CLINICAL VIGNETTE

Myxedema: An Uncommon Diagnosis

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Case Presentation

A 56-year-old male presented to the Emergency Department with a 2-3 year history of behavioral changes, mild to moderate headaches, and fatigue. He reported these symptoms worsened in the past 2 months with onset of marked gait disturbance in past 2 days.

He had been seen at an outside urgent care clinic several days earlier and was noted to have high blood pressure and high cholesterol. His wife provided most of the current history and also reported 20-25 lb. weight gain in 2-3 months, cognitive difficulties, excessive sleeping, cold intolerance, and lack of libido. The patient denied any chest pain, dyspnea, nausea, vomiting, fevers, or abdominal pain. The patient was aware of why he was being evaluated, although he lacked insight into the severity of his symptoms.

On physical exam, the patient was afebrile with severely elevated BP of 183/134. The patient was in no acute distress; however, he had cognitive slowing with mild somnolence. He had mild periorbital edema as well as pretibial edema. His tongue was not grossly enlarged, but he had scanty hair on his limbs with dry, coarse skin and hair. His gait was broad based, and he was moderately ataxic.

His labs were most significant for a high TSH of 55.41 mIU/mL (0.55 - 4.78). Other results included creatinine at 1.3 mg/dL, sodium 132 mmol/L, and total cholesterol of 387 mg/dL LDL of 306 mg/dL. His CK level was slightly elevated at 544 U/L (40 – 280).

Discussion

Hypothyroidism is a relatively common disorder affecting approximately 4.6% of the general population.[1] With widespread availability of laboratory testing for TSH, severe hypothyroidism is a very uncommon presentation and even more so, myxedema coma.

Given the rarity of long-standing disease, its insidious nature and its widespread but chronic and varying effects on multiple organ systems, the diagnosis of myxedema can be difficult or delayed, but it must be considered in any patient with subacute or chronic behavioral or cognitive changes. The collateral history provided by a caregiver was essential in providing an accurate depiction of the acuity and chronology of the patient's symptoms given the patient's cognitive limitations.

In combination with physical exam findings, the diagnosis becomes rather apparent.

One of the hallmarks of myxedema is an associated dermopathy, thought in part due to an accumulation of glycosaminoglycan deposits in the dermis, although the exact pathophysiology is not entirely clear. [2,3] This can be expressed as puffiness of the face and hands, pretibial non-pitting edema, macroglossia, and coarse skin and hair. Without collateral history or images of a patient prior to disease onset and progression, these findings may not be marked and clinical hypothyroidism may go unrecognized. Comparing a photo of a patient's driver's license or ID card taken years prior may be clinically useful in unclear cases.

The most generalized manner in which thyroid deficiency affects the body is in the overall slowing of metabolic processes across different organ systems. The metabolic rate of the patient is slowed leading to weight gain, fatigue, cold intolerance, and in its extreme hypothermia. The patient may have slowed speech and cognition, as well as a delay in deep tendon reflexes. Bradycardia, dyspnea, constipation, decreased libido are among other varying manifestations. Less apparent but also common findings include anemia, renal dysfunction, hyponatremia, myopathy, and elevated cholesterol.

Thyroid hormone decreases systemic vascular resistance by dilating the resistance arterioles of the peripheral circulation; as such, thyroid deficiency often causes hypertension as resistance in the peripheral vasculature increases.[4] Diastolic relaxation is also impaired resulting in more apparent elevations in diastolic blood pressure. In conjunction with bradycardia, overall cardiac output is diminished; however because of lower oxygen demand and increased afterload due to vascular resistance, congestive heart failure is uncommonly seen.

Rarely, severe hypothyroidism presents as myxedema coma, better-termed myxedema crisis, a true medical emergency with a 25-60% mortality rate.[5] Frank coma is not required for this diagnosis; however, an acute alteration in mental status accompanied by hypothermia is the hallmark of the disease. Investigation of an acute precipitating factor such as an infection, myocardial infarction, GI bleeding, withdrawal of thyroid medication, or administration of certain medications such as opioids is indicated. Patients are critically ill, possibly hypotensive, bradycardic, hypoglycemic, and the profound

state of mental depression may be accompanied by centrally-mediated hypoventilation, requiring intubation.

Given its high mortality, when myxedema crisis is suspected, treatment, which includes intravenous thyroid therapy, should be started without delay. However, rapid administration of thyroid hormone can lead to atrial arrhythmias or even myocardial infarction. Stress steroids for possible concomitant adrenal insufficiency and empiric antibiotics should be strongly considered.

Clinical Course and Outcome

Our patient's overall clinical picture was most consistent with long-standing untreated hypothyroidism otherwise known as myxedema. His gait abnormalities, however, were not explained by thyroid disease. Given his elevated blood pressure and ataxia, an MRI showed T2 flair hyperintensities within the white matter thought to be related to long-standing untreated hypertension, although this was still unclear at follow-up. Of note, his MRI changes resolved on subsequent imaging.

The patient was admitted and his blood pressure was carefully monitored. He was additionally found to have renal artery stenosis contributing to his hypertension during his admission. The patient was diagnosed with severe thyroidism and not myxedema crisis, given his lack of hypothermia, bradycardia and hypotension.

It was thought that his mental status changes were more chronic, and his gait ataxia was more directly related to his uncontrolled hypertension and his MRI findings. The patient was started on oral levothyroxine and neither intravenous thyroid repletion nor empiric steroid therapy was warranted. The patient's overall condition improved after one week of therapy; he was discharged home with close follow-up. After several months of oral thyroid replacement, the patient's cognitive function dramatically improved, and his physical changes (e.g., puffy facies and coarse skin and hair) reverted to his previous appearance.

References:

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