

CASE REPORT

Myxedema Coma with Reversible Cardiopulmonary Failure: a Rare Entity in 21st Century

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ABSTRACT

Myxedema coma, a rare entity in 21st century in developed nations, is a decompensated phase of hypothyroidism with high mortality rates. We describe a young woman with myxedema, who developed respiratory failure, congestive heart failure and significant pericardial effusion, some of the uncommon manifestations. Decreased cardiac contractility can result in cardiomyopathy and heart failure. As illustrated by this case, myxedema can also result in significant pericardial effusion due to increased vascular permeability. Myxedema can further be complicated by alveolar hypoventilation and respiratory failure secondary to the lack of central drive as well as respiratory muscle weakness. Prompt therapy with thyroid hormone replacement, glucocorticoid therapy, aggressive supportive care and management of the precipitating event can save lives and reverse the cardiopulmonary symptoms, as in our patient. Hence, physicians should have a high index of suspicion for myxedema coma in patients with unexplained cardiopulmonary failure. Our report is, therefore, aimed at bringing awareness about the rare but fatal manifestations of myxedema coma.

INTRODUCTION

Myxedema coma is a severe and life threatening form of decompensated hypothyroidism. It presents with multiple organ dysfunction and progressive mental deterioration (1). The term is largely a misnomer because quite a few patients are ob-

tunded, rather than truly comatose (2,3). Because of its insidious onset and non-specific symptoms, diagnosis can be difficult at times. Altered mental status, impaired thermoregulation and the presence of a precipitating event are the key elements used to diagnose myxedema coma (4). The mortality rates may be as high as 30–60%, particularly with delayed treatment (1). Here, we describe a relatively

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young woman with myxedema coma, who developed respiratory failure and congestive heart failure, two of the uncommon manifestations.

Case Report

A 44-year-old obese woman presented to the emergency department with a 3-day history of progressively worsening dyspnea. She complained of decreased oral intake, hair loss, menstrual irregularities and weakness for the last 4 months. In addition, she had abdominal distension, and lower extremity swelling. Past medical history was significant for panic attacks. On examination, she had a blood pressure of 153/86 mmHg, heart rate of 65/min, respiratory rate of 12/min, temperature of 98.2°F and oxygen saturation of 98% on ambient air. She was also found to have anasarca, abdominal distension, left pleural effusion and proximal myopathy.

Laboratory investigations revealed TSH of 46 μ IU/mL, free and total T4 of 0.0 μ g/dL and T3 of 37 ng/dL. Brain natriuretic peptide was 541 pg/mL. Other tests revealed sodium of 137 mmol/L, potassium of 4.1 mmol/L, blood urea nitrogen of 19 mg/dL, creatinine of 0.94 mg/dL, chloride of 100 mmol/L, bicarbonate of 27 mmol/L, calcium of 9.2 mg/dL, glucose of 70 mg/dL, and normal complete blood count. Anti-thyroid peroxidase antibody was negative. Cardiac enzymes, EKG and venous duplex of bilateral lower extremities were normal. Echocardiogram showed ejection fraction (EF) of 25% with moderate diffuse hypokinesia; peak systolic pulmonary artery pressure of 50-60 mmHg and mild pericardial effusion.

With a diagnosis of myxedema, the patient was started on oral levothyroxine 25 mcg daily. On day 2, she became comatose with a Glasgow coma score of 9 (E2 V3 M4) and went into respiratory failure. Arterial blood gases revealed pH of 7.03, PaO₂ of 60 mmHg, PCO₂ of 126 mmHg and bicarbonate of 33 mmol/L. She was intubated and parenteral L-thyronine 10 mcg and hydrocortisone 100 mg were administered. There was improvement of clinical status and she was extubated the next day. But she still remained dyspneic and developed progressive anasarca over the next 2 weeks despite aggressive diuresis. A repeat echocardiogram showed worsening of pericardial effusion without features of cardiac tamponade and EF of 10%. A pericardial window was performed with drainage of 350 mL of transudative fluid

immediately and an additional 750mL over next 3 days. Bacterial and fungal cultures of pericardial fluid, AFB stain and cytology were negative. An echocardiogram five days later showed EF of 40-45 % with small pericardial effusion. Thereafter, the patient was continued on oral levothyroxine, and she demonstrated significant improvement over next two months.



DISCUSSION

Myxedema coma can affect virtually any organ system of our body, and although uncommon, may lead to multi-organ failure (5). It occurs almost exclusively in persons 60 years and older, and most of them are female (6). Our patient presented with typical features of hypothyroidism, however, elderly patients may have nonspecific and atypical symptoms such as decreased mobility (7). A precipitating event in the background of long-standing, untreated or inadequately treated hypothyroidism may result in myxedema coma. Infections, drugs such as narcotics, anesthetics, and sedatives, lung diseases, congestive heart failure and gastrointestinal bleeding may be some of the precipitating events (8). In our case, no precipitating factors could be identified.

The cardiovascular manifestations in myxedema coma tend to be especially severe and life threatening in comparison to other symptoms (9). Early symptoms of the disease comprise bradycardia and low cardiac output due to decreased cardiac contractility. Frank congestive heart failure is a rare occurrence (10). Diastolic hypertension has been observed in early stages due to peripheral vasoconstriction and central shunting (8). However, as precipitating event disrupts homeostasis maintained in these cases, there may be hypotension and shock later, exacerbated by reduction in blood volume, bradycardia and decreased cardiac output (2,11). Pericardial effusion may also be present in such cases due to increased vascular permeability (12). Actually, hypothyroidism should be ruled out in all patients with unexplained pericardial effusion as it is an important cause of moderate to severe pericardial effusion (13). It can also lead to cardiac tamponade, although it is rare (13). After diagnosis with echocardiography, early pericardiocentesis should be done to relieve mechanical constriction in such cases (14). There was a significant

improvement in clinical symptoms and EF, as in our patient after the drainage of pericardial fluid.

Myxedema can also be complicated by alveolar hypoventilation and respiratory failure. Alveolar hypoventilation occurs due to decreased ventilatory response to hypoxia and hypercapnia (15,16). There is weakness of respiratory muscles as well as a decrease in the central drive (15,17). In addition, myxedema of tongue and pharynx, obesity, obstructive sleep apnea, any superimposing pneumonia, pericardial and pleural effusions, and ascites can contribute to the respiratory failure (17-19). These respiratory abnormalities respond promptly to aggressive thyroid hormone replacement, as highlighted in this case report (16,20-22). Need for mechanical ventilation is one of the predictors of outcome in such cases (23).

The diagnosis of myxedema coma is primarily clinical with supportive evidence from thyroid function tests (24). Our patient had low levels of T3 and T4 and high TSH levels, along with classical features of myxedema coma. With typical clinical features, laboratory diagnosis should not delay the initiation of treatment (25). The approach to treatment consists of a) thyroid hormone replacement therapy; b) cardiovascular, pulmonary and neurologic support; and c) management of precipitating events (24). However, the drug type, dose, frequency, and mode of administration for thyroid hormone replacement is controversial (26). The optimal treatment is still uncertain because of the rarity of cases and difficulties with performing controlled trials (26,27). Glucocorticoid therapy is advocated in all patients because hypoadrenalism may co-exist with hypothyroidism, and treatment of hypothyroidism alone may precipitate adrenal insufficiency (25). Moreover, cardiovascular and pul-

monary monitoring in intensive care unit with ventilatory support is often needed (1). Our patient was administered parenteral L-thyroxine and glucocorticoid therapy initially. She also required mechanical ventilation, pericardial window for severe pericardial effusion, and other supportive care. After she stabilized few days later, she was maintained on oral levothyroxine.

Hypotension, bradycardia at presentation, need for ventilator support, hypothermia unresponsive to treatment, sepsis, intake of sedative drugs, lower Glasgow coma score, and high APACHE II scores are some of the predictors of mortality in myxedema coma (23). Mortality is between 25% and 52%, with sepsis being the predominant cause of death (23, 28). Successful treatment depends on clinical suspicion, early diagnosis, prompt thyroid hormone replacement and adequate supportive measures (11). □

CONCLUSION

Myxedema coma is a decompensated phase of hypothyroidism with high mortality rates. It usually presents with altered mental status and hypothermia precipitated by an event. Early identification and treatment of hypothyroidism have fortunately made severe cardiopulmonary complications rare in 21st century. Myxedema should be considered in patients with unexplained cardiopulmonary failure. Such cases require prompt therapy with thyroid hormone replacement, glucocorticoid therapy, aggressive supportive care and management of the precipitating event.

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