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Sarcomatoid Anaplastic Carcinoma of the Thyroid: A Case Report

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ABSTRACT

Describe the modalities for the management of anaplastic thyroid carcinoma in its sarcomatoid form. We report the case of a 45-year-old woman who presented with anaplastic thyroid carcinoma in its sarcomatoid form. This is a 45-year-old patient admitted to the emergency department for the management of inspiratory dyspnea on a large mass in the anterior cervical region that has been developing for more than 3 months. A cervicothoracic CT scan was performed urgently showing an enormous thyroid mass of malignant appearance at the expense of the left thyroid lobe with extension to the other lobe, associated with thrombosis of the internal jugular vein and the superior vena cava, as well as a right pulmonary embolism. A surgical decision for tumor reduction with extemporaneous pathological examination associated with a tracheostomy was taken. The definitive anatomopathological examination revealed after immunohistochemical study anaplastic thyroid carcinoma in its sarcomatoid form. The therapeutic approach to anaplastic cancers has changed in a few decades. Surgical treatment of goiters has reduced the incidence of these cancers, access to preoperative cytological diagnosis sometimes allows early management, at the stage of emerging intrathyroidal anaplastic foci in differentiated cancers. Combined treatments give back a place to surgery.

Introduction

Anaplastic thyroid cancers represent the final form of the dedifferentiation of follicular-strain thyroid tumors and are one of the most serious cancers in humans.

They represent less than 2% of all thyroid cancers and their annual incidence is of the order of two per million inhabitants. They are largely predominant in subjects generally between the sixth and eighth decade.

The usual clinical presentation is a rapidly evolving, invading, and compressive mass of the neck with lymphadenopathy and metastases present from the outset once in three. Although the cytological study may suggest the diagnosis of anaplastic thyroid cancer, histological, and even immunohistochemical confirmation is necessary to rule out lymphoma and validate aggressive treatment.

Patients with anaplastic thyroid cancer must be admitted urgently to a specialized department (oncology, otorhinolaryngology, and endocrinology) where treatment protocols adapted to their general condition can be discussed and carried out quickly. Combination treatments can slow the progression of the cervical disease through the combined action of radiotherapy, cytotoxic radiosensitizers, and excisional surgery if possible.

However, they are difficult to use in elderly patients and do not prevent fatal progression to metastatic disease. The prognosis for anaplastic cancers is poor: the average survival of patients varies from four to nine months after diagnosis. Prolonged survival of several years only concerns patients who had been operated on early for a localized tumor who had received adjuvant chemotherapy or radiotherapy treatment.

Therapeutic research is exploring methods of tumor redifferentiation and targeted therapies that specifically block the EGF receptor or inhibit neoplastic angiogenesis. Prevention of this fatal disease consists of the appropriate management of differentiated thyroid cancers and goiters in elderly patients.

Case Report

This is a 45-year-old patient, with no notable pathological history, admitted to the emergency department for the management of inspiratory dyspnea on a large mass in the anterior cervical region that has been developing for more than 3 months, without the concept of dysphagia, dysphonia, or dysthyroidism. The evolution is marked by a deterioration of the general condition with anorexia and weight loss estimated at 17 kilograms in 3 months.

The clinical examination found a conscious patient with inspiratory dyspnea without any notion of signs of respiratory struggle, the cervical examination found a large mass in the anterior cervical region greater on the left than on the right (approximately 10 cm).

A cervicothoracic CT scan was performed urgently showing an enormous thyroid mass of malignant appearance at the expense of the left thyroid lobe with extension to the

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other lobe, associated with thrombosis of the internal jugular vein and the superior vena cava, as well as 'a right pulmonary embolism (trunk of the right pulmonary artery and the ipsilateral lower lobe branch).

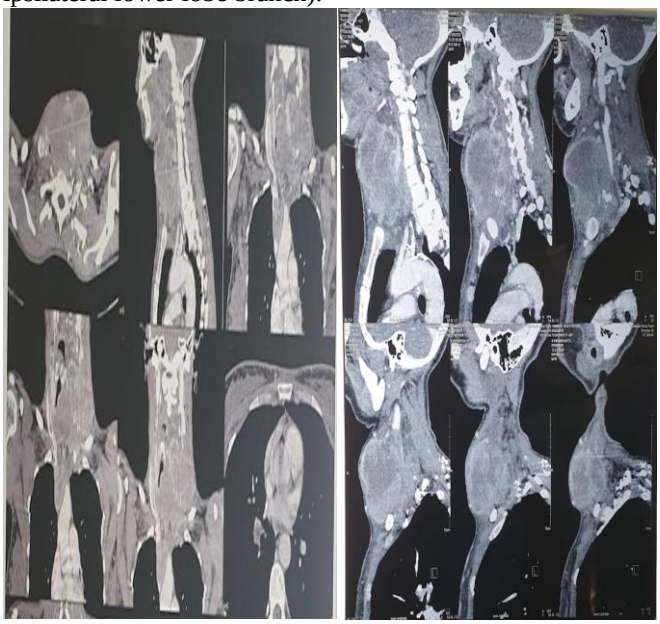

Figure 1. Images of a cervicothoracic CT scan with an injection of contrast product in axial coronal and sagittal slice showing a cervical mass depending on the left thyroid compartment with homolateral internal jugular vein thrombosis and right pulmonary embolism

Nasofibroscopy was performed showing reduced mobility in the two vocal cords more important to the left.

Biologically, the patient presented microcytic hypochromic anemia at 10g / dl of hemoglobin, the thyroid workup was normal, and the CRP elevated to 119. The patient was started on Lovenox 0.6 twice a day at a curative dose.

A surgical decision for tumor reduction with extemporaneous pathological examination associated with a tracheostomy was taken. An exposure Kocher-type cervical surgical incision was chosen, and a huge whitish mass was found deviating from the aerodigestive axis encompassing the trachea and having an uncontrollable inferior extension. The extemporaneous pathological examination is inconclusive.

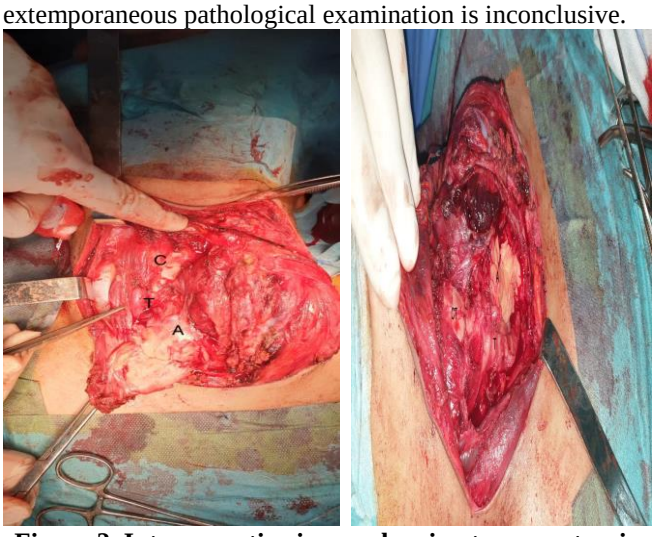

Figure 2. Intraoperative image showing tumor extension on aerodigestive. A: tumor, C: cricoid cartilage, CT: thyroid cartilage, T: trachea.

The immediate post-op was marked by the installation of a cervical hematoma which was evacuated. The overlaps with Anti Vitamin K were administered on the fifth postoperative day under INR control. A post-operative antibiotic treatment based on protected amoxicillin at a rate of 1 gram every 8 hours for 10 days is prescribed.

The definitive anatomopathological examination revealed after immunohistochemical study anaplastic thyroid carcinoma in its sarcomatoid form (using anti Vimentin antibodies (positive), anti Actin antibodies (positive), anti CD31 antibodies (negative), CD34 (negative), Calcitonin (negative), TTF 1 (negative) , AE1 / AE3 (negative), EMA (negative), PS100 (negative), desmin (negative), anti PAX8 antibody (negative), ERG (negative), Thyroglobulin (negative), and complement: H-caldesmon (weak positive), P63 (positive)). The patient was referred to the radiotherapy department and then lost to follow-up.

Discussion

Macroscopically, anaplastic carcinoma is infiltrating, invasive, rearranged by hemorrhagic and necrotic areas.

Histologically, the tumor is formed by the proliferation of spindle-shaped, polygonal, or giant cells. The presence of squamous cells, osteoclastic, or sarcomatous in appearance is possible.

In immunohistochemistry, the tumor no longer expresses thyroglobulin or other markers of thyroid differentiation (NIS, RTSH, TTF1). On the other hand, keratin is frequently present, like the p53 protein or the proliferation markers: Ki67, proliferative cell nuclear antigen (PCNA) [21].

The presence of a differentiated cellular contingent is usual in anaplastic cancers, which argues in favor of the transformation of differentiated cancer. Conversely, the diagnosis of anaplastic cancer should be made as soon as a contingent of undifferentiated cells is present in a papillary or vesicular tumor. Particular attention should be taken concerning undifferentiated small cell cancers which may correspond to lymphomas, spinal cord epitheliomas, poorly differentiated vesicular cancers, or intrathyroidal metastases [2].

The place of surgical treatment is controversial: essential when the ablation of the tumor can be complete in a patient capable of receiving upstream or downstream treatment by chemoradiotherapy [25,35,36], unnecessary and harmful in the event of cervical mass infiltrating the endotracheal axis in an elderly subject in general condition already compromised [5,37].

In localized forms of anaplastic cancer, surgery aims to remove the macroscopic tumor to increase the effectiveness of radiotherapy to preserve noble structures. However, the resection of invaded muscle flaps or the sacrifice of nerves already infiltrated by the tumor may be necessary. Thus, when the surgical treatment is complete, supervised by other therapeutic modalities [33], patient survival can be prolonged [25,36]. Surgery does not dispense with additional treatment by radiochemotherapy, because thyroidectomy alone, even almost total, does not modify the prognosis of patients [14]. Likewise, small anaplastic cancers discovered incidentally and whose surgical treatment appears complete at first glance may present with local relapses or a metastatic course [35,38].

In the event of a locally invasive tumor deemed from the outset inextirpable, first chemoradiotherapy could make it possible to select the patients in whom a surgical treatment carried out in a second phase may be beneficial. The theoretical advantage is to limit very early the development of metastases by a systematic treatment, to increase the

possibilities of complete resection if an objective tumor response is obtained [17,26].

For some authors, even at the stage of metastatic disease, tumor reduction surgery associated with radiochemotherapy may be offered in some patients for palliative purposes, to avoid death from asphyxia [33]. The chemotherapy methods and the irradiation delivered will then be adapted to the age and general condition of the patient to limit toxicities.

A temporary or permanent tracheostomy, the installation of an esophageal stent, or even nutrition by gastrostomy are sometimes essential in the event of major compression of the upper aerodigestive tract. Before starting external radiotherapy, in particular, the observation of a precritical reduction in the size of the laryngeal system and the worsening of swallowing disorders, lead to anticipate these procedures to anticipate post-radiotherapy complications and improve the comfort of the life of the patient.

Conclusion

The therapeutic approach to anaplastic cancers has changed in a few decades. Surgical treatment of goiters has reduced the incidence of these cancers, access to preoperative cytological diagnosis sometimes allows early management, at the stage of emerging intrathyroidal anaplastic foci in differentiated cancers. Combined treatments give back a place to surgery.

Radiotherapy may provide a benefit preoperatively with an intensified dose schedule. Paclitaxel is an attractive alternative to doxorubicin-based multidrug therapy. The prognosis of these cancers, especially in the event of metastases, remains however bleak.

Only patients treated with localized disease can hope for better survival at the cost of heavy treatment. This is why the importance of rapid therapeutic implementation in a specialized setting must be emphasized.

Much is hoped for from combretastatin A4 derivatives, tyrosine kinase inhibitors, and farnesyl transferase inhibitors which have shown promising results at the experimental stage and whose place and mode of use will remain to be defined compared to conventional therapies.

The best approach to anaplastic cancers is preventive, consisting early in the thyroidectomy of goiters as soon as they are symptomatic, progressive, with clinically, sonographically, or cytologically suspect nodules.

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