Anaplastic thyroid carcinoma: a case of a young women that reports a survival exceeding 6 years

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Abstract. – Anaplastic thyroid carcinoma (ATC) is an uncommon and highly aggressive malignancy. Differentiated follicular and papillary thyroid carcinomas account for most (80-90%) thyroid malignancies with ATC accounting for less than 5%. A diagnosis of ATC is usually fatal with a mean survival of 3-9 months and only 10-15% alive at 2 years. Histologic examination reveals that many of them contain a papillary structure or follicular components in focal areas and genetic alteration is the driving force for genesis of cancer and progression. These studies showing that ATC represents a terminal “de-differentiation” of pre-existing differentiated carcinoma.

Most of patients are incurable, nevertheless a multimodality approach, incorporating surgery with the aim to obtain complete macroscopic resection and achieve clear resections margins followed by adjuvant treatment RT (radiotherapy) and/or chemotherapy, improve local control and extend the survival.

The aim of the present study has been to review a case of a young women with ATC that reports a survival exceeding 6 years.

Key Words:
Anaplastic thyroid carcinoma, ATC, Thyroid cancer, Survival.

Introduction

Anaplastic thyroid carcinoma (ATC) is an uncommon and highly aggressive malignancy. Differentiated follicular and papillary thyroid carcinomas account for most (80-90%) thyroid malignancies with ATC accounting for less than 5%. A diagnosis of ATC is usually fatal with a mean survival of 3-9 months and only 10-15% alive at 2 years.

ATC is often demonstrated in patients who have longstanding goiters or incompletely treated papillary or follicular thyroid cancers.

Histologic examination reveals that many of them contain a papillary structure or follicular components in focal areas and genetic alteration is the driving force for genesis of cancer and progression. A recent important genetic finding is the oncogenic T1799A transversion mutation of BRAF (the gene for the B-type Raf Kinase) occurring exclusively in papillary thyroid cancers and PTC-derived anaplastic thyroid cancer and is a specific diagnostic marker for this cancer when identified in cytological and histological specimens. Also tumor suppressor gene p53 is rarely mutated in well differentiated papillary and follicular thyroid carcinomas (PTC and FTC) but is mutated in many poorly differentiated PTC and frequently mutated in ATC.

These studies showing that ATC represents a terminal “de-differentiation” of pre-existing differentiated carcinoma.

Anaplastic thyroid carcinoma present annual incidence of 1-2 cases/million is more frequently in older age with a higher prevalence in women and is marked by rapid growth and extensive local invasion.

The diagnosis is usually confirmed by fine-needle aspiration (FNA) biopsy; however, if result of FNA is uncertain, the patient should underwent to incisional biopsy. Morphologic imaging techniques such as ultrasonography or computerized tomography (CT) have a role in the evaluation of ATC.

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therapy) and/or chemotherapy, improve local control and extend the survival.

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**Case Report**

A 29-year old women come to our observation in october 2000 because of a rapidly enlarging anterior neck mass at the last 4 months. Patient didn’t mention about previous irradiations on the neck either familiarity with thyroid and neoplastic diseases.

In the hospital the patient was submitted to hematocellular and instrumental checks. The ultrasonographic evaluation of the thyroid revealed on the right lobe the presence of one solid formation (40 × 18 × 24 mm), hypodense, with irregular edges with intra- and extraleSIONal vascularization; the left lobe revealed a regular aspect. Ecography revealed also some enlarged lymph nodes at the right sides (max 8 mm).

A thyroid fine-needle aspiration biopsy (FNAB) revealed a supposed medullary carcinoma indicating the difficult of a differential diagnosis with other tumours (lymphoma, hemangioblastoma) which anaplastic carcinoma has been erroneously identified.

There was a clear need for surgical removal of the thyroid; at this case it was possible a total thyroidectomy without lymphadenectomy for the normal state of regional lymph nodes also demonstrated in course of the intra-operating exploration: the tumor included the right total lobe with adesivity to the anterior muscular plane from which the neoplasm was easily dissected.

The patient undergone a surgical treatment; the macroscopic features showed one nodule neoplastic on the right lobe almost 3 cm in diameter, capsulate without local infiltration of adjacent anatomic structures. Histological examination showed ATC with atipical, multinucleated giant cells and with necrosis and hemorrhage areas (Figure 1). Immuno-histological analysis demonstrated an intense positivity for vimentin in the neoplastic cells and negative for CD30, LCA, CD3, CD79a, S-100, HMB45, CK, Thyroglobulin, Calcitonin, Cromogranin.

The surgical treatment of the patient is followed by radiotherapy (5,000 cGy) and chemotherapy (adriamycin 10 mg/m² I.V./week for 7 doses) and the patients of this case reported a complete response and survival exceeding 6 years.

The patient is submitted to a post-treatment control (total-body CT and bone scintigraphy) reporting that the most efficacious curative treatment is the multimodal (surgery, radio- and chemotherapy) and a better prognosis is observable in intraglandular forms, small neoplastic focus and young patients.

**Discussion**

Anaplastic carcinoma of the thyroid (ATC) is an aggressive form of cancer of the thyroid gland; it represents less than 2% of all thyroid cancers, and in contrast to well-differentiated thyroid carcinoma has a worst prognosis, with average rate of survival after diagnosis from 4 to 12 months.

Clinically, most patients have a dominant mass of 5 cm or more, multiple nodules in both thyroid lobes and many enlarged lymph nodes; also is often present voice changes or dysphagia. Unusual symptoms included symptomatic bradycardia from compression of the vagus nerve and superior vena cava syndrome.

The major risk factor is an history of previous benign or malignant thyroid disease. Long-standing goiters or benign nodules should be followed carefully and considered for resection if they grow or do not respond to medical ther-
apy, and total thyroidectomy for malignant disease may avoid the development of anaplastic carcinoma.

Anaplastic thyroid carcinoma is a locally and systemically aggressive disease, with long-term survival seen only in those with well-localized anaplastic tumor and young patients.

In conclusion, for the aggressivity of this neoplasm, many patients with ATC will be treated palliatively. However, surgery with the aim to achieve complete resection followed by adjuvant RT/CT (the most efficacious multimodal treatment), might improve local control and extend median survival.

References


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