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# Thyroid Sarcoma: Case Report, Critical Literature Review, and Recent Advances

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ABSTRACT

Thyroid sarcoma is an extremely rare malignant neoplasm, accounting for less than 1% of all thyroid malignancies. This review article presents a clinical case and discusses the epidemiology, histological classification, clinical features, diagnosis, treatment, and prognosis of thyroid sarcoma. It emphasizes the importance of early diagnosis and multidisciplinary management in improving outcomes for affected patients.



## Introduction

Although thyroid neoplasms are common, thyroid sarcomas are exceptionally rare, accounting for an estimated less than 1% of all thyroid malignancies [1]. These mesenchymal tumors may arise from different tissue components of the gland and are characterized by aggressive clinical behavior.

## Case Report

A 52-year-old male presented to the emergency department with a four-month history of a progressively enlarging cervical mass associated with pain, dysphagia to solids, and dyspnea, but no stridor. Physical examination revealed a large, firm, immobile, heterogeneous mass in the anterior neck region, associated with right cervical lymphadenopathy. Neck CT scanning revealed a bulky mass compressing the trachea and esophagus, located in the anterior mediastinum and in relation to the right thyroid lobe (Figure 1).

The patient underwent oncologic thyroidectomy, revealing an enlarged thyroid gland with a right lobe measuring 12 × 10 cm extending into the prevertebral space and across to the left thyroid compartment, displacing the trachea and esophagus anteriorly. The tumor infiltrated esophageal musculature, precluding identification of the recurrent laryngeal nerve due to involvement of the tracheoesophageal groove. Right pre- and paratracheal lymph nodes measuring 1.5 cm were also identified.

The pathology report described an R1 resection (complete macroscopic resection with microscopically positive margins). Mitotic activity was >20 per 2 $mm^2$. Microscopy examination revealed spindle cell proliferation arranged in fascicles accompanied by marked nuclear atypia, frequent mitoses, and necrotic areas. Immunohistochemistry showed positivity for desmin, AML, h-caldesmon, vimentin, S100, CD31, CD56, CD99, and BCL-2, with a Ki-67 index of 80%. p53 showed strong nuclear overexpression consistent with a mutated profile. Negative markers included CKAE1/AE3, EMA, CD57, PAX-8, thyroglobulin, synaptophysin, ERG, and SOX10. Cam5.2 showed aberrant paranuclear expression. The Immunohistochemical findings were consistent with primary thyroid leiomyosarcoma.

One month after surgery, the patient was re-admitted due to persistent, rapidly progressive disease. Contrast-enhanced neck CT demonstrated

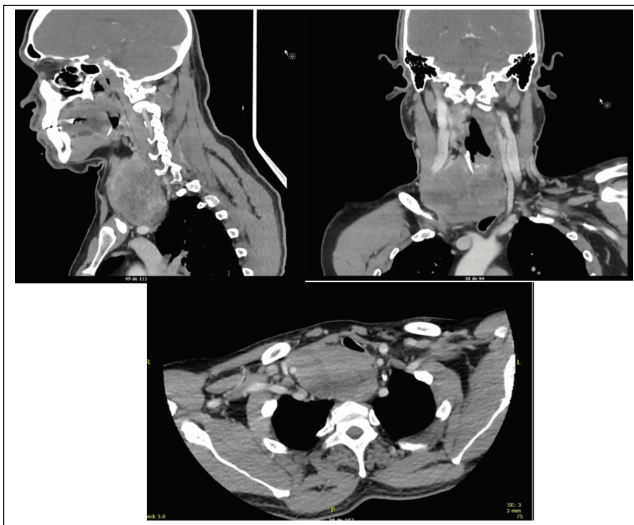

**Figure 1:** Contrast-enhanced CT of the neck: heterogeneous mass with solid and cystic components and heterogeneous enhancement. **Size**: 134 × 85 × 106 mm (longitudinal × anteroposterior × transverse), displacing the cervical esophagus, trachea, and cartilaginous structures. The mass abuts the right sternocleidomastoid muscle and loses the cleavage plane. **A**: Sagittal reconstruction; **B**: Coronal reconstruction; **C**: Axial reconstruction.

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residual tumor within the thyroid bed, extending into the trachea, cervical esophagus, and laryngeal cartilages. Given the resulting airway and esophageal compromise, a tracheostomy and gastrostomy were performed. An interdisciplinary tumor board recommended initiating external beam radiotherapy with intensity-modulated radiotherapy (IMRT) (2 Gy per fraction to a total dose of 60 Gy to the thyroid bed), with subsequent consideration of salvage surgery or systemic therapy with adjuvant chemotherapy, depending on treatment response.

## Discussions

## Epidemiology

Primary thyroid sarcoma is a rare malignant non-epithelial tumor characterized by smooth muscle differentiation, originating from vascular wall smooth muscle within the thyroid gland [1]. It represents only 0.01–1.5% of all malignant thyroid neoplasms [3]. Most cases are diagnosed in patients over 40 years of age [2]. In a series of 142 cases, the peak incidence occurred between 60 and 79 years, with a slight female predominance [3]. A higher incidence has also been noted in patients with a history of cervical radiotherapy [3]. The most common histologic subtypes include angiosarcoma (20.4%), malignant hemangioendothelioma (16.3%), malignant fibrous histiocytoma (14.1%), leiomyosarcoma (11.3%), and fibrosarcoma (9.2%) [4]. These tumors arise from mesenchymal elements of the thyroid and are typically associated with an aggressive clinical course.

### Clinical Manifestations:

Patients with thyroid sarcoma typically present with a rapidly enlarging painless cervical mass, which may be accompanied by dysphagia or dyspnea secondary to compression of adjacent structures. In one case series, 79.6% of patients presented with a painless goiter as the initial symptom [4]. Rapid tumor growth and local invasion are characteristic features of this malignancy.

### Diagnosis

*Imaging.* Imaging techniques play a key role in evaluation of these lesions. On ultrasound, thyroid sarcomas may appear as a hypoechoic or heterogeneous mass with irregular margins [5]. CT typically shows a heterogeneous solid mass with areas of necrosis or hemorrhage and poorly defined borders, features that reflect the rapid proliferation and invasive nature of the tumor. MRI reveals a heterogeneous lesion with variable post-contrast enhancement and is particularly valuable for assessing invasion adjacent structures such as the trachea, esophagus, or cervical vessels [6].

*Biopsy.* Fine-needle aspiration (FNA) is essential for histological diagnosis, although interpretation may be limited by tumor heterogeneity. Histological examination typically demonstrates spindle-shaped or pleomorphic cells arranged in fascicular or storiform patterns, with frequent mitotic figures and extensive areas of necrosis [7]. Immunohistochemistry distinguishes subtypes by expression of markers such as vimentin, desmin, h-caldesmon, CD34, or smooth muscle actin. In selected cases, fluorescence in situ hybridization (FISH) can detect specific genetic translocations, offering both diagnostic confirmation and potential therapeutic guidance [8].

### Treatment

The standard treatment for thyroid sarcoma is complete surgical resection with negative margins. In cases of localized disease, this approach is associated with 5-year survival rates of approximately 40–50%; however, outcomes decline significantly in the presence of metastatic disease or incomplete resection (6). Given its aggressive nature and high local recurrence rate (up to 60%), multidisciplinary management including adjuvant chemotherapy is recommended, particularly for high-grade tumors. Regimens such as ifosfamide–doxorubicin or gemcitabine–docetaxel have shown progression-free response rates of 25–40% depending on histologic subtype [9].

External beam radiotherapy, particularly IMRT, is used in cases with positive surgical margins or locally advanced disease, achieving local control rates of 40–60% (10). Nevertheless, its use carries risks including fibrosis, dysphonia, and hypoparathyroidism [11]. Importantly, neither adjuvant radiotherapy nor chemotherapy has demonstrated a significant improvement in overall survival [11].

Targeted therapies, including tyrosine kinase inhibitors and epigenetic agents, have been explored in selected cases with defined genetic -such as the EWSR1-FLI1 translocation in Ewing sarcoma- although their role in thyroid sarcoma remains experimental [12]. Despite these approaches, overall prognosis remains poor due to rapid disease progression and resistance to conventional therapy.

### Prognosis

Due to its mesenchymal origin, thyroid sarcoma exhibits aggressive clinical behavior, with 5-year overall survival rates ranging from 20% to 50%, depending on histologic subtype [12]. Synovial sarcoma and thyroid angiosarcoma are associated with worse prognosis compared with leiomyosarcoma or malignant fibrous histiocytoma. The lungs represent the most common site of metastasis [3]. Presence of metastatic disease reduces survival to less than 20% at 2 years [13], whereas complete surgical resection with negative margins in non-metastatic cases improves both local disease control and overall survival.

Nodal metastasis is uncommon in most myogenic sarcomas; however, in primary thyroid leiomyosarcoma, nodal involvement has been reported in up to 50% of cases [16]. Local recurrence is also frequent, occurring in up to 60% of cases [14,15], contributing to the high mortality of this entity.

## Conclusion

Thyroid sarcoma is an exceptionally rare malignancy, representing less than 1% of all thyroid cancers. The differential diagnosis includes other aggressive thyroid neoplasms, particularly anaplastic carcinoma. Treatment typically involves radical surgery, often in combination with radiotherapy or chemotherapy; however, prognosis remains poor due to the tumor's aggressive biology. Further research is essential to elucidate the molecular mechanisms underlying these neoplasms and to identify more effective therapeutic strategies. Early diagnosis and multidisciplinary management are key to improving clinical outcomes.

**Ethical Responsibilities:** The authors declare that no human or animal experiments were conducted for this study.

**Data Confidentiality:** The authors declare adherence to their institution's protocols for patient data publication.

**Right to Privacy and Informed Consent:** Written informed consent was obtained from the patient described in this report, and the document is held by the corresponding author.

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