Malignant thyroid teratoma: Case report and review of literature

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INTRODUCTION

Teratomas are defined by the presence of tissue from each of the three primordial germ layers. They are germ cell tumors that are typically encountered in the gonads but can develop along extragonadal sites along the midline. Cervical teratomas more commonly occur in infancy, and are mostly benign in nature. Due to the location near the airway in cervical tumors, possible airway obstruction is a concern in these cases. Adult teratomas are rare and generally are malignant tumors.

METHODS

Case report and literature review

RESULTS

An otherwise healthy 33 year old African-American female was referred for a rapidly enlarging right thyroid mass. It originally was almond-sized but grew significantly over four months. She denied pain, change in weight, dysphagia, dyspnea, fatigue, or hoarseness. She had no family history of thyroid cancer or significant radiation exposure in her past. On physical examination, she had no stridor. She had a large right thyroid nodule that extended to the midline, approximately 8 cm in maximal diameter. The mass was firm, nontender and mobile. She had no palpable lymphadenopathy.

An outside fine needle aspiration was reviewed by pathologists at our institution and revealed a poorly differentiated carcinoma likely of thyroid origin. PET/CT scan demonstrated uptake within the mass in the right thyroid lobe, as well as within two lymph nodes in the right neck. The patient then underwent a total thyroidectomy, central neck dissection, and right neck dissection.

When the pathology for the specimen was reviewed (Figures 1 & 2), immature cartilage components and neuroepithelial elements were noted in the tumor, consistent with teratoma. The teratoma was continuous with the thyroid gland, separated only by a pseudocapsule layer. Two lymph nodes were positive for metastatic teratoma. Greater than four immature elements per each low-power field as well as cellular atypia were found in the teratoma and lymph nodes, meeting criteria for malignancy.

The patient was informed of the results and referred to medical oncology and radiation oncology for further treatment. Her immediate post-operative imaging showed complete removal of gross disease. Unfortunately, she refused adjuvant therapy and developed recurrent bilateral neck masses along with metastatic lesions in the lungs less than 4 months after surgical treatment.

DISCUSSION

Cervical and thyroid teratomas are rarely described, with only 31 cases reported in the literature. Malignant thyroid teratoma is an aggressive disease. It is a rare entity, most commonly described in women of child-bearing age. Typical presentation is a rapidly enlarging neck mass, sometimes associated with dyspnea, stridor or both. Fine-needle aspiration results are often unreliable and diagnosis is made by surgical pathology specimens.

Teratomas are considered to be of thyroid origin if they are (1) present within the gland, (2) the thyroid gland is absent or replaced by tumor or (3) the thyroid gland is continuous with the tumor1. Histology of the tumor correlates with aggressiveness of the disease. Benign teratomas, also known as grade 0, show only mature elements on pathologic review. The increasing number of immature elements and primitive cells correlates with a microscopically malignant teratoma. Grade 1 teratomas have only one immature element per low-power magnification. Grade 2 has 2-3 embryonal-type tissue elements per field. Finally grade 3, which is considered malignant, has more than four immature elements with mitoses and atypia2.

Radiation therapy was first used as an adjuvant treatment in 19533. Since then, radiation has been seen as a palliative effort to reduce symptoms of this grave diagnosis. More recent reports have shown no evidence of disease as long as 10 years after aggressive chemotherapy with cisplatin, bleomycin and etoposide, in regimens based on germ-cell tumors of the gonads4.

A poor prognosis remains for malignant thyroid teratoma despite aggressive multimodality treatment. Death usually occurs due to metastasis to lungs or airway obstruction. While survival times have improved in case reports with chemotherapy and radiation, the mainstay of treatment remains surgical intervention with vigilant follow-up. Given the rarity of the disease, information is limited regarding the appropriate therapeutic algorithm. A summary of all known cases of malignant teratoma of the thyroid is seen in Table 1.

CONCLUSIONS

We present a rare case of a young woman with malignant thyroid teratoma. Malignant thyroid teratoma is a rare disease in adults and has a poor prognosis. Surgery, chemotherapy and radiation have improved survival times. If no intervention is taken, the natural course of the disease results in airway obstruction and metastatic lesions to the lungs.

REFERENCES

5. References for the table above are available upon request.