Parathyroid Carcinosarcoma: A Case Study

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EDUCATIONAL OBJECTIVE: At the conclusion of this presentation, the participants should be able to understand the diagnosis and treatment of parathyroid carcinoma, as well as gain awareness of parathyroid carcinosarcoma, an even more rare clinical entity.

Objectives: Parathyroid Carcinosarcoma was first described by Nacamuli et al. in 2002. We present the second reported case of this entity.

Methods/Study Design: We present a case report of a patient with parathyroid carcinosarcoma and review the relevant literature. A 57 year old woman with long standing right sided vocal cord paralysis in association with a progressive 3.3 x 3 cm mass in the right neck. She had previously undergone a total thyroidectomy at an outside hospital, revealing benign pathology. Parathyroid hormone and calcium blood levels were within normal limits. Results: The mass was surgically removed with negative surgical margins. Histopathology and immunohistochemistry was consistent with parathyroid carcinosarcoma. The patient received adjuvant chemoradiation therapy, and seven months post-operatively she has no evidence of local regional recurrence but has developed progressive pulmonary metastases.

Conclusions: This is only the second reported case of parathyroid carcinosarcoma. In our patient as well as the previously reported case, the disease has shown systemic progression despite aggressive surgical resection and adjuvant therapy.

Introduction

Parathyroid carcinoma is rare, representing less than 4% of cases of parathyroid disease. Characterized by extremely high serum calcium and PTH levels, often along with impairment of vocal fold mobility. Signs and symptoms of this profound hyperparathyroidism include:

- Generalized osteopenia
- Osteoporosis resulting in pathologic fractures
- Gastrintestinal disturbances
- Central nervous system manifestations including weakness, fatigue and depression

Parathyroid carcinoma is even more rare with only one other case being reported in the literature.

In the previously reported case, the serum calcium and PTH levels were mildly elevated.

Case Report

A 57 year old woman with long-standing right-sided vocal fold paralysis underwent a workup by an outside otolaryngologist consisting of a thyroid ultrasound that demonstrated a right-sided thyroid nodule. The patient underwent a total thyroidectomy and initial pathology was found to be benign. Subsequent MRI showed a non-palpable right neck mass. FNA found a neuroendocrine tumor consistent with a parathyroid tissue. Serum calcium and PTH levels were within normal limits (total serum calcium 9.8 mg/dl and serum PTH 47 pg/mL). Pathology

Initial physical exam was notable for dysphonic voice, a 2.5 cm palpable right neck mass in the right paratracheal bed, and an immobile right true vocal fold. CT and MRI demonstrated a 2.8 cm x 1.8 cm mass in the right neck. The mass was subsequently excised.

Histopathologic diagnosis of parathyroid carcinoma is often difficult with criteria first set by Shariz and Castlemann in 1973 describing the presence of fibrous trabeculae, rosette-like cellular architecture, the presence of mitotic figures, and capsular or vascular invasion. Staging and treatment guidelines have been suggested for parathyroid carcinoma; however a lack of experience has made them limited for the treatment of parathyroid carcinoma with chemotherapy and radiation therapy. The American Joint Committee on Cancer (AJCC) has no staging for parathyroid carcinoma because of the rarity of the condition. Even so, some retrospective studies show decreased risk of localized disease recurrence with the addition of radiation therapy, although, these studies lacked statistical power due to small sample size.

Chemotherapy has been tried with limited results with one report showing a reduction in hypercalcemia for 13 months. Other trials have not been as successful. The proposed treatment for parathyroid carcinoma is local resection with post-operative adjuvant therapy. Metastatic lesions are either resected or treated with a combination chemotherapy radiation therapy when resection is not possible. In our patient, this provided adequate locoregional control but did not cure her systemic disease.

Discussion

Tumors of the parathyroid gland usually come to attention as the result of excessive PTH secretion. About 75% are represented by parathyroid adenomas, which are hormonally active, non-malignant tumors. Parathyroid carcinomas (75-85%), followed by parathyroid hyperplasia (15%), and a small minority are the result of parathyroid carcinoma (3%). Notably, the vast majority of those patients with parathyroid carcinoma are women. Thirty percent of patients with parathyroid carcinoma will have elevated calcium and PTH levels, respectively. 15-20% of patients remain asymptomatic. Elevations in calcium and PTH levels are more pronounced in parathyroid carcinoma, in the range of 14 mg/dl for serum calcium and up to 5-fold the normal levels of PTH, where as elevations due to primary hyperparathyroidism are around 12 mg/dl serum calcium and PTH levels double the norm. In the previously reported case of parathyroid carcinosarcoma the patient had mildly elevated PTH and calcium levels of 117 ng/ml and 10.8 mg/dl, respectively. The patient in this study had normal PTH and calcium levels. Up to 75% of patients with parathyroid carcinoma will have a palpable neck mass and according to a study out of the Sloan Kettering Institute recurrent cancer is the most significant indicator of the possibility of malignancy, as was the case with the patient in this study.

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References