American Journal of Surgery Case Reports

Case Report

Novel Surgical Management of Grave's Disease in the Setting of Thyroid Storm

Jordan West[1], Antoinette Esce[2], Ivan Pinon[3], Niels Chapman4, Garth Olson[2] and Nathan Boyd[2]*

[1]School of Medicine, University of New Mexico, USA

[2]Department of Surgery, University of New Mexico School of Medicine, USA

[3]Department of Endocrinology, University of New Mexico School of Medicine, USA

[4]Department of Anesthesia, University of New Mexico School of Medicine, USA

Abstract

Case presentation: A 34-year-old male was diagnosed with Graves' disease and medically managed. Despite aggressive medical management, the patient did not achieve euthyroid status and removal of the thyroid gland became necessary.

Importance: Thyroid storm, the most severe form of thyrotoxicosis, is a rare and life-threatening complication of Graves' disease. Thyroid storm may be caused or worsened by manipulation of the thyroid gland during surgery. Surgical management is typically reserved for patients who have achieved a euthyroid state. However, it is occasionally necessary to perform surgery on a patient who is actively hyperthyroid. Minimizing the risk of thyroid storm is essential in this setting.

Observations: A standard thyroidectomy involves retraction of the thyroid while the major vascular attachments to the gland are still intact. In this case, a novel technique for thyroidectomy was used to minimize manipulation of the gland until its primary vascular attachments were ligated. These modifications included:

1. An extended approach to the central neck which allowed for ligation of the superior and middle thyroid vasculature from an exposure lateral to the strap muscles.
2. Ligation of the sternothyroid muscles bilaterally
3. Primary ligation of the inferior thyroid vasculature bilaterally
4. Manipulation of the gland only after all of the major, named thyroid vasculature was ligated.

Conclusion: Graves' disease can be a life-threatening diagnosis. When medical management fails, surgical management must occasionally be considered even in an actively hyperthyroid patient. Several specific surgical maneuvers may be helpful in minimizing thyroid storm.

Keywords: Thyroid; Hyperthyroidism; Graves; Thyroidectomy; Thyroid Storm; Surgery

Introduction

Graves' disease is an autoimmune disorder in which thyrotropin-receptor antibodies induce overproduction of thyroid hormone [1]. The annual incidence of Graves' related hyperthyroidism is 20-50 cases per 100,000 persons [1]. Thyroid storm, the most severe form of thyrotoxicosis, is a rare and life-threatening complication of Graves' disease. Thyrotoxicosis can also be seen in patients with subacute thyroiditis, toxic multinodular goiter, and toxic adenoma, but it is most commonly seen with Graves' disease. In the United States, thyroid storm is estimated to occur in about 1.2% of patients with some form of hyperthyroidism, with about half of these patients being symptomatic [2]. Thyroid storm may be triggered by acute stress, such as trauma, surgery, or infection, or when a patient's hyperthyroidism is untreated or undertreated [3]. Manipulation of the thyroid gland during surgery can cause a release of thyroid hormone and exacerbate thyrotoxicosis or cause thyroid storm. For this reason, surgical management is typically reserved for patients who have achieved a euthyroid state. However, it is occasionally necessary to perform surgery on a patient who is actively hyperthyroid. Minimizing the risk of thyroid storm is essential in this setting.

Citation: West J, Esce A, Pinon I, Chapman N, Olson G, Boyd N. Novel Surgical Management of Grave's Disease in the Setting of Thyroid Storm. Am J Surg Case Rep. 2023;4(5):1074.

Publisher Name: Medtext Publications LLC

Manuscript compiled: Jun 14th, 2023

***Corresponding author:** Jordan West, University of New Mexico, School of Medicine, MSC08 4720 1 UNM, Albuquerque, NM 87131-0001, USA, Tel: +1-505-414-5393

Case Presentation

A 34-year-old male with a history of tobacco use disorder presented to a community endocrinologist with heat intolerance, tremors, and intermittent tachycardia to 180 beats per minute, muscle weakness, anxiety, and nervousness. Initial laboratory studies showed a Thyroid Stimulating Hormone (TSH) of 0.007 and a Free T4 of 8.0. An ultrasound of the thyroid demonstrated an enlarged, hyperemic gland consistent with Graves' disease, as well as a solitary thyroid nodule. The patient was initially medically managed with methimazole and propranolol. Unfortunately, he had an anaphylactic reaction to this treatment regimen. He was started on several other anti-thyroid medications, but none of them were effective in treating his thyroid symptoms. After four months on various medications, he remained hyperthyroid and surgical management was recommended. He was scheduled for surgery by an outside institution and given Super Saturated Potassium Iodine (SSKI) for one week prior. His surgery was then canceled due to insurance issues. One month later,

the patient presented to an endocrinologist at our institution with ongoing tachycardia, heat intolerance, muscle weakness, anxiety, nervousness, and visible bilateral hand tremors. Labs demonstrated a TSH of 0.007, a Free T4 >8.0, and a TSH receptor antibody >40.00. The patient was sent to the emergency department for management of severe acute thyrotoxicosis.

On admission to the hospital, the patient's Burch-Wartofsky Score was 25, indicating an impending thyroid storm. After eight days of changing and titrating his anti-thyroid and beta-blocking medications to no avail, the decision was made to initiate plasmapheresis. Despite every effort, the patient remained floridly hyperthyroid on the day of surgery, with a free T4 of 5.9. A novel surgical technique was employed to ligate as much of the thyroid vasculature as possible before manipulating the thyroid gland in order to avoid a surge of thyroid hormone in the blood.

Our technique prioritized control of the main thyroid vasculature before gland manipulation by creating excellent exposure lateral to the strap musculature and optimizing access to the central compartment from this approach. First, a pocket was created lateral to the strap muscles and medial to the sternocleidomastoid muscle to provide visualization of the superior pole and middle thyroid vessels and allow them to be ligated without any retraction of the gland. This maneuver was performed on both sides. The strap muscles were then divided in the midline as with a typical thyroidectomy. The sternothyroid muscle was also ligated to improve exposure and the large veins and arteries associated with the cricothyroid membrane and Delphic lymph nodes were carefully ligated, with minimal gland retraction. Finally, the inferior pole vessels were exposed and ligated. In this case, the gland was so hypertrophic that it extended substernally. Kittners were used to apply precise, gentle pressure on the gland to expose and ligate the inferior pole vessels without injuring the recurrent laryngeal nerve or parathyroid glands. At this point, the only remaining thyroid vasculature was assumed to be arising from tracheal perforators and from the region near Berry's ligament. The recurrent laryngeal nerve was exposed and traced to the cricothyroid joint, and the gland mobilized off the trachea. The patient's vital signs and labs were carefully monitored during surgery. At the start of the case, the patient's total T3 was 497 and remained between 427 and 520 for the duration of the procedure. Two hours after surgery, it was 342. During his intraoperative and postoperative course, he remained hemodynamically stable.

In the days following the surgery, the patient's free T4 dropped from 4.0 to 2.1. He was discharged on post-operative day two. At his follow up exam, eleven days after surgery, his only symptom was fatigue. His TSH was <0.007 and Free T4 was 0.5. He was started on weight-based levothyroxine and the dose titrated. Four months after the surgery, he was symptom free, and his lab work was within normal limits.

Discussion

Thyroid storm is a dangerous condition that should be prevented when possible and treated expeditiously when unavoidable. Up to half of patients with thyroid storm can die, and those who survive are at risk for end organ damage and central nervous system dysfunction if not managed properly [2]. Anti-thyroid medication is considered to be first line treatment, while radioactive iodine ablation and surgical management with thyroidectomy are typically reserved for patients who are euthyroid on anti-thyroid medications. Sometimes, anti-thyroid medications are ineffective or poorly tolerated due to side effects [4]. The most severe side effects of methimazole are agranulocytosis and pancytopenia. Less severe side effects include pruritus, allergic dermatitis, nausea, and dyspepsia. Our patient may have experienced these when he was first treated with Methimazole, and other medications proved ineffective. Additionally, our patient did not respond to second line therapy for Graves' disease, plasmapheresis, leaving surgery as his only viable option.

Surgery is typically avoided in patients who are actively hyperthyroid, out of fear that manipulation of the thyroid gland might induce thyroid storm [5]. In this case, surgical management was ultimately unavoidable, and great care was taken during surgery to prevent an acute release of thyroid hormone into the blood by minimizing gland manipulation until all of the major blood vessels feeding the gland were ligated. During a standard thyroidectomy, the major, named thyroid vessels are identified and ligated in a superior to inferior fashion, medially to the strap musculature, which requires significant retraction and handling of the gland for exposure. However, in this case, our technique prioritized control of the main thyroid vasculature before gland manipulation. This was done through an extended access approach to the central neck which allowed for ligation of the superior and middle thyroid vasculature lateral to the strap muscles. And most importantly, handling of the thyroid gland was performed only after all of the named thyroid vasculature was ligated.

From an anesthesia perspective, the primary concern in the management of thyrotoxicosis is focused on cardiovascular status and corresponding physiological reserve. The actual measured hormone levels, the normalization of which would ordinarily be desirable, are less relevant compared to the degree of myocardial compromise and physiological reserve. In this case, with medical management being only partially effective, but echocardiographic data confirming good preservation of cardiac function with an Ejection Fraction (EF) of greater than 50%, prompt definitive management was also a priority from the anesthesiology point of view. With an otherwise benign social and medical history, and an uncompromised airway examination, the anesthetic management included synchronous IV induction of anesthesia and establishment of invasive monitoring. The patient tolerated standard doses of propofol. This was followed by protocolized video laryngoscopy placement of a Nerve Intactness Monitoring (NIM) endotracheal tube facilitated by low dose rocuronium neuromuscular blockade and maintenance of anesthesia using sevoflurane and remifentanil. Intravenous dexamethasone was administered on induction; hemodynamic spikes observed intraoperatively were blunted using esmolol boluses. Only two such doses were administered, the first as an element of the induction of anesthesia, the second, 20 mg, during mobilization of the thyroid gland after near-complete separation of superior pole vessels. Hydrocortisone, 100 mg, was injected empirically following complete devascularization of the thyroid. The patient did not require any vasopressors or inotropes throughout the procedure and the T3 level did not markedly increase during surgery. Emergence from anesthesia and extubation were uneventful.

The endocrinology observations in this case are numerous. For patients who remain hyperthyroid despite maximal medical management or intolerance to anti-thyroid medications, super saturated potassium iodide is given prior to surgery, which induces the Wolff Chaikoff effect. The Wolff-Chaikoff effect occurs when the body stops production of thyroid hormone in the setting of high iodide,

simply by inhibiting enzymatic conversion of iodide to thyroxine and diiodotyrosine [6]. This process acts as a regulator of thyroid hormone production in the setting of high inorganic iodide and is leveraged preoperatively to medically optimize patients for surgery. However, this effect is somewhat transient. If the body is continually exposed to increased iodide, an escape from the Wolff-Chaikoff effect takes place and hormonal biosynthesis is resumed. If high iodide levels persist, this could lead to a hyperthyroid state, as more substrate would be available for conversion to active thyroid hormone.

Our patient received SSKI in anticipation of his first surgery, and then his case was cancelled. When this patient was admitted, he was one month post SSKI administration and still profoundly hyperthyroid. It is unclear if the effect had worn off, leaving him in his baseline hyperthyroid state, or if he had escaped the Wolff-Chaikoff effect while still exposed to the SSKI in his system, further exacerbating his hyperthyroidism. Typically, an iodine load in a euthyroid patient will produce two to three weeks of hypothyroidism, followed by a return to normal function. However, there is still a question about the duration of the effect in a hyperthyroid patient.

Conclusion

This case was unique and particularly challenging for several reasons. The patient did not reach a euthyroid state prior to surgery, despite multiple attempts at medical management. This was complicated by the patient not tolerating anti-thyroid medication, specifically methimazole. It is unclear if the symptoms he experienced after methimazole administration were from a true allergy, or if they were just an adverse effect of the medication. Additionally, it is unclear if he was experiencing the escape from the Wolff-Chaikoff effect when he presented to our hospital.

Fortunately, surgical management of this patient's disease was successful despite his active hyperthyroidism. The careful dissection and ligation of thyroid vessels prior to gland manipulation seemed effective at mitigating potential thyroid storm and subsequent hemodynamic instability, based on vitals, laboratory values, and the patient's clinical course. Graves' disease is difficult to manage and can have dangerous consequences. However, with additional study on safe surgical techniques in hyperthyroid patients, surgery might be considered earlier in disease progression, as opposed to remaining a last resort.

References

1. Smith TJ, Hegedüs L. Graves' disease. N Engl J Med. 2016;375(16):1552-65.
2. Devereaux D, Tewelde SZ. Hyperthyroidism and thyrotoxicosis. Emerg Med Clin North Am. 2014;32(2):277-92.
3. Spitzweg C, Reincke M, Gärtner R. [Thyroid emergencies: Thyroid storm and myxedema coma]. Internist (Berl). 2017;58(10):1011-19.
4. Fiztgerald P. Hyperthyroidism (Thyrotoxicosis). Current Medical Diagnosis & Treatment 2023. 62nd Ed. New York: McGraw-Hill; 2023.
5. Chiha M, Samarasinghe S, Kabaker AS. Thyroid storm: an updated review. J Intensive Care Med. 2015;30(3):131-40.
6. Markou K, Georgopoulos N, Kyriazopoulou V, Vagenakis AG. Iodine-induced hypothyroidism. Thyroid. 2001;11(5):501-10.