Papillary thyroid carcinoma coexists with undifferentiated carcinoma & A case report and review of literature

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Abstract

Coexistence of Papillary Thyroid Carcinoma (PTC) with Undifferentiated Thyroid Carcinoma (UTC) is extremely rare. The etiopathogenesis has not been elucidated though it may be associated with de differentiation. We report a case of an 89 year-old woman with a painless mass of the right neck with 5 months. The patient underwent a lobectomy. The histology showed that the tumor was composed of epithelioid spindle cells with hyperchromatic, karyokinesis obviously and abundant blood sinus. Immunohistochemical studies demonstrated the tumor cells to be Vimentin (+), P53 (+), CK19 (+), CK516 (+), TTF-1 (-), TG (-), CD34 (+) and Ki-67 (+20 %). The patient still alive after 6 months. It is a challenging diagnosis to make due to lack of the recognition. Before making this diagnosis, it is important to strengthen awareness of the disease, especially for the elderly. And this case may be provided further evidence and images of this dedifferentiation.

Introduction

Papillary thyroid carcinoma (PTC) is the commonest thyroid carcinoma worldwide [1], while undifferentiated thyroid carcinoma (UTC), a term we will use as a synonym for anaplastic thyroid carcinoma (ATC), ranks among the most lethal of all human malignancies, and represents nowadays less than 3% of all clinically recognized malignant thyroid neoplasms [2]. PTC, as differentiated thyroid carcinoma (DTC), has good prognosis but 5% of the patients already have distant metastasis at the diagnosis [3]. The incidence of PTC was about 83.7% in thyroid malignancies and increased 5.2% per year, according to the epidemiologic data by Cavalheiro BG et al. [4] And UTC is a rare and extremely aggressive malignancy, with the median survival of less than 6 months due to rapid progression and resistance to multimodal therapies [5]. However, coexistence of PTC with UTC is extremely rare. To the best of our knowledge, very little is known about this condition and no authoritative prognostic data exist. The present study reports the case of an 89 year old female patient diagnosed as coexistence of PTC with UTC and the relevant literature is comprehensively reviewed.

Case report

We present a case of an 89 year old female with a rapidly growing and painless neck mass over a period of five months. No evidence of dyspnea or dysphagia was identified. Her past medical history includes a multinodular goiter diagnosed two years ago with no follow-up. Palpation revealed a large, irregular, firm, tender and painless tumor mass in the right anterior neck. She was clinically euthyroid with normal plasma level of stimulating hormones. Ultrasonography of the thyroid identified a 7.1 x 5.3 cm, ill defined hypoechoic mass in the right thyroid lobe. CT revealed an ill defined, low density mass in the right thyroid lobe that was adjacent to carotid artery. A total body scan did not show any other mass. The patient underwent a surgical exploration. There was a hard, intact capsule tumor of the whole right lobe of the thyroid gland, which was attached to the right carotid sheath. Subsequently, a right thyroid lobectomy was performed. The resected lobe measured 5.5 cm in the largest dimension; No lymph node metastasis was identified.

The resected specimen (4-μm) was fixed in 10% buffered formalin (Sigma Aldrich, St. Louis, MO, USA), processed and embedded in paraffin (Leica, Mannheim, Germany) using standard histological methods. Staining was visualized using an inverted microscope (TE2000-U; Nikon Corporation, Tokyo, Japan). Hematoxylin and eosin (Sigma Aldrich) staining revealed that epithelioid spindle cells with hyperchromatic, karyokinesis obviously and abundant blood sinus were seen in the tumor [Figure 1]. Immunohistochemistry was performed. The monoclonal mouse anti-human vimentin (clone, V9; cat. no. AX074-YCD; dilution, 1:200; BioGenex, USA) was used for vimentin staining. The monoclonal mouse anti-human Ki 67 antigen (clone, MIB-1; cat. no. IR626; dilution, 1:200; Dako) was used for Ki 67 staining. The monoclonal mouse anti human cytokeratin (clone, AE1/AE3; cat. no. IR620; dilution, 1:100; Dako) was used for cytokeratin staining. Immunohistochemical staining was positive for CK19, CK516, p53, vimentin and Ki 67 (20%), and negative for TTF-1, TG and CD34 [Figure 2]. The final diagnosis was papillary thyroid carcinoma coexisting with undifferentiated thyroid carcinoma.
Combining surgery, chemotherapy and radiation therapy, might achieve better results in improving survival in some patients; however, UTC has a very low cure rate even with the extremely radical treatments.

In our case, the diagnosis of PTC coexisting with UTC was made on the pathological, and immunohistochemical features of the tumors, which were similar to those found in the literature [11]. Despite surgical excision, most patients die after 3 to 6 months. As to UTC, the rarity of this malignancy and the rapidity by which it grows has been a major barrier to progress in finding effective therapies. Thus, the treatment that is the current standard of care for these patients is largely palliative, and few are cured. A recent study by Antonelli A et al. [12] shows that the CLM3 (a pyrazolo[3,4-d]pyrimidine compound), as an agent with antitumor and antiangiogenic activity, is very promising in the treatment of UTC, opening the way to a future clinical evaluation. The present case study has been reported, in which the patient was still alive with no evidence of disease, with a follow-up of 6 months. As for the etiology, we agree with Eloy C et al and Evans WD [6,11]. And this case may be provided further evidence and images of this dedifferentiation.

**Conclusion**

In summary, PTC coexisting with UTC of the thyroid is extremely rare. It is a challenging diagnosis to make due to lack of the recognition. Before making this diagnosis, it is important to strengthen awareness of the disease, especially for the elderly. And this case may be provided further evidence and images of this dedifferentiation.

**References**


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