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Undifferentiated Anaplastic Sarcomatoid Carcinoma of the Thyroid: Is Surgical Resection Worth it? – A Case Report and Review of Literature Tracy Nguyen and Subhasis Misra*

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Introduction

Undifferentiated anaplastic thyroid carcinoma is a rare malignancy that occurs in 2-5% of thyroid carcinoma [1]. However, it can contribute to the largest majority of annual deaths from thyroid carcinoma, occasionally as high as 50% of deaths. Anaplastic thyroid carcinoma with sarcomatoid features is even more rare [2,3]. This occurs when carcinomatous epithelial cells possess a morphology that is similar to sarcoma. The majority of cells resemble spindle and giant cells in most cases. This malignancy is known for its very aggressive nature and poor prognosis in the elderly. Most patients diagnosed with this malignancy die within 2-6 months post operatively. This case discusses the outcome of surgical resection in an elderly patient with anaplastic sarcomatoid carcinoma of the thyroid and explores any benefits to surgery.

Case Presentation

A 75-year-old Caucasian female presented to the clinic with complaints of dysphasia and hoarseness due to a progressively enlarging mass on her anterior neck. Patient has a history of hypothyroidism, which she has been taking 0.1 mg of L-thyroxine daily [4,5]. Patient also presents with a history of breast cancer and melanoma of the lower limb and hip. On examination, her left thyroid was firm and round approximating $3.8 \times 2.9 \times 3.3$ cm. A hypoechoic focus was seen on ultrasound measuring $0.8 \times 0.7 \times 0.9$ cm. A palpable left supraclavicular node was felt. Trachea deviation to the right was also noticeable. An ultrasound guided fine needle aspiration biopsy was performed using a 16/18-G needle. After great difficulty due to the solid nature of the mass, a small sample was obtained. Pathology report returned with suspicion of a sarcoma of the thyroid [6,7]. Magnetic resonance imaging of the neck confirmed the left thyroid mass, in addition also highlighted the close proximity of the mass to the left carotid artery. Patient underwent a total thyroidectomy with limited neck dissection with intra operative nerve monitoring. An excision of the left supraclavicular mass and removal of the tumor was performed. The surgery was complicated by the intricate growth of the tumor 180 degree around the left carotid artery.

Discussion

Undifferentiated anaplastic sarcomatoid carcinoma of the thyroid is an extremely rare disease with only a few thousand cases to date. Originally, fine needle biopsy suggested that our patient presented with a sarcoma of the thyroid. However, after resection of our large, hemorrhagic tumor, final pathology determined that our patient presented with undifferentiated anaplastic sarcomatoid carcinoma of the thyroid.

Undifferentiated anaplastic sarcomatoid carcinoma of the thyroid is also an extremely rare malignancy that occurs in less than 2% of all thyroid carcinoma. Undifferentiated anaplastic sarcomatoid carcinoma of the thyroid is known for its aggressive nature [8,9]. Generally affecting the elderly population, undifferentiated anaplastic sarcomatoid carcinoma of the thyroid is relentless and most patients survive only a few months after being diagnosed.

Diagnosis is usually confirmed after visualization of anaplastic spindle cells with atypical cytology and high mitotic activity; as well as immunohistochemical stains showing positivity for the following: vimentin, PAX-8, pan-keratin, and p63. Some anaplastic thyroid carcinoma may show positivity for epithelial membrane antigen and thyroid transcription factor-1. Those tumors that express sarcomatoid features such as our patient, may not express TTF1. Instead, these patients tend to express pan-keratin in most of the cases.

In our case, pathology was confirmed by Emory University to be an undifferentiated anaplastic sarcomatoid carcinoma of the thyroid. Final pathology showed sheets of atypical spindle cells that entrapped nerves. The cells were arranged in an ill-defined fascicular pattern. Cytology showed marked atypia with no defined cell boundaries. Mitotic activity however was not brisk. Immunostains showed expression of pan-keratin and p63 suggestive of undifferentiated anaplastic thyroid carcinoma. In addition, vimentin and CD68 were strongly positive throughout the entire tumor.

Despite the efforts to remove the tumor, some microscopic tumor still remained. Post-operation radiation was advised in addition to chemotherapy. Overall, the leading cause of death in patients is usually due to suffocation. Our patient experienced a severe decline in respiration, which required a tracheostomy. Tracheostomy was performed post operation day 2 due to the development of stridor.

Conclusion

In conclusion, our case outlines the complicated nature of undifferentiated anaplastic sarcomatoid carcinoma of the thyroid and the typical route of treatment most patient experience. The ideal treatment for patients with undifferentiated anaplastic sarcomatoid carcinoma of the thyroid depends on the size of the mass and the stage of the patient. In addition, the amount of sarcomatoid feature of the tumor may play an important role as well, as it can cause future complications and advancement of the tumor. In some cases, if symptomatic relief is not necessary, shrinkage of the tumor with radiation prior to surgery may pose a more optimal outcome for patients. As with our case, surgical resection for symptomatic relief did not increase the survival rate of our patient.

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Received March 19, 2015; Accepted June 09, 2015; Published June 19, 2015

Citation: Nguyen T, Misra S (2015) Undifferentiated Anaplastic Sarcomatoid Carcinoma of the Thyroid: Is Surgical Resection Worth it? – A Case Report and Review of Literature. Endocrinol Metab Synd 4: 179. doi:10.4172/2161-1017.1000179

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