PriMera Scientific Surgical
Research and Practice
Volume 2 Issue 5 Noember 2023
DOI: 10.56831/PSSRP-02-067
ISSN: 2836-0028

PriMera Scientific Publications

Surgical Management of Medullary Thyroid Carcinoma with Retrosternal Extension: A Case Report with Review of Literature

Type: Case Report
Received: September 09, 2023
Published: October 20, 2023

Citation:
Rajesh Kantharia., et al. "Surgical Management of Medullary Thyroid Carcinoma with Retrosternal Extension: A Case Report with Review of Literature". PriMera Scientific Surgical Research and Practice 2.5 (2023): 19-24.

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Abstract

Medullary Thyroid Carcinoma is derived from parafollicular C-cells that normally secretes peptide hormones such as calcitonin, serotonin and vasoactive intestinal peptides and thus it is widely accepted as a neuroendocrine tumor. MTC usually has a good prognosis with a 10 years survival rate of approximately 95.6% in cases restricted to the thyroid gland, but reduces to 40% for those presenting with metastasis. Surgery is considered the primary and best modality of treatment in Medullary Thyroid Carcinoma. Here we present a case of a successful surgical management of a 38 year old female patient of sporadic, medullary thyroid carcinoma with sub sternal extension of the disease.

Keywords: MTC; Sporadic; Total Thyroidectomy; Sub-Sternal Extension

Introduction

Medullary thyroid carcinoma(MTC) accounts for 1-2% of all thyroid malignancies [1]. MTC is usually derived from parafollicular C-cells that normally secretes peptide hormones such as calcitonin, serotonin and vasoactive intestinal peptides and thus it is widely accepted as a neuroendocrine tumor [2]. The hereditary form of MTC is usually concerned with the RET proto-oncogene and account for about 25% of MTC diagnosis, while the remaining 75% are believed to be of the sporadic form.3Familial forms are inherited via autosomal dominance with germline mutations on chromosome 10q11 [3]. This form of MTC is also associated with other endocrine tumors in syndromes known as multiple endocrine neoplasia (MEN) 2A and MEN 2B [4]. Those with MEN 2A, in addition to MTC, have a greater risk of developing pheochromocytoma and parathyroid adenomas. MEN2B, on the other hand, includes MTC, mucosal neuromas, pheochromocytomas, and marfanoid body habitus. Although several forms of biochemical markers such as plasma carcinoembryonic antigen (CEA), serotonin, chromogranin A and tryptase, has been associated with MTC but clinically, calcitonin has been known

to be far superior for diagnosis, maintenance, and prognosis of the disease. Prognostically MTC has a 10 years survival rate of 95.6%in cases restricted to the thyroid gland, but reduces to 40% for those presenting with metastasis [5]. Surgery is considered the primary and best modality of treatment in Medullary thyroid carcinoma. Here we present a case of sporadic, medullary thyroid carcinoma with sub sternal extension.

Case Report

A 38 year old female patient, who underwent a total thyroidectomy, a month back for Medullary Thyroid Carcinoma of the left lobe of thyroid, presented in the department for follow up and second opinion. Patient was asymptomatic when she presented to the department with no complains of change in voice or dysphagia. On neck examination surgical scar was present. No clinically palpable swelling was present on the midline. Fullness in the left to groove Left level II node was palpable, firm with restricted mobility of size approximately 2*3 cm. Hopkin's examination showed equal mobility of the bilateral vocal cords. She had raised levels of serum calcitonin level of 9720 pg/ml. Ultra -sonography of neck showed a large, well defined mass of size (2.8X5X4) cm in the pre and left paratracheal region inferior to the thyroid bed. Enlarged lymph node of size (2.8X2cm) was also noted at left level II. Fine Needle Aspiration Cytology from the same lesion was consistent with recurrence in known case of Medullary Carcinoma of Thyroid. Whole body PET CT scan with FDG-18 was done to rule out any distant metastasis. MRI of the neck extension of the lesion from lower edge of C7 vertebra upto the lower edge of D3 vertebra with medial extension into the trachea-oesophageal groove, compressing the trachea and cervical oesophagus contalaterally. We also did FNAC from the left groove lesion and repeated S. Calcitonin. The patient underwent a completion thyroidectomy along with central compartment clearance and left selective neck dissection(level II-IV) with a single lower neck crease incision. No additional approach or incision was required to expose the sub sternal disease which was retrieved into the neck with the help of blunt dissection. Intra-operative recuurent laryngeal nerve monitoring was done and the tumor was retrieved without damaging the Recurrent laryngeal nerve. The histopathology report came to be pTxN1bMx with completion left side thyroidectomy specimen with organizing granuation tissue and no residual thyroid tissue or malignancy.2/2 central compartment nodes were consistent with metastatic MTC with extranodal extension,1/1 left pre tracheal node was metastatic MTC without extranodal extension and 3/5 left level II-V cervical node was metastatic MTC with extranodal extension. No post operative complications were noted and the patient was discharged on the 5th post operative day after the drain removal. Patient had hoarseness of voice in the post operative period.

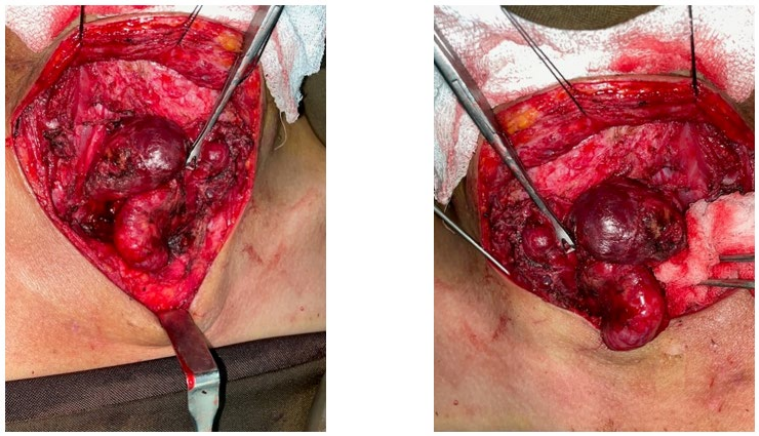

Figure 1a and 1b: Residual thyroid tissue along with the retrosternal nodal mass dissected retrosternal region.

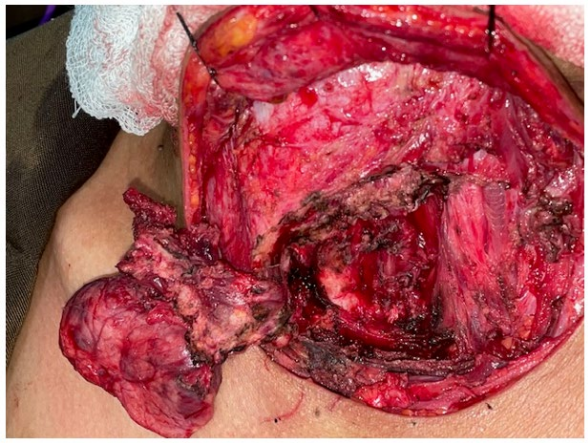

Figure 2: Thyroid bed containing intact recurrent laryngeal nerve along with the residual thyroid tissue and nodal mass +completely dissected from the tumor bed.

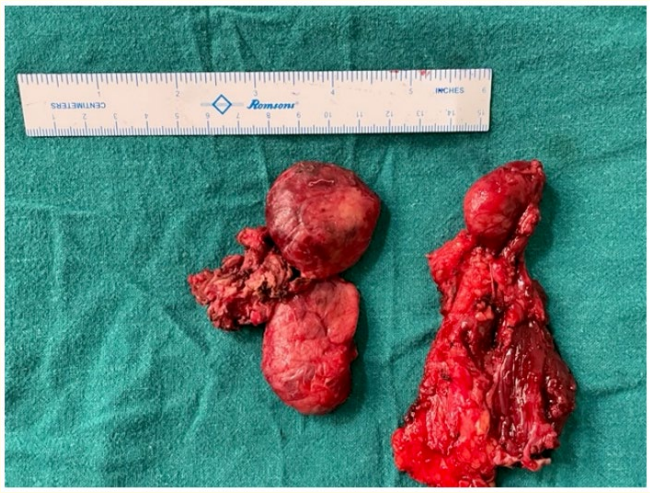

Figure 3: Ressected residual tumor and nodal specimen.

Discussion

In 1959 Hazard and associates provided a definitive histological description of MTC and named it likewise [6, 7]. Sporadic MTC usually occur in fifth or sixth decade of life. However, hereditary MTC is more common in the younger age group, of which multiple endocrine neoplasia type 2A (MEN 2A) and familial thyroid cancer usually arise when patients are in their 30s, and MEN 2B typically presents in those who are <20 years of age. Hereditary forms are transmitted as an autosomal dominant trait either alone as familial MTC (FMTC) or as part of MEN 2A or 2B [8, 9] Germline mutations of the RET proto-oncogene (RET) found on chromosome 10q11 are responsible for FMTC s and may be present in more than 95% of the hereditary MTCs and about 25% of the sporadic MTCs [10]. Our patient was diagnosed at 38-years of age based on a neck swelling and was operated at another centre, followed by which she came for

a second opinion at our centre when final HPR showed Medullary Carcinoma of the thyroid. Although pain or aching is also a common symptom of MTC our patient was totally asymptomatic. Our patient did not have any symptoms of local tumor invasion such as dysphagia, dyspnea, or dysphonia. In MTC solitary thyroid nodules may be associated with palpable cervical lymphadenopathy (15% to 20% of the cases) [11]. The elevated Serum Calcitonin levels along with the USG findings in our patient resulted in an extensive work-up that included a PET/CT scan to exclude any distant metastasis. The PET-CT scan showed moderate grade FDG avid enhancing nodular lesion in the operated thyroid bed suggesting a recurrent/residual disease.No evidence of metabolically active disease was seen anywhere else in the body. An ultrasound was performed to confirm that the focal metabolic uptake seen on the PET/CT was a well defined mass lesion of approximate size(2.8X5X4)cm in the left paratracheal region just below the thyroid bed. Patients with extensive metastatic disease frequently develop diarrhea, which may result from increased intestinal motility and impaired intestinal water and electrolyte flux due to high calcitonin levels [12]. Despite having calcitonin levels of 9720 pg/ml our patient did not have diarrhea or flushing. It has also been noted that 2% to 4% of patients develop Cushing syndrome as a result of ectopic production of ACTH.

FNAC from the left paratracheal region proved to be recurrence in a known case of medullary thyroid carcinoma. The sensitivity of FNA for the diagnosis of MTC is 50% to 80%, though a higher sensitivity can be achieved by adding the immunohistochemical staining for calcitonin [13-15]. In case of high suspicion of MTC, calcitonin can be measured in the washout of the FNA biopsy needle.

The treatment decided for this patient was upfront surgery. Although, recurrences are a poor prognostic factor which may lead to greater morbidity, total thyroidectomy along with central compartment clearance is the treatment of choice according to ATA Guidelines. The incidence of central neck metastasis can be as high as 81% in patients with palpable tumors [16] and thus, central compartment neck dissection provides a better survival and cure rate than total thyroidectomy alone [17]. The role of routine lateral neck dissection in treating patients with MTC is less clear. Ipsilateral lateral nodal metastases may be present in 14-80% of the patients and contralateral lateral nodal metastases have been described in 19-49% of the patients [17, 18]. Since there is a huge risk of neck metastasis, even in tumors Cervical recurrences require early diagnosis and adequate surgical treatment to achieve good oncological results. Thus, in several literatures, more that 90% cure rate have been reported post surgery in recurrent cases.

The current ATA guidelines recommend that patients without nodal metastases on ultrasonography and no distant disease undergo a total thyroidectomy along with unilateral level VI clearance prophylactically. In this scenario, no consensus was reached regarding the optimal management of the lateral compartments, and the guidelines indicate that a prophylactic lateral neck dissection may be considered based on calcitonin levels (recommendation #25 ATA guidelines). Prophylactic dissection of the contralateral neck can be considered if the calcitonin level is greater than 200 pg/mL (23). In our patient no residual disease was found on the thyroid bed but a left central compartment node was present which was extending into the retrosternal area. Hence only left side central compartement clearance was done. Blunt dissection was used to release the node from the adjacent soft tissue, and vessels and the recurrent laryngeal nerve was traced from the trachea-oesophageal groove running upward behind the thyroid gland and the entire dissection was carried out keeping the recurrent laryngeal nerve under vision, thus protecting it from inta-operative injuries. Once the node was dissected from its retro-sternal attachments the entire specimen was pulled up into the neck and specimen was delivered after freeing from the adjacent soft tissue attachments.

Although the recurrent laryngeal nerve was saved intra-operatively, patient had some hoarseness of voice from the post op day 1 which gradually decreased in the subsequent follow ups.

The postoperative follow-up should begin 2-3 months after operation and it is based on serum calcitonin and CEA levels. Patients with undetectable calcitonin levels postoperatively are usually followed with yearly measurements of serum calcitonin and CEA. In addition, ultrasound of the neck can also be done, but appears to have no added advantage. In case of patients with elevated levels of serum markers, ultrasound of the neck or CT/magnetic resonance imaging can be done.

The prognosis for patients with MTC seems good with a 10-year survival rate for patients with MTC being 75-85% [19-21]. About 50% of the patients with MTC have disease restricted to the thyroid gland, and have a 10-year survival rate of 95.6% [20]. Approxi-

mately, one-third of the patients present with locally invasive tumors or with metastasis to the regional lymph nodes. The survival rate of patients with regional disease is 75.5%. Distant metastases may be seen in 13% of the patients at the initial diagnosis and have a poor prognosis, with a 10-year survival rate of only 40%. Radio-active iodine therapy seems to have no role in MTC as the tumor originates from parafollicular C-cells which do not accumulate iodine. Both radiation therapy and conventional chemotherapy have limited place in the treatment of patients with MTC. Vandetanib and cabozantinib are the newer tyrosine kinase inhibitors which have been approved by the US Food and Drug Administration for the treatment of progressive and metastatic MTC.

Conclusion

MTC is an uncommon and a rare thyroid malignancy. Its management is different than that for differentiated thyroid cancers. Early diagnosis offers a higher likelihood of cure and long-term survival. Surgery is the mainstay of treatment followed by regular follow-up to diagnose recurrences early and treat accordingly.

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