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Systemic Amyloidosis Presenting as Adrenal Insufficiency and Multi-nodular Goiter

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Introduction:

Systemic Amyloidosis is a rare disorder in which misfolded proteins deposit in extracellular areas throughout the body. It can involve multiple organs, including endocrine glands. Due to the rare nature of the disease, it can be difficult to diagnose. We present a case of a 69-year-old male with severe orthostatic hypotension and nontoxic multinodular goiter who was ultimately diagnosed with systemic light chain amyloidosis

Case Presentation:

A 69-year-old male presented to the Endocrinology clinic after multiple hospital admissions for severe orthostatic hypotension, weight loss, edema and generalized weakness. On physical exam, he was found to have orthostatic hypotension and large goiter

Laboratory and Radiological Imaging:

Random Cortisol	6.9
Thyroid Function Test	Normal
ACTH Stimulation Test	Secondary Adrenal Insufficiency
Thyroid Ultrasound	Nontoxic multinodular goiter with 2 nodules greater than 5 cm in size (Figure 1 and 2)
FNA of 5.8 cm right thyroid nodule and 5.4 cm left thyroid nodule	Atypia of undetermined significance

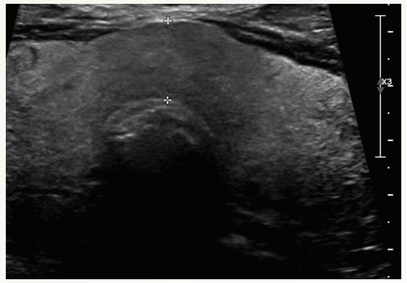

Figure 1. Thyroid ultrasound image with generalized thyromegaly and enlarged isthmus

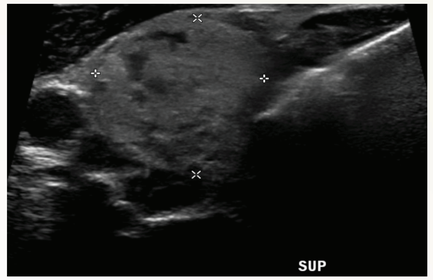

Figure 2. Superior right thyroid nodule

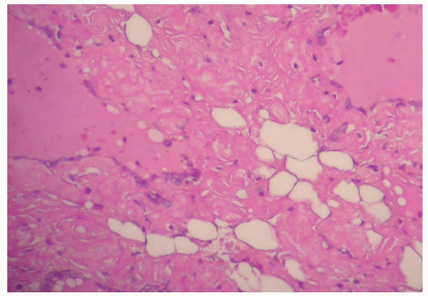

Figure 3. Representative histopathology of amyloid deposition in thyroid gland

Afirma genomic sequence classifier	"suspicious" for malignancy (risk of malignancy ~50%)
Thyroidectomy and Pathology	Benign thyroid parenchyma with dense light chain amyloid deposition

Further workup revealed systemic lambda light chain amyloidosis with renal involvement with nephrotic syndrome and cardiac involvement

Discussion:

The patient was treated with a bortezomib-based regimen for systematic amyloidosis, glucocorticoids for adrenal insufficiency, and levothyroxine for post-operative hypothyroidism. He had an excellent response to the chemotherapy and marked improvement in his symptoms

Conclusion:

There have been few reports of systemic amyloidosis resulting in secondary adrenal insufficiency. A review by Ozdemir found asymptomatic amyloid deposition in 30-80% of patients with systemic amyloidosis. Symptoms of amyloid endocrine involvement can include compressive symptoms and hormone deficiency, although patients with amyloid goiter are typically euthyroid. This case illustrates a rare presentation of systemic light chain amyloidosis, illustrating some of the ways that systemic amyloidosis can impact endocrine organs.

References:

1. Chavez CP, Morales Hernandez MDM, Kresak J, and Woodmansee WW. Evaluation of multinodular goiter leads to a diagnosis of AL amyloidosis. Thyroid Research. 2022;15:7.
2. Ozdemir D, Dagdelen S, Erbas T. Endocrine involvement in systemic amyloidosis. Endocrine Practice. 2010:16:1056-1063. Tsai K, Yu AC, Livhits MJ, Sajed D, Leung AM, Cheung DS.
3. Systemic light-chain amyloidosis incidentally diagnosed after subtotal parathyroidectomy and thyroid lobectomy. BMJ Case Rep. 2021;14:e241282