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Primary Leiomyosarcoma of Thyroid Gland: Case Report and Review of the Literature

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Abstract **Case Report**

Primary thyroid leiomyosarcomas are rarely seen in the thyroid gland. Due to their rarity, clinical case studies concerning management are lacking. The objective of the study is to review the literature an aggressive rare mesenchymal malignant tumor which has high mortality. From the standpoint of treatment and prognosis, the histogenesis of thyroid 'sarcomas' is academic, because these tumors, irrespective of the therapeutic intervention, are lethal. In this study, we report the case of a 28-year-old male affected by a rapidly enlarging mass of the anterior neck, associated with bilateral lung metastases, and increasing dysphagia and dyspnea during the previous 4 months. The prognosis is poor and there is no consensus regarding the treatment. A literature review was performed and thyroid leiomyosarcoma differential diagnoses, management, including alternative treatment strategies, and adjuvant therapy were analyzed. Although an improved multimodal approach is often necessary, TT and neck dissection represent the treatment of choice and are often the only possible therapy. Adjuvant radio-chemotherapy appears to be ineffective and a high mortality rate is observed.

Keywords: Thyroid, Leiomyosarcoma, Anaplastic thyroid carcinoma, Sarcomas.

INTRODUCTION

Sarcomas are an extremely rare group of tumors among all thyroid malignancies [1].

Primary leiomyosarcoma of the thyroid gland is extremely rare and accounts for 0.014% of 28 630 thyroid tumors in one series [2].

According to the histological tumor classification of the World Health Organization (WHO), thyroid leiomyosarcoma is classified as a member of the smooth muscle tumors of thyroid glands [3].

It is difficult to make a preoperative diagnosis of thyroid leiomyosarcoma and differentiate it from anaplastic thyroid carcinoma [4].

A diagnosis of LMS is dependent on the presence of smooth muscle-actin (SMA), which may be identified by immunohistochemical staining. The standard primary treatment for primary thyroid LMS is radical surgery. The long- term prognosis is poor and ~50% of patients succumb to the disease within a short period of time after diagnosis [5, 6].

TL remains a fatal tumor, and innovative and more effective therapeutic strategies to improve management and outcomes are required.

CASE REPORT

In June 2021, a A 29-year-old man complained of a rapidly growing neck mass over a period of 2 months, resulting in dysphagia and dyspnea. There was no history of a previous systemic disease, no toxic habits, and no history of radiation exposure, nor was there another primary tumor.

Ultrasound examination revealed a hypoechoic nodular lesion of the right lobe of the thyroid gland, with irregular margins and a central cystic area (28.6×25.3×31.8 mm) confirmed by a contrast medium

computed tomography (CT) scan, bilateral multiple pulmonary lesions were also identified.

Thyroid-stimulating hormone, calcitonin, thyroglobulin and carcinoembryonic antigen levels were in the normal range.

A Tir4 neoplasm fine needle cytological diagnosis of the right thyroid lobe was reached and the patient underwent total thyroidectomy (TT). Definitive histological examination identified a TL.

There was no history of radiation exposure, nor was there another primary tumor. Palpation revealed a large mass of firm consistency in the left lower neck. Free triiodothyronine and free thyroxine levels were within normal limits but thyroid-stimulating hormone levels were elevated to 5.32 IU/ml. Plasma calcitonin levels were normal. First, clinically anaplastic thyroid carcinoma was suspected.

Computed tomography (CT) demonstrated a low-density mass invading the thyroid cartilage. Dense calcification and necrotic portions were depicted within the tumor. No lymphadenopathy was seen in the neck.

Palpation-guided fine-needle aspiration biopsy revealed spindle-shaped cells with increased chromatin, prominent nucleoli, and nuclear pleomorphism. Anaplastic thyroid carcinoma was suspected but differential diagnosis included medullary carcinoma and leiomyosarcoma. The tumor was found to adhere to the larynx and trachea during surgery, and subtotal thyroidectomy with total laryngectomy was performed.

Immunohistochemical staining revealed reactivity with vimentin, smooth muscle actin, and desmin, and no reactivity with thyroglobulin, cytokeratin, calcitonin, and S-100 protein. Thus, a diagnosis of primary leiomyosarcoma of the thyroid gland was established.

The postoperative course following a total thyroidectomy (TT) was uneventful and the patient was discharged on day 3. Adjuvant postoperative therapy was performed 3 courses of doxorubicine and dacarbazine, the second one was not achived due to the poor general clinical conditions.

Recurrent tumor in the right upper neck and multiple lung metastases were found 3 weeks after the surgery. However, the patient succumbed 55 days after surgery, due to respiratory distress.

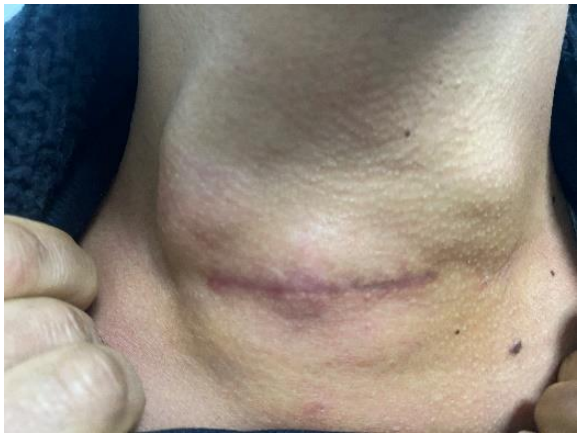

Figure 1: Four weeks recidive after surgery (TT)

DISCUSSION

Extremely rare and aggressive, anaplastic carcinoma, considered a fatal tumor, is associated with a poor survival rate, as reported for sarcomatoid carcinoma of other origins [7]. In the last 10 years, ultrasonography-guided FNAB has allowed a more precocious diagnosis [8].

Primary TL, an extremely rare neoplasm, may originate from smooth muscle cells of the capsula vessels. Metaplasia from a previously existing thyroid anaplastic carcinoma should be considered [9, 10].

At the time of first diagnosis, TL is frequently associated with distant metastases. It is a fatal tumor with a 1- year survival rate of <20%. Grossly, TL is large fleshy white-gray masses, with foci of fresh tumor necrosis and hemorrhage, and a tendency for cystic degeneration. Microscopically, the pattern of growth is usually fascicular, with tumor bundles intersecting each other. Certain tumors also present areas with a whorled appearance. The individual neoplastic cells are elongated, with abundant acidophilic fibrillary cytoplasm; the nucleus is generally centrally located and typically blunt-ended or ‘cigar-shaped’. These features also appear on cytological samples. The degree of nuclear atypia is highly variable and the mitotic activity varies considerably. High mitotic activity is virtually diagnostic of malignancy, although a TL must be strongly suspected for a tumor that is widely necrotic, hemorrhagic and with significant atypia, even if the mitotic index is low [11].

TL preoperative diagnosis can be extremely difficult. It is important to discriminate between primary TL and ‘non-thyroid’ cervical leiomyosarcoma (1% of head and neck sarcoma) and, furthermore, exclude a metastatic origin from stomach, pelvis and soft tissue [12]. CT scan and magnetic resonance imaging are useful for defining the local extent of disease and for identifying distant metastases.

Rapid loco regional infiltration and diffuse brain or lung metastases are responsible for the high

mortality rate. Total or near-total thyroidectomy, for the majority of thyroid pathologies, associated with therapeutic modified radical neck dissection should be considered for intra-thyroidal disease [13, 14]. Chemotherapy has not shown any therapeutic efficacy. Wang *et al.*, and Raspollini *et al.*, reported interesting data in the management of thyroid and uterus leiomyosarcomas through the overexpression of c-Kit proto-oncogene, a tyrosine kinase receptor [15]. However, the use of imatinib mesylate (tyrosine kinase inhibitor) did not prevent the relapse and the fatal outcome in one patient with TL associated with lung metastases [16].

In the case of loco regional infiltrating disease, surgery may be performed to prevent airway or esophageal obstruction. Often, therapies do not produce any clinical benefit, only palliative results.

TL remains a fatal tumor, invariably associated with a dismal prognosis, and, although notable improvements in oncology, an efficacious multimodal treatment protocol is lacking. To modify the poor surgical outcomes, novel and effective adjuvant therapeutic strategies, based on a molecular approach, are required [11].

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REFERENCES

1. Güney, E. (2008). Diger Malignensiler. In: Güney, E, (ed), Tiroid ve Paratiroid Bez Cerrahi Hastalıkları. Kayseri, p. 150.
2. Thompson, L. D., Wenig, B. M., Adair, C. F., Shmookler, B. M., & Heffess, C. S. (1997). Primary smooth muscle tumors of the thyroid gland. *Cancer: Interdisciplinary International Journal of the American Cancer Society*, *79*(3), 579-587.
3. DeLellis, R. A. (2004). Pathology and genetics of tumours of endocrine organs. *World Health Organization.*
4. Day, A. S., Lou, P. J., Lin, W. C., & Chou, C. C. (2007). Over-expression of c-kit in a primary leiomyosarcoma of the thyroid gland. *European archives of oto-rhino-laryngology*, *264*(6), 705-708.
5. Takayama, F., Takashima, S., Matsuba, H., Kobayashi, S., Ito, N., & Sone, S. (2001). MR imaging of primary leiomyosarcoma of the thyroid gland. *European journal of radiology*, *37*(1), 36-41.
6. Wang, T. S., Ocal, I. T., Oxley, K., & Sosa, J. A. (2008). Primary leiomyosarcoma of the thyroid gland. *Thyroid*, *18*(4), 425-428.
7. Conzo, G., Troncone, G., Docimo, G., Pizza, A., Sciascia, V., Bellevicine, C., ... & Santini, L. (2013). Cytologically undetermined thyroid's follicular lesions: surgical procedures and histological outcome in 472 cases. *Annali Italiani di Chirurgia*, *84*(3), 251-256.
8. Troncone, G., Volante, M., Iaccarino, A., Zeppa, P., Cozzolino, I., Malapelle, U., ... & Palombini, L. (2009). Cyclin D1 and D3 overexpression predicts malignant behavior in thyroid fine-needle aspirates suspicious for Hurthle cell neoplasms. *Cancer Cytopathology: A Journal of the American Cancer Society*, *117*(6), 522-529.
9. Chetty, R., Clark, S. P., & Dowling, J. P. (1993). Leiomyosarcoma of the thyroid: immunohistochemical and ultrastructural study. *Pathology*, *25*(2), 203-205.
10. Tulbah, A., Al-Dayel, F., Fawaz, I., & Rosai, J. (1999). Epstein-Barr virus-associated leiomyosarcoma of the thyroid in a child with congenital immunodeficiency: a case report. *The American journal of surgical pathology*, *23*(4), 473-476.
11. Conzo, G., Candela, G., Tartaglia, E., Gambardella, C., Mauriello, C., Pettinato, G., ... & Santini, L. (2014). Leiomyosarcoma of the thyroid gland: A case report and literature review. *Oncology Letters*, *7*(4), 1011-1014. https://doi.org/10.3892/ol.2014.1853 !!
12. Cirocchi, R., Boselli, C., Guarino, S., Sanguinetti, A., Trastulli, S., Desiderio, J., ... & Avenia, N. (2012). Total thyroidectomy with ultrasonic dissector for cancer: multicentric experience. *World Journal of Surgical Oncology*, *10*(1), 1-5.
13. De Bellis, A., Conzo, G., Cennamo, G., Pane, E., Bellastella, G., Colella, C., ... & Bellastella, A. (2012). Time course of Graves' ophthalmopathy after total thyroidectomy alone or followed by radioiodine therapy: a 2-year longitudinal study. *Endocrine*, *41*(2), 320-326.
14. Wang, T. S., Ocal, I. T., Oxley, K., & Sosa, J. A. (2008). Primary leiomyosarcoma of the thyroid gland. *Thyroid*, *18*(4), 425-428.
15. Raspollini, M. R., Amunni, G., Villanucci, A., Pinzani, P., Simi, L., Paglierani, M., & Taddei, G. L. (2004). c-Kit expression in patients with uterine leiomyosarcomas: a potential alternative therapeutic treatment. *Clinical cancer research*, *10*(10), 3500-3503.
16. Day, A. S., Lou, P. J., Lin, W. C., & Chou, C. C. (2007). Over-expression of c-kit in a primary leiomyosarcoma of the thyroid gland. *European archives of oto-rhino-laryngology*, *264*(6), 705-708.