Thyroidectomy in a Patient with Thyroid Storm, Severe Dyspnea, and Cardiac Dysfunction

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ABSTRACT

Thyroid storm (TS) is an endocrine emergency demanding prompt and intensive treatment. Several risk factors are involved in precipitating TS and should be prevented in patients with thyrotoxicosis. Withdrawal of antithyroid drugs is probably the main risk factor for TS. Treatment with oral or intravenous drugs (antithyroid agents, steroids, and iodine) is required. Other measures are needed, such as plasmapheresis, diuretics, supportive care, and management of systemic complications such as cardiorespiratory failure. Finally, precipitating factors must be controlled. Nevertheless, some patients do not respond to therapy and their lives further deteriorate, creating circumstances in which thyroidectomy should be considered. We report the case of a 42-year-old woman with a previous history of thyrotoxicosis, diagnosed 18 years earlier, who was noncompliant with her therapy. Her medication was stopped before she came to our emergency unit with multinodular goiter, severe dyspnea, edema, tachycardia, and malnutrition. Thyroid storm was diagnosed, and she was transferred to the intensive care unit. She was treated with intensive antithyroid drugs, glucocorticoids, iodine solution, a β-blocker drug, and support measures. Due to rapid deterioration of her health in the absence of a reduction in thyroid hormone response, total thyroidectomy was decided. After surgery, the patient was well, with recovery of respiratory function and reduction in thyroid hormone levels. She was discharged with normal parameters. Consequently, if patients with thyroid storm continue to deteriorate despite medical treatment, thyroidectomy should be considered at centers with the necessary expertise.

Keywords: Cardiopulmonary failure, Thyroid storm, Thyrotoxicosis, Total thyroidectomy.


BACKGROUND

The occurrence of thyroid storm (TS) in a patient is a severe, life-threatening condition with an annual incidence of two patients per 1,000,000 and a mortality rate of 11% in patients with thyrotoxicosis.¹ Most sources report that TS accounts for 1–2% of hospital admissions for thyrotoxicosis. The incidence of TS in hospitalized patients has been estimated at 0.2 per 100,000 per year, and 5.4% of hospitalized patients with thyrotoxicosis.¹ ²

Consensus exists that therapy should be started as soon as the diagnosis is suspected, to target the synthesis and release of thyroid hormones, and minimize the systemic effect of circulating hormones. Although surgery is an option in hyperthyroidism, the indication for this intervention in TS is still under discussion.

Patients with TS should receive prompt and vigorous treatment in an intensive care unit. We report the case of a woman with a hyperfunctioning thyroid nodule, history of noncompliance with antithyroid therapy, and drug failure. She presented severe TS with respiratory and cardiovascular failure but was managed successfully with total thyroidectomy.

CASE DESCRIPTION

Our patient was a 42-year-old woman with a history of arterial hypertension and smoking, which was diagnosed 18 years earlier of hyperfunctioning thyroid nodule by her primary care physician. Her adherence to the prescribed antithyroid treatment during the following years was deficient.

She stopped taking treatment on her own volition 2 years before this recent visit. She was transferred to the emergency unit of our 12 de Octubre Hospital severe anasarca and intense asthenia. On physical examination, her body temperature was 38.7°C, and she had tachycardia (140 bpm), tachypnea (25 rpm), goiter grade III, dyspnea at rest, severe lower limb edema, blood pressure 170/90 mm Hg, and signs of malnutrition (Fig. 1). The chest radiograph showed bilateral inferior lung edema and pericardial effusion.

Laboratory Analyses

Blood analysis showed suppressed thyroid-stimulating hormone (TSH) <0.005 μIU/mL (normal range: 0.57–5.92 μIU/mL), elevated free T4 7.3 ng/dL (normal range: 0.72–2 ng/mL), glycemia 221 mg/dL, and hyperbilirubinemia 4.2 mg/dL (normal range <2 mg/dL). There were findings indicative of malnutrition (total proteins 7.6 g/dL and albumin 2.7 g/dL), thrombocytopenia (109,000 L), and a tendency to coagulopathy (prothrombin time test 48%, partial thromboplastin time test 31 seconds, and international normalized ratio 1.61). Viral hepatitis serology and autoimmune antibodies were negative. She was diagnosed of TS. Using the Burch and Wartofsky criteria, she had a score of 60 points out of a total 100 points.³
Thyroid Storm and Total Thyroidectomy

She was admitted to the intensive care unit with the diagnosis of TS, where vigorous medical therapy was initiated with methimazole, potassium iodide, glucocorticoids, β-blockers (esmolol and propranolol), and diuretics. She also underwent plasma exchange to remove thyroid hormones from plasma. Cholestyramine was given as adjunctive therapy to reduce thyroid hormone by interfering with the enterohepatic circulation and increasing fecal excretion, without effect. Abdominal ultrasound revealed severe ascites, without other findings. One liter of fluid was removed by paracentesis.

Two days after admission, she continued to develop progressive respiratory distress, shock, and altered consciousness, with increasing bilateral pleural effusion, leading to great physical deterioration. Orotracheal intubation was performed.

Computed tomography showed a large multinodular goiter with a hypodense cystic nodule (8 × 5.9 × 8 cm) in the lower right thyroid lobe that displaces and compresses the trachea and intrathoracic extension. Cardiac and pleural effusion, free peritoneal fluid, and a congestive liver were confirmed.

Fine needle aspiration biopsy on the thyroid nodule yielded 120 mL of serohematic fluid. The histology showed hyperfunctional hyperplasia without malignancy signs. In about 24–48 hours, the cystic nodule was refilled.

Given the resistance of thyroid hyperfunction to methimazole and other adjuvant treatment, increased progressive respiratory distress, and rapid deterioration in the patient’s conditions, surgery was recommended by the team.

Total thyroidectomy of a multinodular goiter with intrathoracic expansion was performed (Fig. 2). Histopathology confirmed the diagnosis of multinodular toxic goiter. Total thyroidectomy was unaccompanied by hypocalcemia or vocal hoarseness. Replacement treatment with levothyroxine was initiated. Four months after thyroidectomy, the patient was well, and thyroid hormone levels and liver function tests were within normal range.

**DISCUSSION**

Thyroid storm has a high mortality rate and requires urgent treatment. Our patient had a long history of inadequate use of antithyroid drugs, completely omitting them in the last 2 years. Therapeutic noncompliance is the most common trigger factor implicated in the literature in decompensated thyrotoxicosis.4

Although she was hyperglycemic in the emergency department, she had no history of diabetes. At discharge, with acceptable thyroid control, her glucose homeostasis was normal. Among the other factors that can precipitate TS, the most frequent are surgery, trauma, infection, acute iodine load, or childbirth.3

The pathophysiology of TS is not clearly known. Patients with TS typically have exaggeration of the usual symptoms of hyperthyroidism. A rapid rate of increase in thyroid hormone and enhanced free thyroid hormone binding to receptors, possibly combined with heightened response, have been postulated.6 Reduced thyroid hormone-binding proteins could also contribute to increasing free thyroid hormone concentrations in blood.7

A single sensitive TSH assay and free T4 are recommended for the initial evaluation of thyroid function.8 Keep in mind that some drugs can directly affect thyroid function studies: glucocorticoids and dopamine inhibitors can lower TSH levels. Seventeen percent of hospitalized patients with non-thyroid illness have been found to have lower TSH levels, and 85% of these patients have normal TSH levels after follow-up.9 Our patient had almost undetectable TSH values, but they normalized after surgery and levothyroxine replacement.

The diagnosis of TS can be supported using the quantitative diagnostic score proposed by Burch and Wartofsky.3 Our patient’s score was 50, which exceeds the score of 45 that suggests TS on this scale. This diagnostic criterion considers the presence of fever, central nervous system (CNS), gastrointestinal, liver, cardiovascular, and precipitating factors and does not require abnormal thyroid hormone values. Akamizu et al. in 2012, in a Japanese survey, developed a diagnostic criterion consisting of elevated serum thyroid hormone levels and other CNS, CHF, and gastrointestinal/hepatic manifestations.1 Our case also met this criterion (low TSH plus elevated free T4, fever, and cardiac failure) for the diagnosis of TS. Both criteria are helpful for supporting the diagnosis, but are not decisive, as they have not been validated in prospective studies.

Our patient had elevated conjugated bilirubin values, without clear jaundice, which has been reported in 18% of patients with TS.10 However, the liver biopsy in some patients is normal. Serum bilirubin levels in our patient normalized after surgery achieved euthyroidism. Unconjugated bilirubin levels were not elevated, which rules out Gilbert syndrome.
The therapy of patients with TS should be aggressive and initiated as soon as the diagnosis is suspected. The mortality associated with TS is as high as 37.5%, although recent reports indicate a lower rate, 10.7%, probably due to the prompt use of effective therapeutic strategies. The combination of antithyroid drugs, inorganic iodide, and corticosteroids is considered the first-line approach to this disease. The American Thyroid Association (ATA) recommends using propylthiouracil because it can block the conversion of T4 to T3 in addition to inhibiting new hormone synthesis. However, recent studies do not confirm this nor do they report significant differences in mortality or disease severity between patients treated with methimazole or propylthiouracil. In some critically ill patients, like ours, initial parenteral administration can be preferable to the oral route.

The rate of remission in patients with nodular disease treated with antithyroid drugs is lower, and discontinuation of treatment is often followed by relapse. Our patient did not accept lifelong antithyroid treatment.

Patients with hyperthyroidism who are unresponsive to aggressive medical therapy with methimazole and corticosteroids should undergo thyroidectomy as recommended in ATA guidelines, especially if they experience excessive deterioration of health and uncontrolled thyroid function. Our patient’s worsening respiratory status and congestive heart disease despite intensive medical therapy made us consider surgery as a way to rapidly resolve TS.

After discussion of the case, total thyroidectomy was chosen as treatment in this case. Recurrences can be avoided and the rate of complications, in the hand of expert surgeons, is the same as with subtotal thyroidectomy. Radioactive iodine is an alternative for toxic nodular goiter, but not for TS, because retreatment has 20% risk. Euthyroidism is not immediately achieved as with surgery, and the prevalence of hypothyroidism can reach 64% at 24 years post-surgery.

Scholz et al., retrospectively analyzed the outcome of early thyroidectomy in 10 older critically ill patients with TS, thyroid autonomy, and severe cardiorespiratory and renal failure with cardiac arrhythmia, coronary artery or chronic obstructive pulmonary disease, or acute inflammation. Late postoperative mortality in these patients was 20% in contrast to previous reports of 43%. Scholz et al. suggested that total thyroidectomy is a treatment of choice in older chronically ill patients with TS associated with cardiorespiratory and renal failure.

More recently, Chiha et al. classified when patients require an urgent surgical intervention into three categories: (1) patients who clinically deteriorate or do not improve within 24–48 hours, despite intensive medical treatment; (2) patients who develop side effects from treatment; and (3) patients who need prompt resolution of hyperthyroidism due to severe underlying cardiac or pulmonary comorbidities. Our patient continued to have life-threatening deterioration in cardiovascular and lung functions after intensive drug therapy.

**Conclusion**

In conclusion, early thyroidectomy should be considered in patients with TS, organ failure, and/or non-compliance with medical therapy due to the high morbidity and mortality of this condition. A multidisciplinary team in an intensive care unit should evaluate candidates for this approach.

**Informed Consent**

Written informed consent was obtained from the patient for publication of this manuscript and any accompanying images.

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**References**

