CASE REPORT

Carcinoma Showing Thymus-like differentiation (CASTLE) - A Rare Thyroid Carcinoma

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ABSTRACT

This is a case report of a rare thyroid tumor "Carcinoma showing thymus-like differentiation" (CASTLE). These tumors are usually in the lower part of the thyroid gland. They present like other advanced thyroid cancers but it is important to differentiate CASTLE from these. CASTLE tumors of the thyroid have better prognosis than thymic carcinoma, squamous cell carcinoma and anaplastic carcinoma and hence curative surgery and adjuvant radiotherapy lead to over 80% 10 year survival rate. This case report describes how emergency surgery on a dyspneic patient followed by external radiation therapy was successful.

Keywords

Thyroid Carcinoma showing thymus-like differentiation (CASTLE)

Introduction

CASTLE, a rare thyroid tumor was first described in 1985 by Miyauchi et al as an "intrathyroid epithelial thymoma Carcinoma"[1]. In 1991 Chan and Rosai coined the term "Carcinoma showing thymus-like differentiation" (CASTLE) for this rare tumor of the thyroid gland. They also proposed that it originated from ectopic thymus, or from thymopharyngeal duct or branchial pouch remnants in the thyroid gland and neck[2]. These tumors are usually in the lower part of the thyroid gland though they can arise in the extra thyroidal soft tissue of the neck[3-4]. There are very few reported cases of CASTLE [1-25]. Due to their rarity there are no official guidelines about their management. The prognosis is favourable compared to thymic carcinoma, squamous cell carcinoma and anaplastic carcinoma; hence it is important to differentiate it from these and to treat it aggressively.

Case report

A 55 year old female in respiratory distress, presented in the emergency room with a neck swelling since 35 years, dry cough since 3 months and progressive dyspnea since 1 month. On examination she had stridor, a large nodular goiter, distended right neck veins, and the right side much larger than the left with trachea grossly displaced to the left side and carotid displaced posteriorly. Lower border of the goiter was not palpable. There were no palpable lymph nodes. She was euthyroid. She had with her 1 week old Ultrasound scan, FNAC, Chest X ray reports. She had refused surgery then out of fear of death at surgery.

Basic blood tests, cross match were done, high risk consent taken and she was taken up for surgery immediately. After Bronchoscope guided intubation, through a mid-neck skin crease incision near total thyroidectomy was performed leaving residual tissues on the arch of the aorta and right brachiocephalic trunk to which the hard right lower pole of thyroid was stuck. The goiter had the appearance and feel of a multinodular goiter with secondary changes except the right lower pole that looked and felt like an anaplastic carcinoma and had to be shaved of the trachea and carotid artery. Frozen section facility was not available at

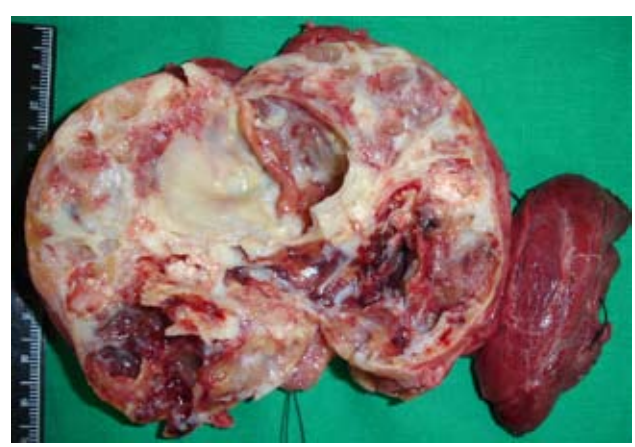

Figure 1: Cut section of right lobe of thyroid with silk suture on lower pole and an intact left lobe

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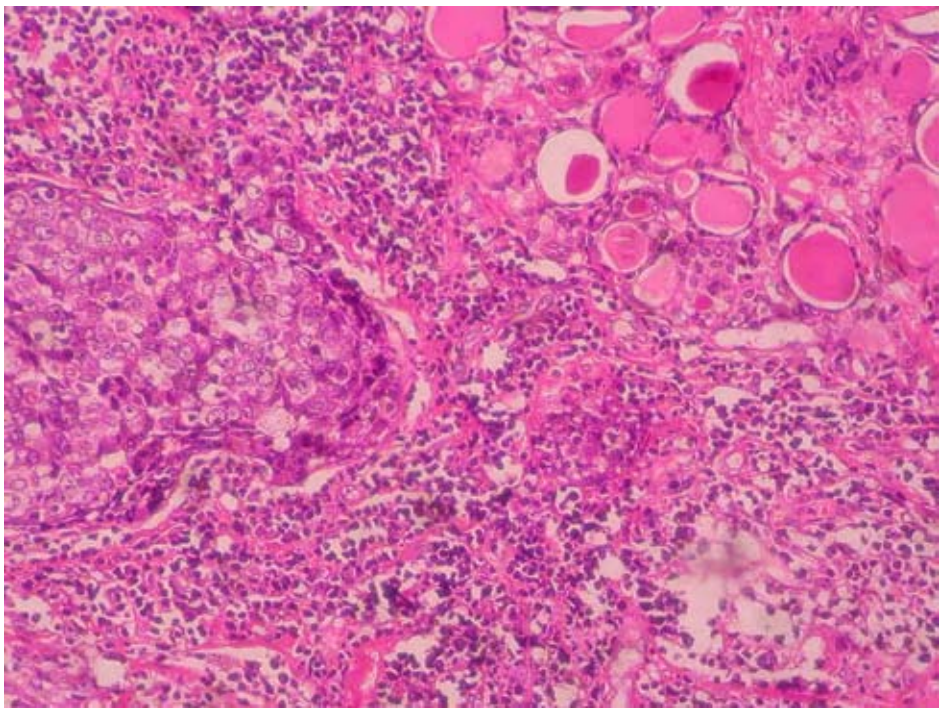

Figure 2: Section at 10x magnification showing tumor infiltrating thyroid follicles

night during surgery. One unit of whole blood had to be transfused. Post op recovery was rapid and there was no voice change or hypocalcemia.

On gross examination of the specimen the right lobe was 10x5x3cms, the left lobe 6x4x2cms. Cut section revealed nodular grey white to grey brown areas with cystic changes and a 2x1cms grey white nodule in the right lower pole. Microscopic examination revealed features of multinodular goiter except in the right lower pole. Highly pleomorphic cells infiltrating into the thyroid follicles along with lymphocytes and plasma cells were seen in the right lower pole. Immunohistochemistry (IHC) staining was positive for EMA, Cytokeratin, CD117, BCL2 and negative for CDS, CEA, LCA and TTF. A final diagnosis of intra-thyroidal epithelial carcinoma showing thymus-like differentiation (CASTLE) was reported.

After returning to her home in the state of UP, she underwent external radiation therapy and was asymptomatic three years later as per telephonic message from her son.

Discussion

CASTLE usually arises in the lower part of the thyroid gland, in the age group 40 to 50 years, and presents as a painless, slow-growing cervical mass [14]. Preoperative diagnosis is very difficult, because it presents just like other advanced thyroid cancers as a hard mass with poor mobility due to invasion of adjacent organs and enlarged regional lymph nodes. It cannot be diagnosed on imaging either. A solid, hypoechoic mass without calcification but with moderate vascularity is seen on sonography.

Usually these tumors on gross section are well circumscribed, well separated from surrounding thyroid tissue and have a firm, gray cut surface. Cytological appearance of undifferentiated epithelial cells with lymphoid cells is similar to that of a carcinoma in the background of lymphocytic thyroiditis. It closely resembles thymic carcinoma in histopathology and on IHC staining. CASTLE must be differentiated from lymphoepitheliomas, malignant lymphoma, poorly differentiated squamous carcinoma, thymic carcinoma and anaplastic carcinoma.

CASTLE tumors grow less rapidly than squamous cell carcinoma of the thyroid[1]. In a collaborative study by Ito et al the 5- and 10-year cause-specific survival rates were 90% and 82%, respectively. Nodal metastasis and tumor extension predicted a worse prognosis. Of 22 patients who had curative surgery, 10(45%) underwent adjuvant radiation therapy, and no locoregional recurrence was seen in any of them[14]. Due to their rarity there are no official guidelines about their management. The prognosis is favourable compared to thymic carcinoma, squamous cell carcinoma and anaplastic carcinoma; hence it is important to differentiate it from these. Total thyroidectomy, resection of invaded adjacent organs and neck dissection should be performed followed by external radiotherapy to prevent locoregional recurrence and to improve long-term survival.

Conclusion

This case report confirms earlier literature on CASTLE tumors of the thyroid having better prognosis than thymic carcinoma, squamous cell carcinoma and anaplastic carcinoma. It supports other reports of good survival rate with curative surgery and adjuvant radiotherapy.

Acknowledgement: I acknowledge the efforts and contribution of Dr. K.M. Prathima, former senior consultant pathologist at Gokula Metropolis clinical laboratories in arriving at the diagnosis of CASTLE.

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