Medullary Thyroid Carcinoma Metastatic To The Parotid Gland

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Abstract

Educational objective
At the conclusion of this presentation, the participants should be able to recognize the potential for unusual presentations of metastatic disease in patients with medullary thyroid carcinoma.

Objectives
Medullary thyroid carcinoma is an aggressive malignancy of the thyroid gland with a propensity for widespread metastasis both at the time of diagnosis and following treatment. Clinicians should be aware of unusual presentations of this malignancy to ensure timely identification of recurrent disease.

Methods
We present a rare case and review the associated literature. Our patient had a history of familial medullary thyroid cancer previously managed with total thyroidectomy and paratracheal neck dissection. She now presented five years later with a new right parotid mass. Laboratory values showed an elevated calcitonin of 1112 and a CEA of 36. Fine needle aspiration was consistent with metastatic medullary thyroid carcinoma. CT neck confirmed the presence of a right parotid mass but showed no cervical lymphadenopathy. PET, octreotide, and chest CT scans showed no lesions.

Results
The patient was managed surgically with a superficial parotidectomy and right level II-IV selective neck dissection. Final pathology confirmed a diagnosis of metastatic medullary thyroid carcinoma in the parotid gland; no tumor was present in the contents of the right neck dissection. One year from the time of surgery she remained free of clinical disease.

Conclusions
To our knowledge this is the second reported case of medullary carcinoma metastatic to the parotid gland. Knowledge of this malignancy’s potential for unusual and aggressive patterns of spread is essential to ensuring early identification of recurrent disease.

Case Presentation

A 19 year old female presented with a right parotid mass for 2 months. Her past medical history was significant for multifocal medullary thyroid carcinoma diagnosed at age 14 after development of a thyroid nodule. She was subsequently found to be positive for the germline RET protooncogene mutation. She also had a personal and family history of cutaneous lichen amyloidosis, but screening for other disorders associated with multiple endocrine neoplasia syndrome was negative. At the time she was managed with a total thyroidectomy and central paratracheal neck dissection. Following surgery, she was monitored for recurrence with periodic physical examinations and laboratory testing of calcitonin and CEA levels.

On physical exam, the patient had a firm, mobile right facial mass measuring approximately 1.5 centimeters in the region of the parotid gland. Her facial nerve function was normal and she had no other palpable lymphadenopathy.

Laboratory values revealed a significantly elevated calcitonin level of 1112 pg/mL, increased from a value of 35 immediately following her initial surgery. Additionally, CEA level was similarly elevated at 36 ng/mL from 1.4 over the same time period. The patient underwent a fine needle aspiration of the parotid mass, and results were consistent with metastatic medullary thyroid cancer. CT scan of the neck was additionally performed which confirmed the presence of a 1.5 centimeter mass in the right superficial parotid gland, with no other radiographic evidence of pathologic lymphadenopathy. PET, octreotide, and CT chest scans showed no lesions.

The patient was managed surgically with a superficial parotidectomy and right level II-IV selective neck dissection. Final pathology confirmed a diagnosis of metastatic medullary thyroid carcinoma in the parotid gland; no tumor was present in the contents of the right neck dissection. One year from the time of surgery she remained free of clinical disease.

Discussion

Medullary thyroid carcinoma (MTC) accounts for approximately 3-10% of all thyroid cancers and arises from parafollicular c-cells of neural crest origin. It is the most aggressive of the well-differentiated thyroid cancers, with cause-specific mortality at five years reported between 13-32%. Sporadic MTC accounts for approximately two-thirds of these tumors, and the remainder are familial. Familial MTC generally has a more favorable prognosis and is associated with germline mutations of the RET proto-oncogene. It may occur in isolation or as part of multiple endocrine neoplasia syndrome type 2.

Lymph node metastases occur in 25-63% of patients with palpable MTC. Cervical lymph node metastasis most commonly presents in the central neck compartment, and less often in the lateral parajugular nodal levels. Distant metastatic MTC occurs in up to 10-15% of cases and has a predilection for liver, lung, and bone. Recurrent disease is likewise expected to present at the same regional or distant metastatic sites.

To our knowledge this is only the second reported case of MTC metastatic to the parotid gland. The most common metastatic lesions to the parotid gland are cutaneous squamous cell carcinoma and melanoma of the face and scalp, which is consistent with known patterns of lymphatic drainage to the parotid nodal basin. As the lymphatic flow from the thyroid gland is to central, lateral, and caudal neck regions, metastatic spread of primary thyroid cancers is unlikely to occur superior to the mandible. However, rare sites of metastatic spread from MTC to the pituitary gland and skin of the neck, chest, and scalp have been reported. Likewise, uncommon metastases to the parotid gland may occur, including those from breast or Merkel cell carcinoma primary tumors.

Treatment of recurrent MTC relies primarily upon surgical management. Tumors do not concentrate radioactive iodine, and responsiveness to external beam radiation is variable. Patients with clinical evidence of recurrent neck disease by either physical exam or imaging typically undergo resection via formal central (level VI-VII) and lateral neck dissection (level II-V) if not previously done so at initial surgery. For patients having persistently elevated calcitonin but without other clinical evidence of recurrence, options include observation with serial imaging or surgery. Several reports have demonstrated success at reducing or even normalizing calcitonin levels when reoperation is performed in this group of patients, although the overall impact on long-term prognosis remains unclear. Further studies are needed to determine the survival benefit afforded by repeat operation in patients with recurrent MTC.

Conclusions

This represents only the second reported case of medullary thyroid carcinoma metastatic to the parotid gland. Knowledge of this aggressive tumor’s capability for unusual patterns of metastatic spread will aid in timely identification and treatment of residual or recurrent disease.

References