CASE REPORT

Carcinosarcoma of the thyroid: a case report

Naqiyah I¹, Zulkarnaen A N², Rohaizak M¹, Das S³

¹Department of Surgery, ²Department of Pathology, ³Department of Anatomy Faculty of Medicine, Universiti Kebangsaan Malaysia, Jalan Yaakub Latiff, Bandar Tun Razak, 56000, Kuala Lumpur, Malaysia

Abstract

Thyroid carcinosarcoma (TC) is a very rare, aggressive thyroid malignancy with a clinical course similar to anaplastic carcinoma. A thorough search of the literature reveals limited information regarding its behavior and treatment modalities. TC has a high mortality rate despite the multi-modal approach. A 54-year-old Chinese gentleman with a long history of a nodule in the right lobe of the thyroid presented with recent history of hemoptysis and shortness of breath. Cytology was suggestive of follicular neoplasm. However, histopathological examination of the total thyroidectomy specimen confirmed TC. This case reports discusses the clinical course and management of TC, which can cause a diagnostic dilemma. Hippokratia 2010; 14 (2): 141-142

Key words: thyroid, carcinosarcoma, undifferentiated, follicular, cancer.

Corresponding author: Rohaizak Muhammad, Department of Surgery, Faculty of Medicine, Universiti Kebangsaan Malaysia, Jalan Yaakub Latiff, Bandar Tun Razak, 56000, Kuala Lumpur, Malaysia, tel: 603-91702315, fax: 603-91737831, e-mail: rohaizak@hotmail.com

Thyroid carcinosarcoma (TC) is a rare entity as it comprises less than 0.1 percent of all thyroid malignancies. While other thyroid carcinomas arise from the epithelial component of the gland, TC originates from both the epithelial and stromal components. It is typically observed in old-aged females and carries grave prognosis, similar to the anaplastic thyroid carcinoma.

Case report

Prior ethical consent was taken for the case. A 59-year-old man, retired carpenter, was first seen with symptoms of cough and hemoptysis over a span of 3 months. This was associated with dyspnea at rest. The patient also claimed to have a slow growing anterior neck swelling over 30 years, which did not cause any troublesome symptoms. He was a chronic cigarette smoker but never had respiratory ailments before. He denied having previous neck irradiation. His family members were free from any type of malignancy.

Local examination of the neck revealed a firm, well-defined right thyroid nodule measuring 3 x 4 cm. There was no cervical lymphadenopathy. Bilateral crepitation was heard over his lungs. Serum investigations that included full blood count, electrolytes and thyroid function tests were normal. Ultrasound of the neck revealed a nodule with characteristics that made it suspicious for malignancy. A subsequent thyroid scan detected a 'cold' nodular lesion. Multiple "cannon ball" lesions were observed in the chest radiograph, suggestive of metastases. A computed tomography of the abdomen did not demonstrate any other lesions. The fine needle aspiration cytology (FNAC) of the thyroid nodule showed a follicular lesion. A clinical diagnosis of follicular thyroid carcinoma

was set and the patient underwent total thyroidectomy. At surgery, there was extensive adhesion surrounding the nodule but the trachea and the major vessels were free from invasion. Cervical lymph node enlargement was not encountered. The patient had an uneventful post-operative recovery.

The histopathological examination of the thyroid gland revealed a 3.5 x 2.5 cm tumor, with two groups of cellular populations. One comprised of large spindle cells with pleomorphic hyperchromatic nuclei containing priminant nucleoli and eosinophilic cytoplasms, while the other comprised of carcinomatous cells and exhibited pleomorphic vesicular nuclei with occasional prominent nucleoli and clear cytoplasm. Histochemical staining showed that the spindle cells were strongly positive for vimentin and negative for calcitonin or neuroendocrine markers while the carcinomatous cells were positive for thyroglobulin (Tg). Hence, a diagnosis of carcinosarcoma was made. Extrathyroidal extension was also observed.

The patient underwent adjuvant radioactive iodine-131 (RAI) therapy. Positive uptake was observed. He remained asymptomatic until seven months later when he presented with local recurrence over the right and posterior aspect of his neck. A palliative excision of the lesion was performed due to its ulcerative nature. The histological report was similar to the primary tumor. The patient succumbed to the disease one month later.

Discussion

Undifferentiated thyroid malignancy consists of 2% of all thyroid malignancies. Unlike the well-differentiated thyroid carcinoma, which have excellent prognosis, these undifferentiated tumors has poor survival rate of

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less than 6 months. In an earlier study involving 422 patients with thyroid malignancies, 91 cases (21.5%) were undifferentiated and only 1 case (0.2%) was diagnosed as carcinosarcoma¹. TC is more common in females over 50 years of age. Its typical physical history is that of a rapidly enlarging thyroid nodule that may obstruct breathing. However, there are reports of TC arising in pre-existing multi-nodular goiters by means of neoplastic metaplasia². Metastases to cervical lymph nodes are also common.

A previous study reported that exposure to phosphorus, RAI and carmustine therapy may be contributing factors for the development of TC². Pre-operative diagnosis of TC by FNAC is difficult. It is usually diagnosed by histopathological examination when both entities of malignant mesenchymal and epithelial components are discovered. This is confirmed with immunohistochemistry staining of the carcinomatous cells for Tg while the mesenchymal cells are strongly positive for vimentin and S100 protein³⁻⁵.

As in all thyroid malignancies, a total thyroidectomy and complete removal of the tumors is usually recommended for TC whenever feasible. This is performed along with complete adjacent lymph nodes excision². Local recurrence with invasion to the surrounding structures is common and this is said to be associated with the aggressiveness of the tumor ²⁻⁵. Complicated airway obstruction and metastatic lesions in the lungs usually follow the recurrence and most patient succumb to this.

Due to its rarity, the adjuvant treatment for TC is not well-studied. External beam irradiation and chemotherapy had been recommended but these modalities do not seem to arrest the disease progression or improve survival. A previous study reported a series of different modality approaches to treat anaplastic thyroid carcinoma, another poorly differentiated thyroid tumour. These modalities included biopsy alone, biopsy and chemoradiation, debulking procedure, debulking with chemoradiation and complete excision with chemoradiation. There was no significant difference in the outcome and survival rate among the different treatments.

External beam radiotherapy has beneficial effect in locally advanced follicular thyroid carcinoma. In a pre-

vious study, high-dose external beam radiotherapy (60 Gy) has been proven to be effective for regional control against aggressive tumours? However, similar response is yet to be proven for TC. The benefit of RAI in well-differentiated thyroid cancers is well-established, as it results in significant survival prolongation and good regional control⁸. It may be sensible to use RAI in TC with positive uptake but to the best of our knowledge, there is paucity of literature regarding this. In addition, extensive pulmonary metastases, as demonstrated in this case, may deem RAI unsuccessful due to the large tumor burden.

Conclusion

TC is rare and the treatment of this aggressive disease still remains unclear. Surgical removal of the primary thyroid tumor remains the only method of management despite early advance of metastatic disease.

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