MEDULLARY CARCINOMA OF THYROID WITH HASHIMOTOS THYROIDITIS - A RARE CASEREPORT

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Abstract:
Medullary carcinoma of thyroid in association with Hashimotos thyroiditis is a very rare entity. It is reported with an increased predilection for women in their fifth decades. These tumors have an intermediate prognosis. Hereby, we report a case of medullary thyroid carcinoma with Hashimotos thyroiditis in a 50 year old woman who presented with neck swelling of four months duration. Fine needle aspiration cytology suggested the possibility of medullary carcinoma of thyroid. Serum calcitonin levels were markedly elevated. Total thyroidectomy was done. Histopathological examination and immunohistochemistry confirmed the diagnosis.

Keyword:
Medullary carcinoma, thyroidectomy, Hashimotos thyroiditis, calcitonin

INTRODUCTION:
Medullary carcinoma of thyroid is a malignant tumor exhibiting parafollicular C-cell differentiation. It comprises approximately 2.5% of all thyroid cancers.1 Hashimoto’s thyroiditis is predominantly a disease of women over 40 years of age2 and the overall association of medullary thyroid carcinoma with Hashimoto’s thyroiditis is very rare that constitutes to about 0.35%.3 Herein, we report a case of medullary carcinoma of thyroid in a 50 year old woman with Hashimoto’s thyroiditis.

CASE HISTORY:
A 50 year old female was admitted in the surgery ward with complaints of swelling in the anterior aspect of neck of four months duration. On examination, a butterfly shaped swelling was seen in front of the neck, firm in consistency and movement on deglutition.

An Initiative of The Tamil Nadu Dr. M.G.R. Medical University
University Journal of Pre and Para Clinical Sciences
was noticed. USG neck revealed a well defined heterogenous hypoechoic nodular mass involving the left lobe of thyroid gland and a possibility of malignancy was suspected. Fine needle aspiration cytology of the swelling showed features of medullary carcinoma of thyroid. Serum calcitonin levels were found to be markedly elevated (1164pg/ml) with the normal reference range being 14 to 16 pg/ml. Total thyroidectomy was done and the specimen was sent for histopathological examination. **FNAC THYROID**: Ø Aspirated cellular smear composed of plasmacytoid and spindle cells arranged in dyscohesive clusters and as dispersed single cells. The plasmacytoid cells displayed eccentric nucleus, a moderately abundant dense cytoplasm with well-defined cell borders. The spindle cells were elongated with pale nuclei and uniform stippled nuclear chromatin with indistinct cytoplasm. (Figure 1)

**Figure-1**: FNAC 40x-Plasmacytoid and spindle cells with fine stippled nuclear chromatin measuring 3x2x1cm and isthmus measuring 1x1cm. Cut surface of the left lobe revealed fairly circumscribed gray-white areas measuring about 4x1cm with adjacent normal thyroid parenchyma. Cut surface of isthmus and right lobe revealed gray-tan areas with vague nodularity. Serial sectioning also revealed the same features. (Figure 3)

**FNAC 40x-Binucleated forms of neoplastic cells.**

**Impression-Medullary Carcinoma of thyroid.**

**GROSS:**

Received total thyroidectomy specimen, with the left lobe measuring 8x5x3cm, smaller lobe measuring 3x2x1cm and isthmus measuring 1x1cm. Cut surface of the left lobe revealed fairly circumscribed gray-white areas measuring about 4x1cm with adjacent normal thyroid parenchyma. Cut surface of isthmus and right lobe revealed gray-tan areas with vague nodularity. Serial sectioning also revealed the same features. (Figure 3)

**Figure-3**: Gross-Total thyroidectomy specimen with cut surface of the left lobe reveals a relatively circumscribed gray white area with adjacent normal thyroid parenchyma. Cut surface of the right lobe and isthmus reveals diffuse, gray brown areas with vague nodularity.

**MICROSCOPIC FEATURES**: Sections studied from the left lobe of thyroid showed a neoplasm composed of tumor cells arranged in sheets and solid nests separated by delicate fibrovascular septa. The individual cells were polygonal to spindle with round to oval nuclei, finely stippled chromatin and indistinct nucleoli with finely granular cytoplasm. (Figure 4)
Massive deposits of amyloid were seen as amorphous eosinophilic material in the intercellular stroma. Sections from the right lobe of thyroid revealed a lesion composed of atrophic thyroid follicles with extensive lymphocytic infiltration of the stroma, formation of germinal centers and oncocytic change of the follicular epithelium. (Figure 5)

**Figure - 5: 10x-Germinat centers, oncocytic follicular epithelium**

With these features, histological diagnosis of medullary carcinoma with Hashimoto’s thyroiditis was arrived at.

**SPECIAL STAIN:** CONGORED stain highlights the amyloid deposits (Figure 6)

**Figure - 6: 10x- Amyloid deposits, Congo red stains the amyloid**

**IMMUNOHISTOCHEMISTRY:**

The tumor cells showed immunocytochemical positivity for chromogranin and calcitonin. (Figure 7,8)

**DISCUSSION:**

Hashimoto’s thyroiditis, also known as “struma lymphomatosa” is a form of autoimmune thyroiditis predominantly seen in women over 40 years of age. Grossly, it presents as diffuse and symmetrical enlargement of the thyroid gland. Cut surface is gray-tan, vaguely nodular with firm consistency reminiscent of a hyperplastic lymph node. Microscopically, the two characteristic features are lymphocytic infiltration of the stroma and oxyphilic change of the follicular epithelium. Most of the
follicles are atrophic with prominent germinal centers. The rate of occurrence of thyroid malignancies in patients with Hashimoto’s thyroiditis is about 7-10% with the most common reported malignancies being papillary carcinoma and lymphoma. The association of medullary thyroid carcinoma with Hashimoto’s thyroiditis is very rare. Medullary carcinoma of thyroid is a malignancy of parafollicular C-cells, also termed as C-cell carcinoma and solid carcinoma with amyloid stroma. It accounts for 2.5% of all thyroid malignancies. Grossly, the tumor is solid, firm, relatively well circumscribed with a gray to yellow cut surface. Most tumors are located in the midportion of the gland, the region where the C-cell density is highest. Microscopically, the tumor is composed of solid nests or sheets of round to polygonal cells traversed by delicate fibrovascular septa. The tumor cell nuclei are oval with finely stippled nuclear chromatin and indistinct nucleoli. The cytoplasm is amphophilic and finely granular. Extensive amyloid deposits are seen in about 80% of the cases. The typical immunohistochemical profile of medullary carcinoma of thyroid is its positivity for pan-neuroendocrine markers, cytokeratin, calcitonin and carcinoembryonic antigen. The majority of medullary thyroid carcinomas are sporadic in occurrence (70-80%). The hallmark of these tumors is the presence of increased serum calcitonin levels. The prognosis is considered to be of intermediate type. It is suggested that the prognosis of patients with carcinoma of thyroid with co-existing Hashimoto’s thyroiditis is better than that of patients with carcinoma of thyroid gland alone.

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