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THYROID CARCINOSARCOMA: A CASE REPORT

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ABSTRACT Thyroid carcinosarcoma is a rare and aggressive neoplasm of the thyroid gland, with a recurrent character and a bad prognosis, despite the introduction of a multimodal treatment that is not yet codified. We present a case of thyroid carcinosarcoma, number 32 described in the literature, in a 64-year-old man complicating a multinodular goiter that had been evolving for 10 years and which relapsed 6 months after total thyroidectomy. On the anatomopathological level, thyroid sarcoma was mentioned before the microscopic aspect of the tumor and confirmed by the immune histochemical study. The recurrence was massive requiring the use of tumor reduction surgery that allowed the avoidance of a tracheotomy and a good locoregional control of the tumor. And yet the patient died of lung metastases 4 months later.

KEYWORDS : Thyroid, Carcinosarcoma, cancer, undifferentiated,

INTRODUCTION

Differentiated thyroid cancers are the most common cancers of the thyroid gland; they are dominated by papillary carcinoma, which accounts for 80% of all thyroid cancers [1]. However Epithelial and non-pepithelial thyroid carcinomas such as carcinosarcomas are rare and clinically aggressive tumours with poor prognosis. Surgery is the standard treatment for these carcinomas [2]. The exact pathogenesis of thyroid carcinosarcoma is still unknown.

Histologically, carcinosarcomas have an independent malignant double component: carcinoma and sarcoma [3]. It is generally seen in eldery women. It is known for its early metastatic power and local aggressiveness [2].To date, thirty-one cases of thyroid carcinosarcoma have been reported, and this is number thirty-two [1].

OBSERVATION

This is a 64-year-old male patient living in a known mountainous area endemic to goiter, with no particular pathological history including no cervical irradiation in childhood or similar cases in the family. This patient was referred to the ENT consultation after a rapid and recent increase in the volume of a goiter that had been evolving for 10 years, and which was responsible for a compressive symptomatology made of dysphagia and respiratory discomfort without associated dysphonia. Furthermore, no clinical signs of dysthyroidism were noted.

An initial cervical ultrasound already performed (June 2018) showed a nodule occupying almost the entire right thyroid lobe, roughly oval, with blurred boundaries in places (at loboisthmic extension level), moderately hypoechogenic heterogeneous with presence of multiple micro calcifications responsible for posterior attenuation, measuring 5.6 * 3 centimetres.

Thus allowing the classification of this nodule in EU-TIRADS V. On the other hand, the left thyroid lobe was occupied by multiple infracentimetric nodules classified EU-TIRADS II. No cervical

lymphadenopathy was noted. The patient had a total thyroidectomy 7 months after his first ultrasound.

The anatomopathological study of the suspected nodule showed macroscopically a whitish and microcystic nodule, located close to the thyroid capsule and taking the entire lobe with the presence of hemorrhagic reshaping (Figure 1)

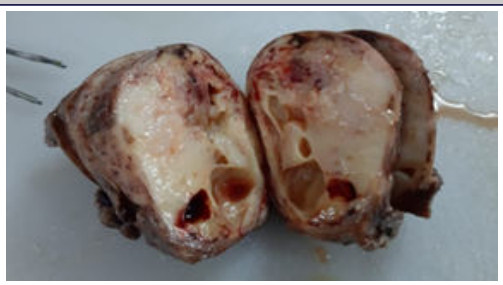

Figure 1: ***macroscopic appearance of tumor thyroid nodule.***

Whereas on the microscopic plane, the examination of the nodule spotted shows a very dense tumor proliferation with double component; a made of fusiform and ovoid cells showing marked atypia and mitoses with a rather clear osteogenesis made of osteoid spans often immature, while the second shows cartilaginous differentiation with atypical cells often elongated to hyperchromatic nuclei. At the periphery, proliferation penetrates the adjacent thyroid parenchyma in places and the two components are intertwined with thyroid vesicles often of small size. Moreover, no vascular embolus was noted. The morphological aspect was in favor of a sarcomatoid malignant tumor process with osteoid and cartilaginous differentiation whose immunohistochemical study is compatible with thyroid carcinosarcoma associating sarcomatoid carcinoma with neuroendocrine differentiation (calcitonin -, thyroglobulin +, EMA +, chromogranine + and synaptophysine +) and osteosarcoma. There was a lack of Pancytokeratin (clone AE1/AE3) marking of tumour cells, with a focal PS100 marking of the osteoecartileginous zones, while Ki67 marking is estimated at 50 (Figure 2).

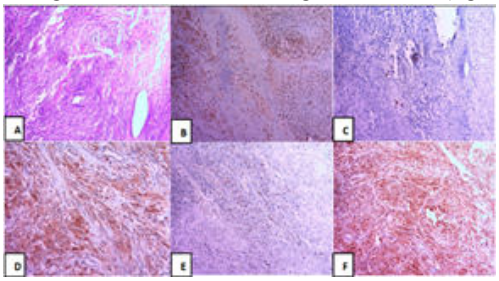

Figure 2: Anatomopathological and immunohistochemical study of the thyroid nodule

A: proliferation of fusiform cells with cytonuclear atypia: mean haetinine eosin (HE) coloration magnification B: thyroglobulin: diffuse positivity of tumour cells with anti-thyroglobulin antibody with cytoplasmic and membrane marking C: focal positivity of tumour cells with anti-cytokeratin antibody D: diffuse positivity of tumour cells with anti-cytokeratin antibodysynaptophysin with cytoplasmic marking E: KI 67: increased in tumour proliferation with nuclear marking indicating aggressive tumour proliferation F: Embryonic membrane antigen (EMA): diffuse proliferation of tumour cells with anti-EMA antibodies with membrane marking.

After having retained the diagnosis of thyroid carcinosarcoma, the patient was referred to the university hospital for further management. The patient was lost of sight and did not consult until after six months of surgery, and it was on the occasion of the appearance of a hard and motionless anterior cervical mass at the clinical examination and covered with normal skin.

An ultrasound with cervical CT was performed. Cervical ultrasound showed 3 contiguous oval anterior cervical masses well limited in relation to tumor recurrence. Cervical CT described a right-sided tumour-like lesional process of the thyroidectomy chamber at the cervical level, measuring 60*78*65 mm, which had a mass effect on the trachea without edging separation and without endotracheal invasion, arriving in intimate contact with thyroid cartilages and cricoids without lysis or condensation and repressing the jugulo-carotid axis which remains permeable. At the bottom, it overflows the upper orifice of the thorax by 2 cm and arrives at the contact without edging of separation of the right inominated venous trunk which remains permeable. There is also a small right-hand Level III EPA of 12 mm (Figure 3).

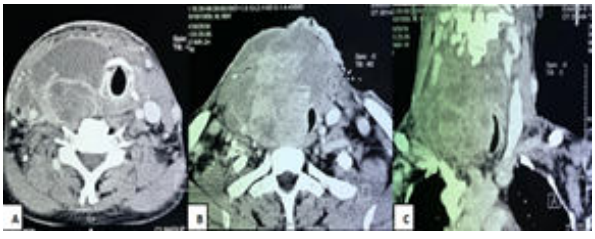

Figure 3: *cervical CT slices of tumor recurrence in axial (A-B) and coronal (C) slices showing a heterogeneous mass compressing the trachea without infiltrating it, pushing back the vascular axis, and having intimate contact with the cricoid and thyroid cartilages plunging into the chest.*

At the thoracic level, the CT shows a probable secondary lung nodules. An ascites of great abundance on the upper abdominal scannographic sections was also objectified. A osseous scan was performed as part of the extension assessment and did not note any fixation abnormalities related to secondary lesions. The evolution was rapidly progressive, marked by the rapid increase in volume with massive skin invasion (Figure 4).

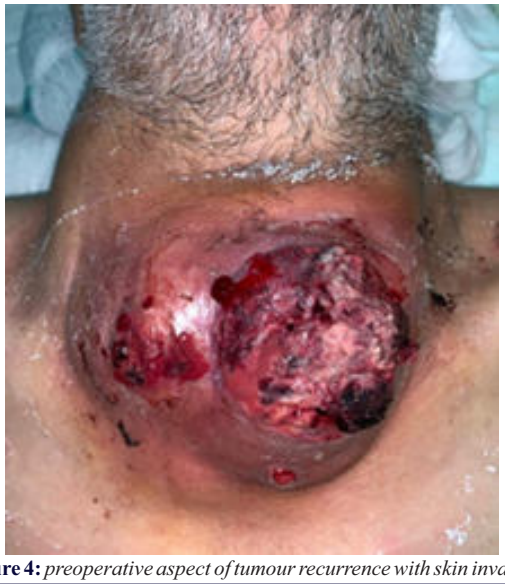

Figure 4: *preoperative aspect of tumour recurrence with skin invasion*

A tumor reduction surgery was decided, consisting of excision of the muscles sub-hyoid necrotic and infiltrated by the tumor, as well as the recurrent mass at the level of the right thyroid chamber by sacrificing the recurring nerve from the same place. The remaining of the plunging mass was removed while preserving the right vascular axis and without resorting to the tracheotomy, but the trachea remained slightly infiltrated (Figure 5).

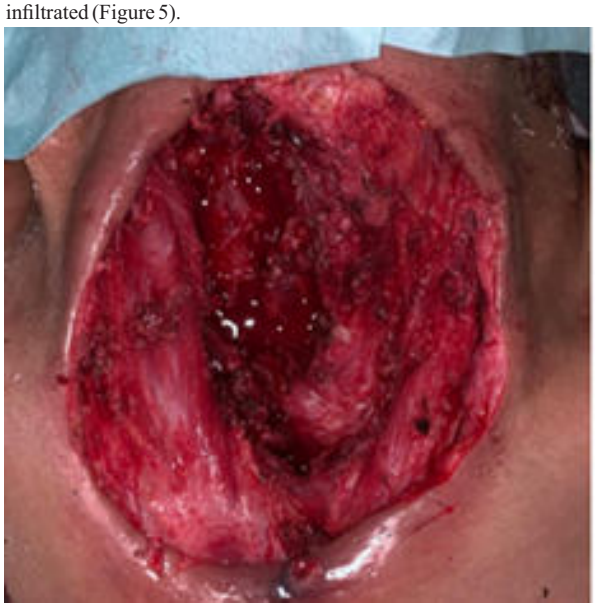

Figure 5: ***intraoperative aspect after tumor reduction surgery***

The anatomopathological study of this mass showed a morphological aspect compatible with previously diagnosed carcinosarcoma (Figure 6).

Subsequently, intensive radiotherapy was introduced at a dose of 60 gray allowing in addition to the surgery a good locoregional control. The patient died 4 months after the restart and 15 months after his first consultation because of the metastases especially pulmonary.

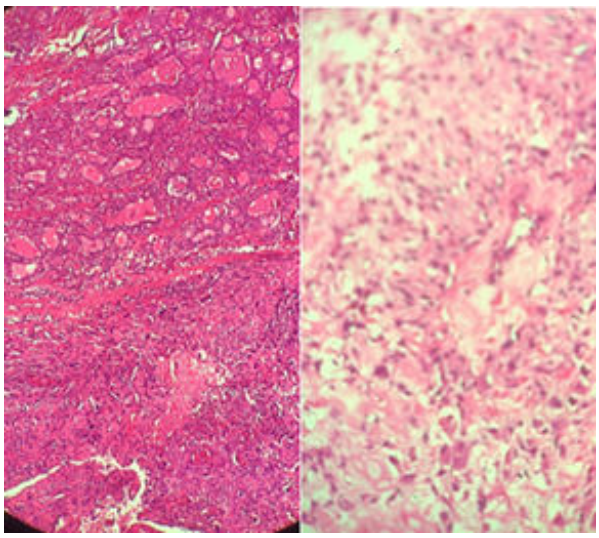

Figure 6: *Malignant tumor proliferation, often pleomorphic, composed of large cells with large nuclei, sometimes of medium size, suggesting a prominent nucleoli. They are often in mitosis. The cytoplasm of the cells is abundant, basophilic. The tumor is often little differentiated with some sites with chondroid differentiation or osteosarcomatous. This proliferation reshaped by large layers of neutrophil polynuclear, infiltrates the skin covering in the eye, the adjacent striated muscle tissue and a residual thyroid parenchyma site, without clearly individualized vascular emboles.*

DISCUSSION

Thyroid carcinosarcoma is a very rare tumour that accounts for only 0.1% of all malignant thyroid tumours. It is an undifferentiated thyroid gland malignancy that originates from both epithelial (carcinoma) and stromal (sarcoma) elements of the thyroid [3]. In fact, this term

"carcinosarcoma" was first used by Rudolf Virchow in 1865 [1]. This type of tumour was considered a variant of anaplastic carcinoma according to the latest World Health Organization (WHO 2017) classification of thyroid tumours. However, Agrawal and al. proposed that because of the double origin of this tumour, it should be considered as a separate entity from anaplastic carcinoma [2], [4]. Undifferentiated malignancies and non-epithelial thyroid tumours remain rare tumours.

Undifferentiated thyroid carcinomas account for 2% of all thyroid malignancy while thyroid sarcomas account for only 1% [3], [5]. Thirty-one cases of thyroid carcinosarcoma have been reported in the literature, and we are reporting a new case affecting this gland [1]. Thyroid carcinosarcoma is most frequently observed in women over the age of 50. Its usual evolution is that of a thyroid nodule that rapidly increases in volume, that can become compressive. However, there are reported cases of thyroid carcinosarcoma occurring on pre-existing multi-nodular goiters (GMNH) due to neoplastic metaplasia.

This case is atypical because of its evolution over 10 years in a man with a GMHN that increased its volume significantly within 3 months before his first consultation. Carcinosarcoma develops in other organs and clinical progression varies with the location of the tumour [5]. For example, patients with esophageal carcinosarcoma have a better prognosis than patients with pancreatic carcinosarcoma who are very aggressive. The site most frequently affected by this type of cancer is the uterus, which is also a rare tumour [6]. These undifferentiated tumours are poor prognoses with a low survival rate of less than 6 months, unlike well differentiated thyroid carcinomas, which have an excellent prognosis [3], [5], [6].

Exposure to phosphorus, adjuvant therapy for radioactive iodine-131 (RAI) and carmustin was considered in one study as contributing factors to the development of carcinosarcoma. Thyroid cytopuncture has no place in the preoperative diagnosis of thyroid carcinosarcoma. On the other hand, the histopathological study of the excised tissues generally allows the diagnosis of CT by showing the two mesenchymal and epithelial components. This is confirmed by immunohistochemical staining of cells carcinomatous to Thyroglobulin while mesenchymal cells are strongly positive for vimentine and S100 protein [5].

As in all thyroid malignant tumours, total thyroidectomy and complete removal of tumours are generally recommended for thyroid carcinosarcoma as long as possible. However, it is often very difficult to perform such surgery due to extensive extra-thyroid spread and regional metastases. This is done with a complete curettage of the adjacent lymph nodes [3], [5]. However, there is no consistent consensus on the therapeutic approach for thyroid carcinosarcoma due to its very low incidence, its aggressive nature with poor prognosis, and the consequent lack of large clinical series.

Adjuvant chemotherapy, radiotherapy and immunotherapy were not beneficial. Local recidivism with invasion of surrounding structures is common, probably related to the aggressiveness of the tumor. Airway obstruction and metastatic lesions in the lungs usually follow recurrence, and the prognosis is unfortunate [5]. In this reported case, local recurrence is observed only after 6 months of total thyroidectomy as a large fixed median cervical mass invading the skin and trachea.

Our patient had a tumour reduction surgery that allowed good locoregional control of the tumour and a better quality of life by avoiding the use of a tracheotomy, however the remote lung and hepatic metastases were evolving in parallel, and the patient died after 5 months of tumor reduction surgery and 11 months after total thyroidectomy.

Because of its rarity, adjuvant treatment of thyroid carcinosarcoma is not well studied [5]. Radiotherapy remains a highly recommended complementary treatment by different authors, but there is little evidence available to support this recommendation. In the case of primary thyroid sarcomas, these remain more radiosensitive than secondary or metastatic tumours, while mixed-cell sarcomas have a better radiosensitivity than fibrosarcomas [6]. Other studies have shown the effectiveness of external high-dose radiotherapy (60GY) in the regional control of locally advanced aggressive tumours. However, a similar response has not yet been proven for thyroid carcinosarcoma [5]

As for the response to chemotherapy, little information is available [6]. The use of iratherapy (RAI), can be useful in case of thyroid carcinosarcoma, but until now, there is little literature on this subject. On the other hand, in the case of extensive lung metastases, RAI is often unnecessary because of the large tumour load [6]. Despite the good locoregional control of the tumor proved by the unnecessary use of tracheotomy during the evolution of this patient, the patient died from distant metastases, not suffocation, which is the most common cause of death in patients with undifferentiated and anaplastic thyroid carcinoma [3]

CONCLUSIONS

Tyroidian carcinosarcoma is a very aggressive recurrent malignant tumour with a high metastatic power, with a bad prognosis. Until then, there is no therapeutic consensus, but total thyroidectomy with lymph node cleaning remain the basic treatment. Adjuvant treatment is highly recommended. The use of cleanliness surgeries after tumor recurrence are to try as much as possible for better local control of the tumor and to avoid tracheotomy and death by choking.

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