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Medullary carcinoma of thyroid: Retrospective analysis

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Abstract

Objective: To describe outcome of patients with medullary carcinoma of thyroid.

Methods: Retrospective descriptive study of patient record from 1999 to 2011. Study includes patients treated in surgical oncology department of NIMS, Hyderabad

Result: Surgical resection is the mainstay of the treatment. Perineural invasion is a adverse prognostic feature in addition to positive cervical nodes and positive margin.

Conclusion: Medullary carcinoma thyroid are rare neuroendocrine tumor of thyroid. Treatment is complete surgical resection which may require viscera resection. Adjuvant RT should be given in case of adverse prognostic factors like perineural invasion, positive margins and positive nodes.

Keywords: Medullary carcinoma thyroid

1. Introduction

Medullary carcinoma of thyroid is uncommon neuroendocrine tumor arising from parafollicular C cells^[1]. Calcitonin is secreted by C cells and it primarily inhibits osteoclastic bone resorption and promotes calcium excretion by kidney^[2]. They account for 5-10% of the thyroid cancer. Medullary thyroid cancer was first described by Hazard *et al.*^[3] Most of them are sporadic but around 25% are due to mutation in ret proto-oncogene. Hereditary medullary carcinoma thyroid is found in association with type 2A or type 2B^[4]. Type 2B has earlier onset and aggressive course. They can also occur as familial syndrome in small percentage of the case. Histopathologically it may be encapsulated or un-encapsulated with extension to adjacent parenchyma. Amyloid is seen in 80% of the tumor^[5]. On IHC the tumor cell stains positive for calcitonin, CEA and chromogranin A. They are known for early metastasis to the regional lymph nodes. In advanced cases they can invade local structures like trachea, esophagus, strap muscle and recurrent laryngeal nerve. Distant site of metastasis includes liver, lung and bone.

Surgery in form of total thyroidectomy with central compartment dissection is the treatment when basal calcitonin level is normal and stimulated calcitonine level is elevated. Patients with stimulated calcitonin level > 560 pg/ml or having palpable disease are considered for lateral neck dissection. Mediastinal nodes detected on imaging should undergo dissection via trans-sternal approach. In our retrospective analysis we reviewed our data from 1999 to 2011.

2. Material and methods

The study included patient treated at Nizam's Institute Of Medical Sciences; Hyderabad in Surgical Oncology Department. It is a retrospective descriptive study of patient record from 1999 to 2011. The data were entered in excel sheet and mean and percentage was calculated.

3. Results

Total numbers of patients were 21; 6 (28%) males and 15 (71%) females. Mean age was 35.2 years. 3 had history of previous thyroid surgery in the form of hemithyroidectomy. The average duration of symptoms was 48 months. Painless lump in neck was commonest presenting symptoms. Three patients presented with local compressive symptoms. 11 patients had palpable lymph node at presentation.

All patients were evaluated with neck ultrasound and diagnosed with FNAC. Serum calcitonin was also evaluated in all patient and those with markedly raised values underwent additional chest and abdominal CT scan. One had liver metastasis. One patient was found to have MEN 2A syndrome.

All patients underwent total thyroidectomy with central and or lateral neck dissection. In 2 patients RLN was sacrificed. Two patient required tracheal resection.

Histopathologically average tumor size was 2 x 3 cm. 4 patient (19%) had bilateral tumor. One patient had multifocal tumor. 3 patients had margins positive along with perineural invasion. 14 patients had involved nodes and average numbers of positive nodes were 13. Tumor size was found to be important predictor for lymph node involvement. Postoperatively 6 patients underwent RT for positive margins or extensive soft tissue infiltration. 2 patients lost to follow up. 3 patients died after 2 years of average follow up. Three patients had recurrence in contra lateral neck node and were salvaged with surgery. All others were doing well at the time of writing the article.

4. Discussion

Medullary thyroid cancer is rare neoplasm of thyroid. Females are affected more commonly as compared with males. Our literature shows that most of the patient presents late in their course of disease. The most common site for spread is central and lateral compartment nodes. Liver is the common site for distant metastasis. Tumor size is an important predictor for lymph node involvement and those with size greater than 2 cm frequently have lymph node metastasis. Bilateral involvement is seen in small percentage of the patient. Surgery is the only curative treatment.

Lymph node metastasis is most important prognostic factor. Perineural spread is adverse prognostic factor in our study along with positive margins. Those with residual disease unsuitable or re-resection and those with extensive lymph nodal metastasis or soft tissue involvement along with perineural spread should undergo adjuvant RT. Serum calcitonin is helpful in monitoring the course of disease and prognosis.

5. Conclusion

Treatment for medullary carcinoma thyroid is complete surgical resection which may require visceral resection in form of tracheal resection. Every effort should be made to preserve the parathyroids and RLN. Central compartment nodes are commonly involved and should be dissected during primary surgery. Perineural invasion is also an adverse prognostic factor in addition to positive lymph node and positive margin. RT should be used as adjuvant in these cases.

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