

A Case of Medullary Carcinoma of Thyroid

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ABSTRACT

A 50-year-old diabetic female attended in September, 2021 with a huge swelling in front of her neck, respiratory distress, hoarseness of voice, & generalized weakness for 1 month. All the biochemical tests were inconclusive but S. calcitonin was high(>0.08ng/ml), Fiber optic laryngoscopy revealed left vocal cord paralysis, X-ray chest revealed right sided tracheal deviation, CT-scan of Neck revealed large solid mass in thyroid with mediastinal extension & cervical lymphadenopathy. Patient undergone total thyroidectomy. The specimen sent for histopathology and revealed medullary carcinoma of thyroid gland.

Key words: Medullary carcinoma, stridor, total thyroidectomy, mediastinal extension.

Introduction

Medullary carcinoma of Thyroid (MTC) is about 5% of all thyroid neoplasms with the pathology of para follicular C cell and a higher calcitonin level. Genetic (MEN IIA, MEN IIB) association and family history are also common etiology in this type. The tumor being aggressive in nature, mediastinal, lung and bone metastasis are common features of it. Sometimes in spite of not having any endocrinopathy, post-surgical follow up for calcitonin level is a necessary need for recurrent or residual tumors. Clinically dyspnea, dysphagia, voice hoarseness are clues to suspect this variety of neoplasm of thyroid but Fine Needle Aspiration cytology (FNAC), Serum calcitonin and exclusion of genetic predisposition and family history is confirmatory. However, surgery is the mainstay of treatment in this type of tumor with removal of nodes up to level VI even if not involved. Radioactive iodine and chemotherapy playing no role here despite recently advanced developments of treatment technologies has been in progress like being in researches that some targeted therapies like tyrosine

kinase inhibitors can reduce symptomatic metastatic neoplasms. Other than differentiated thyroid cancers, MTC cells doesn't uptake iodine, and iodine-131 treatment is not effective¹. In this case report, total thyroidectomy relieved the left recurrent laryngeal nerve involvement and tracheal compression with the stridor but there was no need of post-operative tracheostomy and naso-gastric feeding tube insertion, with a good voice is a remarkable achievement.

Case Report:

A 50-year-old diabetic, euthyroid, normotensive female came with the complaints of a very big neck swelling that grew very rapidly in last 1 month. She also complained of dysphagia, respiratory distress, hoarseness of voice and generalized weakness of same duration. She has no history of diarrhea, cough or joint pain. She has no family history of any cancer, no history of significant weight loss. She was on metformin 500mg twice daily but her sugar profile was always in normal range.

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On clinical examination, a huge swelling (8cm x 6.5cm x 5cm) on the anterior aspect of neck in the midline mostly in upper and middle portion of thyroid gland is obvious in inspection without and engorged vein or visible pulsation. On palpation, the swelling was hard, fixed with underlying structure, immobile, normal surface temperature also with mediastinal extension. There were also palpable lymph nodes (level II-V). Trachea was shifted to the right. On auscultation there was no aortic bruit, no rhonchi or crepitation sound on chest but stridor present. On investigation, CBC revealed high ESR (110mm), Serum calcitonin was 10.89ng/ml which is significantly higher than normal (0.08ng/ml).

On Chest x ray, there was clear lung field but trachea was extremely deviated on right side due to the compression of the mass, fiber optic laryngoscopy revealed left vocal cord paralysis, USG of neck revealed a large solid mass of about 6cm X 5cm sized multinodular goiter suggesting FNAC. On CT-scan of neck there was finding of a large mass of about 8cm X 6.5cm X 5cm in the left lobe of thyroid gland with remarkable rightward compression over trachea and mild intra-thoracic extension. With the respiratory distress and dysphagia patient undergone total thyroidectomy and also intra-thoracic extension was cleared and ipsilateral elective neck dissection (level II-V) was done, total specimen was sent for histopathology and revealed medullary carcinoma of thyroid gland.

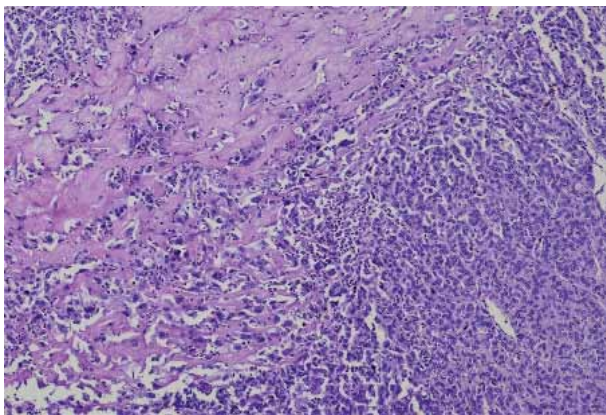


Figure 1. Histopathological photomicrograph of Medullary Carcinoma of Thyroid.

Discussion:

Medullary carcinoma of thyroid originated from para follicular C cells of thyroid. Most of medullary carcinoma are sporadic (80%), although minority of them may occur in hereditary form (Multiple Endocrine Neoplasm type-2 (MEN2)². High calcitonin and carcino-embryonic antigen are key to diagnose this cancer variety but FNAC and elevated calcitonin are the hallmark.

Histopathology report revealed a malignant tumor composed of solid proliferation of round to polygonal cells with amphophilic cytoplasm arranged in nested trabecular and pseudopapillary patterns and deposition of amyloid in extensive areas within tumor, also with some extensive areas of necrosis (Figure 1). Clinical picture of medullary carcinoma resembles pain, dyspnea, dysphagia, voice hoarseness, diarrhea, palpable neck nodes. Lymph node, blood, lung, mediastinum metastasis is common afterwards. Recurrence of the tumor is also characteristic.

In our case, patient had dyspnea, dysphagia, hoarseness, palpable lymph nodes on left side (level II-V). In terms of physical exam, before observing the lab results swelling was palpated suspicious for malignancy and then we also went through fiber optic laryngoscopy that revealed left vocal cord palsy, then for diagnostic purpose calcitonin was sent and proceed for CT-Scan of Neck. Higher Calcitonin levels being the hallmark and Ct-Scan Resulted in a massive mass with retro-sternal extension and deviating trachea on right and with other remaining normal investigations required for general anesthesia fitness. We decided the case was obvious for surgical exploration with central clearance.

So, we undertook Total Thyroidectomy with selective neck dissection (level II-V) and along with central clearance which is the preferred procedure. After surgery, the specimen was sent enbloc for histopathology and revealed medullary carcinoma of thyroid gland. Medullary carcinoma accounts for as much as 13% of all thyroid cancer-related deaths^{3,4}. Survival rates for 10 years range from 56% to 96%

respectively with adequate treatment. Proper identification through imaging, calcitonin level and proper exclusions of family history and MEN IIA, MEN IIB genetic predisposition are necessary too. Serum calcitonin and carcino embryonic antigen levels are measured every three-months after surgery, the NCCN guidelines recommends yearly serum calcitonin and CEA test⁵.

Conclusion:

If proper management or surgery can be done in early time, survival rate will be more. Our patient was good till the last follow-up.

Conflicts of interest: None

References:

1. Martins RG, Rajendran JG, Capell P, Byrd DR, Mankoff DA. Medullary thyroid cancer: options for systemic therapy of metastatic disease? *J Clin Oncol.* 2006; 24(11): 1653–5.
2. Saad MF, Ordonez NG, Rashid RK, Guido JJ, Hill Cs, Jr, Hickey RC, et al. Medullary carcinoma of the thyroid. A study of the clinical features and prognostic factors in 191 patients. *Medicine (Baltimore)* 1984; 63(6): 319–42.
3. Hung WW, Wang CS, Tsai KB, Ou-Yang F, Shin SJ, Hsiao PJ. Medullary thyroid carcinoma with poor differentiation and atypical radiographic pattern of metastasis. *Pathol Int* 2009; 59: 660-3.
4. Hsieh MH, Hsiao YL, Chang TC. Fine needle aspiration cytology stained with Rius method in quicker diagnosis of medullary thyroid carcinoma. *J Formos Med Assoc* 2007; 106: 728-35.
5. Ha EJ, Na DG, Baek JH, Sung JY, Kim JH, Kang SY. US Fine-Needle Aspiration Biopsy for Thyroid Malignancy: Diagnostic Performance of Seven Society Guidelines Applied to 2000 Thyroid Nodules. *Radiology.* 2018; 287(3): 893-900.