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RESEARCH ARTICLE

THYROID ANGIOSARCOMA: A CASE REPORT AND REVIEW OF LITERATURE

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Abstract

Thyroid angiosarcoma is a rare and highly aggressive vascular malignancy associated with poor prognosis due to rapid local invasion and early metastasis. We report the case of a 68-year-old Moroccan woman presenting with a rapidly enlarging cervical mass associated with dyspnea and dysphonia evolving over 3 months. Thyroid function tests were within normal limits. Cervical computed tomography revealed a heterogeneous bilateral thyroid mass measuring 8 cm, with tracheal compression but no clear vascular invasion or lymph node involvement The patient underwent total thyroidectomy. Histopathological examination showed malignant vascular proliferation. Immunohistochemistry demonstrated strong positivity for ERG, CD31, and CD34, while epithelial markers (cytokeratin AE1/AE3, TTF-1) were negative, supporting the diagnosis of primary thyroid angiosarcoma. Surgical margins were close (<1 mm). Adjuvant radiotherapy was administered to the thyroid bed, delivering a total dose of 66 Gy in 33 fractions. However, pulmonary metastases developed within one month, and the patient died shortly thereafter. This case highlights the aggressive behavior of thyroid angiosarcoma and emphasizes the importance of accurate diagnosis and multidisciplinary management despite limited therapeutic efficacy.

Introduction:-

Primary thyroid angiosarcoma is an exceptionally rare malignancy, accounting for less than 1% of thyroid cancers worldwide [1]. It has been predominantly reported in Alpine regions, where iodine deficiency and endemic goiter have been suggested as potential risk factors [2].Histologically, this tumor is characterized by malignant endothelial proliferation and demonstrates strong expression of vascular markers such as CD31, CD34, and ERG, which are essential for diagnosis [3]. However, its morphological overlap with anaplastic thyroid carcinoma represents a major diagnostic challenge and requires careful immunohistochemical differentiation [4].Clinically, thyroid angiosarcoma exhibits highly aggressive behavior, with rapid local invasion and early distant metastasis, leading to poor prognosis despite multimodal treatment [5–7]. Previous studies have consistently reported limited survival outcomes, often less than one year [8].Although initially considered geographically restricted, more recent reports have documented cases outside endemic regions, suggesting a broader epidemiological distribution and highlighting the need for increased awareness among clinicians [9,10].

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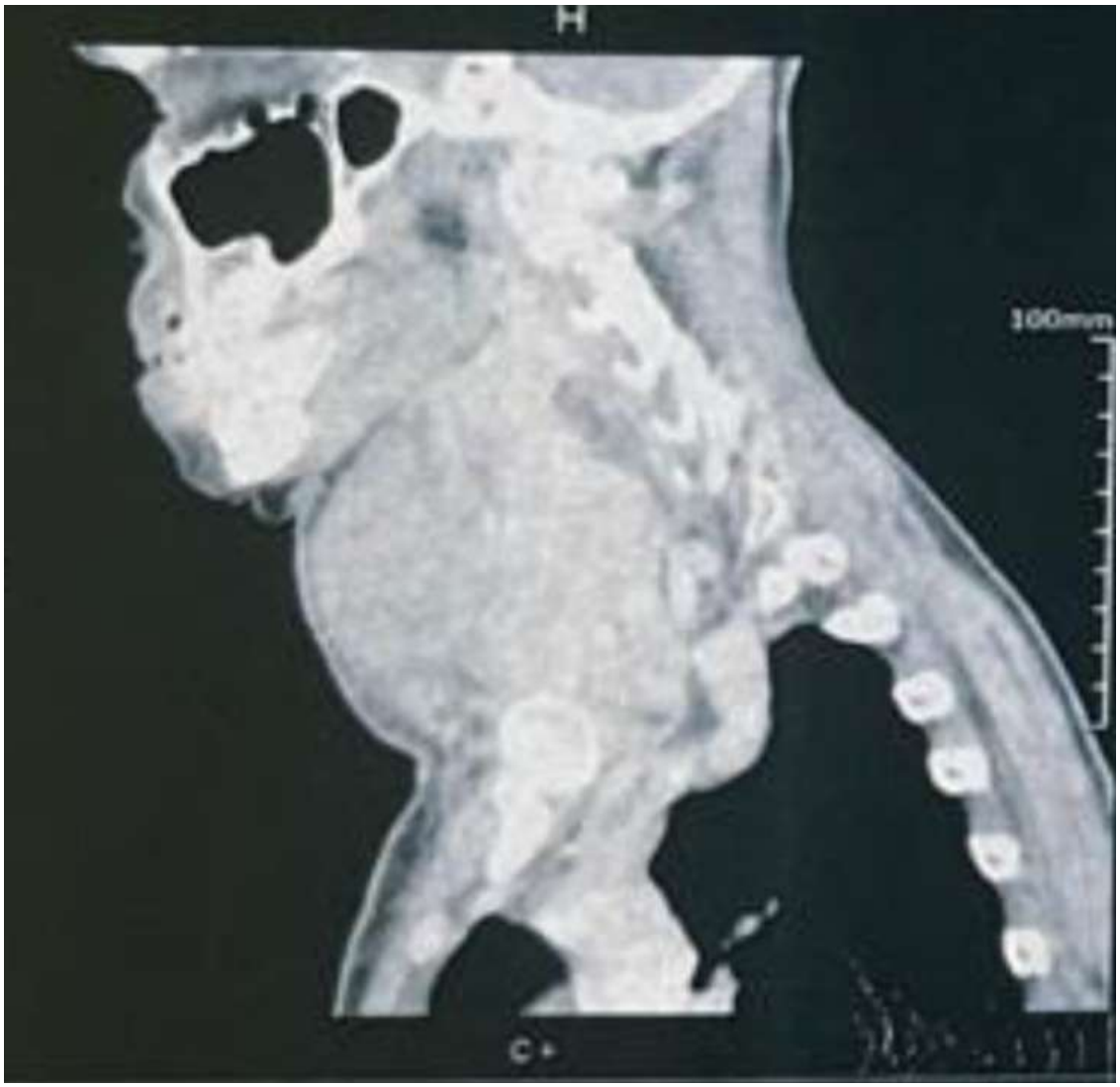
Case presentation:

We report the case of a 68-year-old Moroccan woman presenting with a rapidly enlarging anterior cervical mass, evolving over three months, associated with dysphonia and dyspnea. There was no history of radiation exposure or thyroid disease.Clinical examination revealed a firm, fixed thyroid mass without palpable lymphadenopathy. Thyroid function tests were normal.Cervical CT showed a heterogeneous mass measuring 8 × 6 cm involving both thyroid lobes, with tracheal compression but no vascular invasion or lymph node involvement.



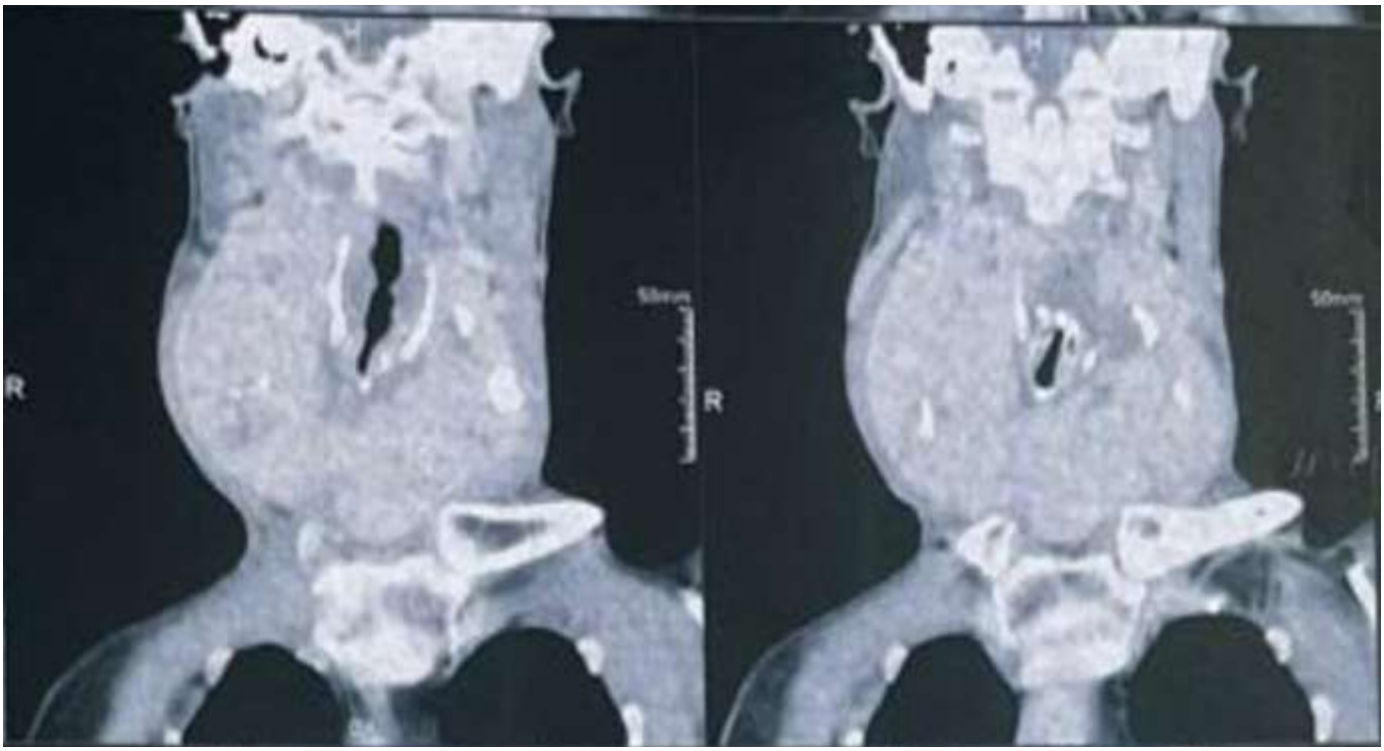


Figure1: sagittal and transverse section of the cervical CT scan

The patient underwent total thyroidectomy. Histopathological examination demonstrated malignant vascular proliferation, and the diagnosis of thyroid angiosarcoma was confirmed by strong immunohistochemical expression of endothelial markers including ERG, CD31, and CD34 [4]. Surgical margins were close (<1 mm).

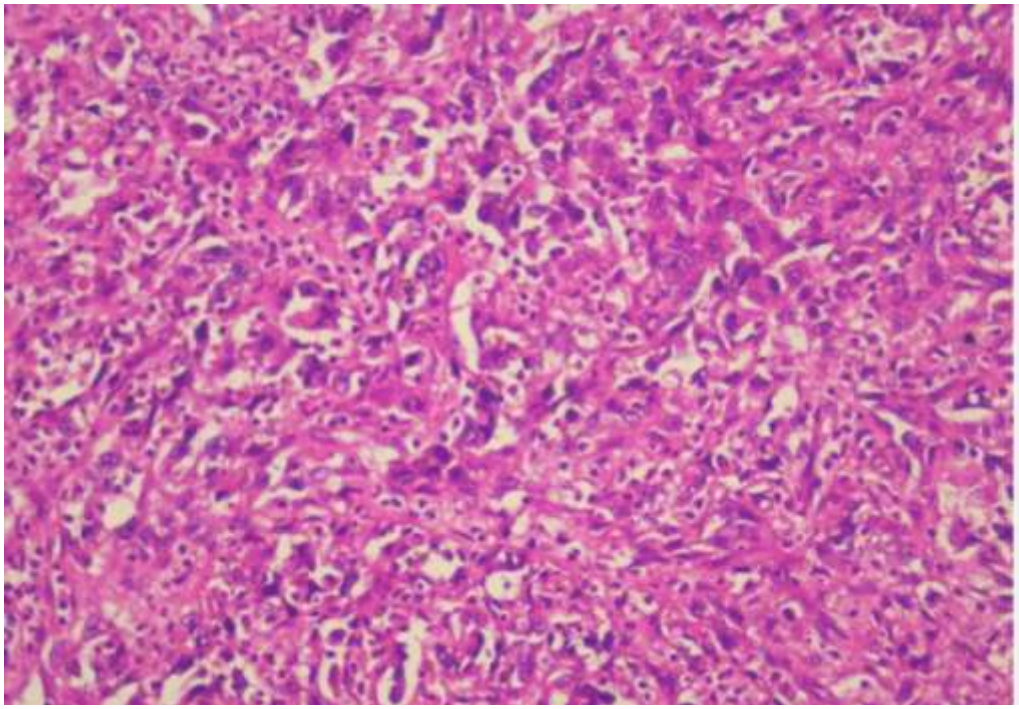


Figure2: histological image of the thyroid tumor

Adjuvant radiotherapy was administered to the thyroid bed, delivering a total dose of 66 Gy in 33 fractions.; however, early pulmonary metastases developed, and the patient died one month after completion of treatment [3].

Discussion:-

Primary thyroid angiosarcoma is an exceptionally rare and highly aggressive vascular malignancy, historically described in Alpine regions but increasingly reported worldwide [2,9,10]. While iodine deficiency and long-standing goiter have been proposed as potential risk factors, the occurrence of cases in non-endemic areas, such as Morocco, suggests that additional environmental or molecular mechanisms may be involved.Clinically, thyroid angiosarcoma typically presents as a rapidly enlarging cervical mass associated with compressive symptoms, including dyspnea and dysphonia, as observed in our patient. This rapid growth reflects the highly proliferative nature of the tumor and often leads to delayed diagnosis due to its resemblance to more common thyroid malignancies. Radiological findings are non-specific and frequently mimic anaplastic thyroid carcinoma, with large heterogeneous masses showing necrosis and local invasion [4]. Therefore, imaging alone is insufficient for diagnosis and must be complemented by histopathological and immunohistochemical evaluation.

From a pathological perspective, the main diagnostic challenge lies in distinguishing thyroid angiosarcoma from anaplastic carcinoma and other high-grade thyroid neoplasms. Immunohistochemistry plays a central role, with strong expression of endothelial markers such as CD31, CD34, and ERG confirming vascular differentiation [3]. The absence of epithelial markers, including cytokeratins and TTF-1, is crucial to exclude anaplastic carcinoma [4]. This diagnostic approach is essential, as misclassification may lead to inappropriate management strategies.The prognosis of thyroid angiosarcoma remains extremely poor due to its aggressive biological behavior. Several studies have demonstrated a high rate of early hematogenous dissemination, particularly to the lungs, which represents the most common site of metastasis [5–7]. Reported survival is typically less than one year, even with multimodal treatment [8]. In our case, the patient developed pulmonary metastases shortly after treatment and died within two months, highlighting an even more aggressive clinical course.

Therapeutically, complete surgical resection remains the cornerstone of management when feasible. Adjuvant radiotherapy is often recommended to improve local control, particularly in cases with close or positive margins. However, its impact on overall survival remains limited due to the high risk of systemic relapse [5,7]. Chemotherapy has not demonstrated consistent benefit, and there is currently no standardized systemic treatment. Recent advances in oncology suggest that anti-angiogenic therapies and targeted agents may represent promising approaches, given the vascular origin of the tumor, although clinical evidence remains limited.Overall, this case is consistent with previously reported data but also underscores the extreme aggressiveness and rapid progression of thyroid angiosarcoma. It highlights the importance of early diagnosis, accurate immunohistochemical characterization, and multidisciplinary management. Nevertheless, the lack of effective systemic therapies remains a major limitation, emphasizing the need for further research to better understand tumor biology and develop more effective treatment strategies.

- **Methods for Literature Review:-**

A literature search was conducted using PubMed and Scopus databases. Studies reporting clinical data on primary thyroid angiosarcoma were included.

- **Results of Literature Review:-**

Thyroid angiosarcoma predominantly affects elderly patients, with slight female predominance. Presentation typically involves a rapidly enlarging neck mass with compressive symptoms.Immunohistochemical staining consistently shows positivity for endothelial markers (CD31, CD34, ERG), essential for diagnosis [3].Prognosis remains poor, with early metastasis and median survival often less than one year despite treatment [6–8].

Table 1. Clinicopathological features of reported cases of thyroid angiosarcoma

Study	Age	Region	IHC Profile	Treatment	Outcome
Papotti et al. [1]	60–80	Alpine	CD31+, CD34+	Surgery ± RT	<12 months
McCully et al. [2]	~70	Europe	ERG+, CD31+	Multimodal	Poor
Zöller et al. [4]	65	Non-Alpine	CD31+, CD34+	Surgery + RT	Metastatic
Randle et al. [5]	Variable	USA	Variable	Multimodal	Poor

Conclusion:-

Primary thyroid angiosarcoma is a rare and highly aggressive malignancy associated with poor prognosis due to rapid local progression and early metastatic dissemination [5–8]. Diagnosis relies on histopathological examination supported by immunohistochemical confirmation of endothelial differentiation [3,4]. Despite multimodal management combining surgery and radiotherapy, outcomes remain unfavorable, mainly due to early distant relapse [5,7]. This case highlights the importance of early diagnosis, accurate pathological characterization, and multidisciplinary management. However, current therapeutic strategies remain insufficient, underscoring the need for further research and the development of more effective systemic treatments.

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