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Primary Thyroid Lymphomas: A Decade of Clinical Experience through Eight Case Reports

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Abstract | **Case Report**

***Background*:** Primary thyroid lymphoma (PTL) is a rare malignancy, accounting for less than 5% of all thyroid cancers and less than 2% of extranodal lymphomas. It typically affects elderly individuals and is often associated with chronic lymphocytic thyroiditis, particularly Hashimoto's disease. The diagnosis can be challenging due to overlapping clinical and imaging features with other aggressive thyroid malignancies such as anaplastic carcinoma. ***Objective*:** This study aims to present and analyze eight cases of primary thyroid lymphoma managed in our department, highlighting the clinical presentations, radiological features, histopathological subtypes, therapeutic approaches, and patient outcomes. ***Methods*:** We conducted a retrospective analysis of eight patients diagnosed with PTL over a five-year period. Clinical symptoms, imaging findings (ultrasound and CT scan), histopathological and immunohistochemical results, treatment modalities, and follow-up outcomes were documented. EU-TIRADS classification was used to stratify the ultrasound features of the thyroid lesions. ***Results*:** The patients (5 men and 3 women) ranged in age from 45 to 78 years. All presented with a rapidly enlarging neck mass, with compressive symptoms in six cases and systemic B symptoms in three. Two patients had superior vena cava syndrome, and one had Claude Bernard-Horner syndrome. Ultrasound revealed hypoechoic, heterogeneous lesions with irregular margins in all cases, and EU-TIRADS scores ranged from 4 to 5. CT imaging showed large thyroid masses with mediastinal extension in four cases. Histological analysis identified diffuse large B-cell lymphoma (DLBCL) in all eight cases. All patients received R-CHOP chemotherapy, and three received adjunctive radiotherapy. Despite treatment, two patients died due to severe local complications before completing therapy. The remaining six showed good clinical and radiological response with long-term remission in four. ***Conclusion*:** Primary thyroid lymphoma, though rare, must be considered in elderly patients with a rapidly growing thyroid mass, especially in the context of autoimmune thyroiditis. Early diagnosis through adequate imaging and biopsy is crucial for initiating timely treatment. Chemotherapy, particularly with R-CHOP, remains the mainstay of management. A multidisciplinary approach improves outcomes, although prognosis remains guarded in advanced cases with compressive complications.

Keywords: Primary Thyroid Lymphoma, B-Cell Non-Hodgkin Lymphoma, Thyroiditis, Hashimoto's Disease, Neck Mass.

INTRODUCTION

Primary thyroid lymphoma is a rare form of thyroid malignancy, posing diagnostic and therapeutic challenges due to its rapid progression and resemblance to anaplastic carcinoma or advanced Hashimoto's thyroiditis. Despite its rarity, timely recognition is crucial, as PTL is highly responsive to nonsurgical treatments such as chemotherapy and radiotherapy. We present eight cases collected over a decade, emphasizing diverse presentations and management outcomes to contribute to the evolving understanding of this rare entity.

PATIENTS AND METHODS

Between March 2014 and June 2024, eight patients were diagnosed with primary thyroid lymphoma at the ENT department of Mohammed VI University Hospital. The cohort comprised six males and two females, with a mean age of 54 years (range: 46–71 years).

Only one patient had a prior history of Hashimoto's thyroiditis, managed medically for over ten years. Two others had comorbid diabetes, and one patient had a history of controlled hypertension. The remaining four patients were otherwise healthy individuals. All presented to the emergency department with rapidly expanding anterior neck masses, developing over weeks in some and a few months in others.

Five patients experienced progressive dyspnea and inspiratory stridor, while four reported dysphagia to solids and dysphonia. Cervical examination revealed massive goiters in all cases—immobile in four, mobile in the others—with palpable cervical lymphadenopathy. Two patients exhibited superior vena cava syndrome (figure 1), and one presented with a complete Claude Bernard-Horner syndrome (ptosis, miosis, and enophthalmos), reflecting deep local invasion (figure 2).

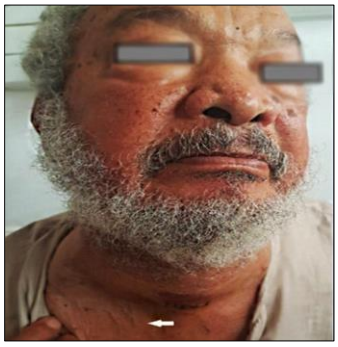

Figure1: Clinical presentation of a patient with a cervico-mediastinal mass and superior vena cava syndrome, demonstrating orbital edema. The white arrow highlights the presence of collateral venous circulation

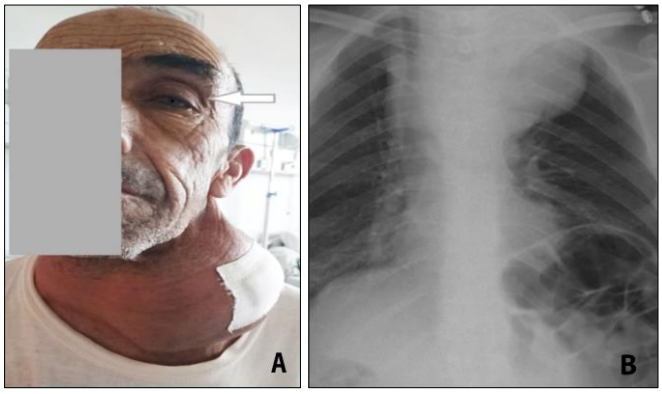

Figure 2: Patient presenting with a large cervical mass associated with Claude Bernard-Horner syndrome. A: Clinical representation showing ptosis, indicated by the black arrow. B: Chest X-ray of the same patient demonstrating a sizeable compressive cervico-mediastinal mass

Nasofibroscopy consistently revealed unilateral vocal cord immobility. All patients were admitted to the intensive care unit for initial stabilization. Corticosteroids were initiated, and one patient required emergency tracheostomy for airway compromise (figure 3).

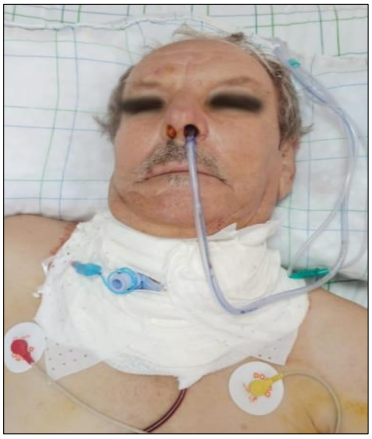

Figure 3: Clinical representation of a patient with a cervico-mediastinal mass required emergency tracheotomy

Contrast-enhanced CT scans showed bulky thyroid masses with I nfiltration of the prevertebral fascia and compression of the trachea, esophagus, and major vascular structures. The average disease duration before admission was seven months, ranging from 4 to 23 months (figure 4).

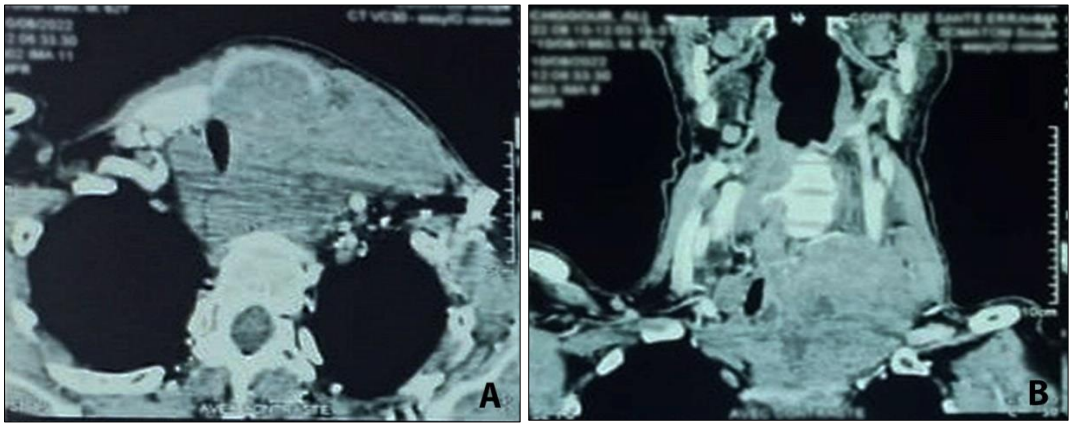

Figure 4: CT scan images demonstrating a voluminous and compressive goiter in a patient with thyroid disease, A: axial cuts, B: frontal reconstructions

All patients had normal thyroid hormone profiles, but elevated Erythrocyte Sedimentation Rate (ESR) and lactate dehydrogenase (LDH) levels were noted. Open biopsy was necessary due to non-diagnostic fine-needle aspiration, and histopathology confirmed large B-cell non-Hodgkin lymphoma in every case (figure 5).

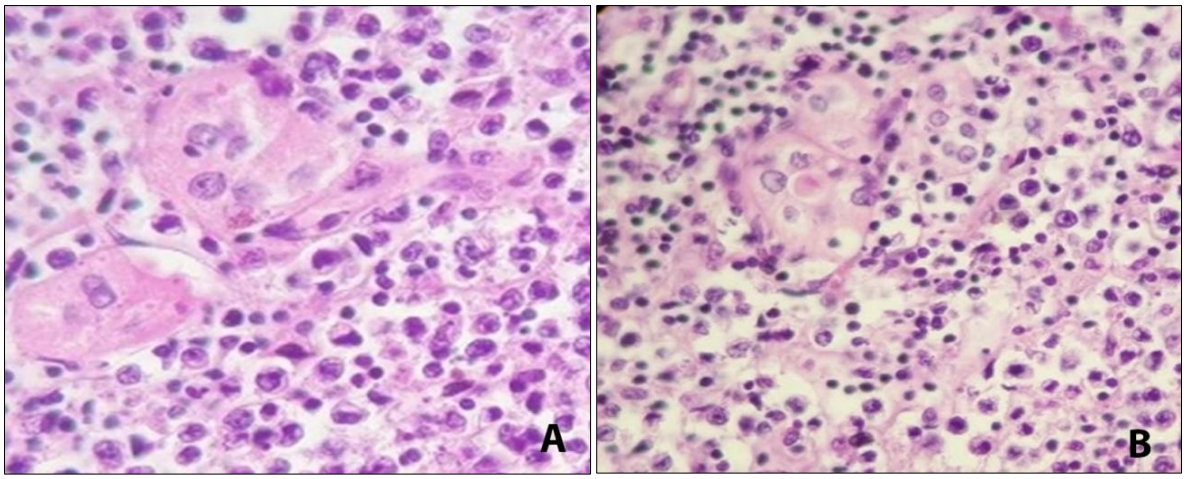

Figure 5: Histological representations with hematoxylin-eosin staining, magnification ×10, A: The presence of large lymphomatous cells characterized by a high nucleocytoplasmic ratio, anisokaryotic nuclei with prominent nucleoli, and observable mitotic figures. B: Thyroid localization of diffuse large cell lymphoma proliferation

Staging workups—including chest-abdominal-pelvic CT scan, bone marrow biopsy, and laboratory panels—revealed stage IE disease in three patients and stage IIE in five, based on the Ann Arbor classification. Patients were referred to the hematology unit and started on R-CHOP chemotherapy.

Unfortunately, one patient with Superior Vena Cava syndrome died from complications during the first chemotherapy cycle. Another died after six months of therapy. Two patients are currently under active treatment and follow-up, while four others are in confirmed remission (two at 18 months and two at 24 months).

Table 1: Summary of Clinical, Radiological, and Therapeutic Features of the 8 Patients with Primary Thyroid Lymphoma (Abbreviations: DLBCL: Diffuse Large B-Cell Lymphoma, SVC: Superior Vena Cava, FNAC: Fine Needle Aspiration Cytology, RT: Radiotherapy, IE/IIE: Ann Arbor stages)

Patient	1	2	3	4	5	6	7	8
Age	46	49	53	55	57	60	67	71
Sex	M	M	F	M	M	M	M	F
History	None	Type 2 Diabetes	Hashimotos (10 yrs)	None	Type 2 Diabetes	None	HTA	None
Clinical Signs	Rapid goiter, dysphagia, dysphonia	Rapid goiter, dyspnea, SVC syndrome	Rapid goiter, dysphagia	Cervical mass, Claude Bernard-Horner syndrome	Rapid neck swelling, dysphonia	Goiter, stridor, required tracheotomy	Rapid goiter, cervical pain, fatigue	Goiter, dysphagia, mild dyspnea
Ultrasound (EU-TIRADS)	Hypoechoic diffuse mass, cervical adenopathies — EU-TIRADS 5	Heterogeneous mass, enlarged nodes — EU-TIRADS 5	Solid hypoechoic mass, irregular borders — EU-TIRADS 5	Mixed echogenicity, posterior extension — EU-TIRADS 4	Multinodular hypoechoic thyroid — EU-TIRADS 4	Solid isoechoic mass, compressive signs — EU-TIRADS 5	Hypoechoic nodular lesion, intrathyroidal — EU-TIRADS 4	Diffuse thyroid enlargement, microcalcifications — EU-TIRADS 5
CT Scan Findings	Large thyroid mass compressing trachea; cervical lymph nodes	Invasive mass to upper mediastinum, SVC compression	Bilateral cervical mass with tracheal displacement	Large mass invading carotid space and upper mediastinum	Mass compressing larynx; enlarged paratracheal nodes	Central mass compressing laryngotracheal axis	Localized thyroid lesion with level II adenopathies	Thyroid mass with level III lymph nodes
Biopsy	Open	Open	FNAC + Open	Open	Open	Open (emergency)	Open	Open
Histology	DLBCL	DLBCL	DLBCL	DLBCL	DLBCL	DLBCL	DLBCL	DLBCL
Stage	IE	IIE	IE	IIE	IIE	IIE	IE	IIE
Treatment	R-CHOP x6	R-CHOP (1 cycle)	R-CHOP x6 + RT	R-CHOP x6	R-CHOP x6	R-CHOP x6	R-CHOP x6	R-CHOP x6
Evolution	Complete remission at 24 months	Deceased during first cycle	Remission at 18 months	Deceased at 6 months (progressive disease)	Under treatment (12 months)	Under treatment (12 months)	Remission at 24 months	Remission at 18 months

DISCUSSION

Primary thyroid lymphoma (PTL) remains a rare but clinically significant entity that must be recognized promptly due to its aggressive presentation and the potential for curative treatment. Though uncommon, it should always be considered in the differential diagnosis of rapidly enlarging thyroid

masses, particularly in older patients or those with a history of autoimmune thyroiditis.

Many clinicians tend to assume that a thyroid nodule or mass with a high EU-TIRADS score, particularly EU-TIRADS 5, is of epithelial origin—such as papillary or anaplastic thyroid carcinoma—which is accurate in the majority of cases. However, this assumption can lead to misdiagnosis or delayed identification of less common entities like thyroid lymphomas. Though rare, both primary and secondary lymphomas must be considered in the differential diagnosis of a rapidly enlarging thyroid mass. The essential criterion to define a lymphoma as "primary" is the confinement of disease to the thyroid gland and, potentially, the regional lymph nodes, without evidence of systemic involvement or another primary site. Full-body imaging and bone marrow biopsy are thus crucial for excluding secondary lymphoma involvement and accurately staging the disease [1].

The majority of primary thyroid lymphomas are of the B-cell non-Hodgkin subtype, especially the diffuse large B-cell lymphoma (DLBCL), which is known for its aggressive behavior. T-cell lymphomas and Hodgkin's disease within the thyroid are far less common. One of the key predisposing factors is chronic autoimmune thyroiditis, particularly Hashimoto's thyroiditis. Although the thyroid gland lacks native lymphoid tissue, prolonged autoimmune stimulation in Hashimoto's disease results in lymphocytic infiltration, which may undergo malignant transformation over time. This explains the strong epidemiological association: patients with Hashimoto's thyroiditis have a 60- to 80-fold increased risk of developing primary thyroid lymphoma compared to the general population [1, 2].

Primary thyroid lymphoma is typically observed in women in their sixth or seventh decades of life, with a female-to-male ratio of around 8:1. This trend is believed to reflect the higher incidence of autoimmune thyroiditis in women. Interestingly, our series diverges from this pattern, showing a male predominance. Clinically, patients often present with a rapidly enlarging cervical mass, sometimes associated with compressive symptoms such as dyspnea, dysphagia, hoarseness, or stridor. Systemic symptoms like weight loss, fever, and night sweats (B symptoms) are less frequently observed. In our study, two patients presented with superior vena cava syndrome—a life-threatening complication—while one developed Claude Bernard-Horner syndrome, reflecting mediastinal invasion. Such features, although rare, highlight the aggressiveness and local invasiveness that thyroid lymphomas can display [3-9].

Ultrasound remains the first-line imaging tool for evaluating thyroid pathology. In primary thyroid lymphoma, sonographic patterns vary and may include diffuse hypoechoic infiltration, solitary or multiple nodular lesions, or a combination of both. EU-TIRADS classification, although primarily designed for epithelial thyroid nodules, can still suggest suspicious features such as irregular margins, marked hypoechogenicity, and internal vascularity. However, EU-TIRADS cannot differentiate between carcinoma and lymphoma. Fine needle aspiration cytology (FNAC) is often performed but may be insufficient for subtyping lymphomas due to the need for architectural evaluation and immunophenotyping. In such cases, a core needle or open surgical biopsy remains the gold standard to establish a definitive diagnosis [4-8].

Diffuse large B-cell lymphoma is the most common subtype of primary thyroid lymphoma, often presenting as a bulky mass with rapid progression. Other subtypes include mucosa-associated lymphoid tissue (MALT) lymphoma, which tends to follow a more indolent course. Rarer types include T-cell lymphomas, Burkitt's lymphoma, plasmacytoma, and Hodgkin's lymphoma. Histological classification is critical, as it guides therapeutic decisions and prognosis. The combination of chemotherapy and, in some cases, radiotherapy remains the cornerstone of treatment. The R-CHOP regimen—comprising rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone—has become the standard of care for DLBCL. MALT lymphomas may respond well to radiotherapy alone. Our patients were uniformly treated with R-CHOP, reflecting the aggressive histology seen in all eight cases [6, 7].

Staging follows the Ann Arbor classification, which remains widely used despite the emergence of newer functional imaging modalities. Stage IE refers to disease confined to the thyroid gland; stage IIE includes involvement of adjacent lymph nodes; stages IIIE and IVE denote more extensive nodal or extranodal spread. Our series included three patients with stage IE and five with stage IIE disease. Prognosis is significantly influenced by staging: reported five-year survival rates are around 80% for stage IE, 50% for IIE, and less than 36% for advanced stages. Timely initiation of systemic therapy is key, as delays—especially in patients with compressive symptoms or vascular complications—can be fatal, as seen in two of our cases. Early diagnosis through vigilant clinical assessment and appropriate use of imaging and biopsy techniques can markedly improve patient outcomes [4-6].

CONCLUSION

Primary thyroid lymphoma, though rare, should be considered in patients with rapidly enlarging goiters, particularly in those with a history of autoimmune thyroiditis. Prompt diagnosis using biopsy, followed by chemotherapy and radiotherapy, can achieve excellent outcomes, especially in early-stage disease. A multidisciplinary approach involving ENT surgeons,

radiologists, pathologists, and hematologists is vital for optimal patient care.

Ethical Statement:
Compliance with Ethical Standards: The study was conducted in compliance with ethical standards.

Funding: This research received no external funding.

Conflict of Interest: There are no conflicts of interest to declare related to this research.

Ethical Approval: While formal ethical approval was not obtained for this study, we ensured that all aspects of the research were conducted ethically and with respect for the rights and well-being of the participants.

Informed Consent: Informed consent was obtained from all participants involved in the study, and this information has been appropriately included in the manuscript.

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