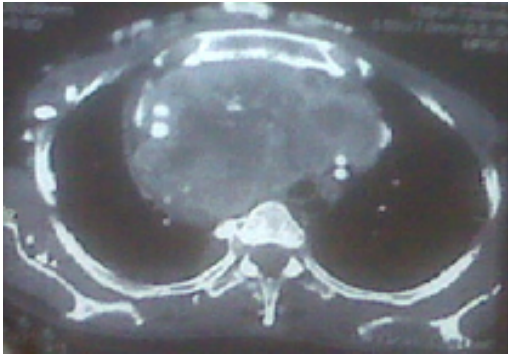


Figure II: CT scan shows mass with retrosternal extension.

#### DISCUSSION:

MTC constitutes approximately 4% of all thyroid cancers in the United States.<sup>4</sup> Mutations in the *RET* proto-oncogene is seen in most cases of familial medullary carcinoma which confer varying degrees of risk<sup>5</sup> and prophylactic thyroidectomy can now be offered to specific types of patients with this genetic abnormality. Peak incidence of isolated medullary thyroid carcinoma occurs in the fifth or sixth decade of life, and the peak incidence of MTC associated with multiple endocrine neoplasia (MEN) 2A or 2B occurs during the second or third decade of life.

The clinical presentation usually differs; however, one or more of the following symptoms may be observed. Patients may present with lump in the neck, hoarseness, dysphagia, or respiratory difficulty. Although uncommon, they may show symptoms of Cushing or carcinoid syndrome. Diarrhea may occur from increased intestinal electrolyte secretion secondary to high plasma calcitonin levels. Symptoms of weight loss, lethargy, and bone pain are observed if there are distant metastases.

Surgery is the mainstay therapy for the MTC. Prior to surgery, it is important to rule out a concomitant pheochromocytoma. Total thyroidectomy and a central neck dissection are recommended for all patients with MTC. A modified radical neck dissection is recommended for cervical metastasis. Selected

patients although controversial, may benefit from external beam radiation therapy and chemotherapy postoperatively.<sup>6,7</sup> Postoperatively, a serum calcitonin level should be obtained. Elevation of the serum calcitonin level indicates persistent disease. Our case was one of its kind and literature search of literature did not reveal such huge tumour extending from neck to mediastinum.

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