

Myasthenic Crisis Manifesting as Postoperative Respiratory Failure following Resection of Unsuspected Intrathoracic Thymic T-Cell Lymphoma during Thyroidectomy for an Adjacent Large Retrosternal Goiter

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What Is Known about This Topic

- There has been one previous case report on the coexistence of thymoma and intrathoracic goiter with an uncomplicated postoperative course. Myasthenic crisis occurs postoperatively in one third of thymoma patients.

What This Case Report Adds

- The presence of thymoma in this patient's large intrathoracic goiter was recognized only postoperatively upon the development of myasthenic crisis. Preoperative symptoms of fatigue and dysphagia and application of a predictive score for myasthenic crisis might have alerted the treating physician at an earlier stage.

Key Words

Thymoma · Goiter · Mediastinal neoplasms · Substernal goiter · Myasthenia crisis

Abstract

A middle-aged female with a goiter of 10 years' duration presented with progressive pressure symptoms, nocturnal

choking and dyspnea on exertion for 5 months. Physical examination demonstrated a large simple multinodular goiter. Imaging revealed a deep retrosternal goiter extending below the tracheal bifurcation with marked tracheal deviation. Total thyroidectomy was carried out via a cervical approach and a median sternotomy. Extubation was not possible, and the patient had to be kept intubated. She then went into a myasthenic crisis. Initial ventilatory support was followed by

intravenous immunoglobulin, steroids and pyridostigmine. The patient had complete remission and was asymptomatic 18 months later. Histopathology showed a T-cell-rich thymoma in addition to a nodular colloid goiter.

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Introduction

Goiter is endemic in many parts of the Sudan and thyroidectomy is a common elective surgical procedure in Khartoum [1]. The presence of a retrosternal opacity on a chest X-ray with a cervical swelling always points to a retrosternal extension of the thyroid. Thyroidectomy via a cervical incision is often possible, however in a deep-seated goiter below the tracheal bifurcation, median sternotomy may be resorted to [2]. The coexistence of a retrosternal goiter and thymoma is an interesting combination that has not been reported before. We present a case in whom a thymoma associated with retrosternal goiter was initially missed and was diagnosed only when the patient went into a myasthenic crisis from which she recovered after treatment and remained well 18 months postoperatively.

Case Report

A 32-year-old Sudanese female presented with a goiter of 10 years' duration. She had experienced pressure symptoms for the last 5 months with nocturnal choking, shortness of breath on lying flat and had to use two pillows while sleeping. She also had fatigue towards the end of the day along with progressive dysphagia for solids. Upper gastrointestinal endoscopy was normal. There was some degree of ptosis that was detected retrospectively after the final diagnosis was made. General examination was unremarkable. Cervical examination showed a simple multinodular goiter measuring 14 × 10 × 8 cm with tracheal deviation to the left and dull percussion note over the upper sternum indicating retrosternal extension. Upper gastrointestinal endoscopy was normal. Investigations showed a normal blood picture, Hb 13 g/dl, total WBC 6,000 cells/mm³, creatinine 1.0 mg/dl, Na 139 mmol/l, K 4 mmol/l, thyroid hormones T₃ 95 nmol/l (normal range: 80–220), T₄ 9 nmol/l (normal range: 4.5–12.5), and TSH 2.5 mU/l (normal range: 0.3–3.3). Plain chest X-ray showed a deep retrosternal mass compressing the trachea and with marked left side deviation (fig. 1). CT scan showed a homogeneous anterior mediastinal mass with distinct outlines and no calcification or contrast enhancement which could be due to a deep retrosternal extension of a goiter below the tracheal bifurcation or a lymphoma (fig. 2).

The patient had total thyroidectomy via both cervical collar incision and a median sternotomy. We started by a cervical approach; the superior pole of the thyroid was ligated and transected. A trial of cervical delivery by finger sweeping in the line of cleavage and gentle pulling was attempted on the left lobe but was abandoned because the lower end had a broad base and was adherent to the



Fig. 1. Tracheal narrowing due to side-to-side compression and deep retrosternal extension of a goiter and presence of a thymoma.

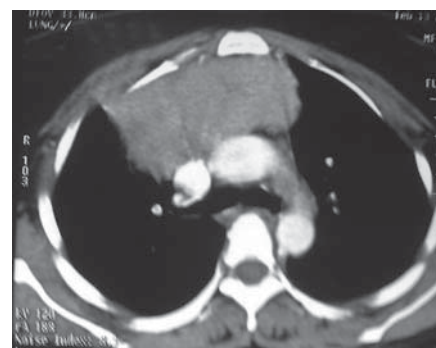


Fig. 2. CT scan showing a retrosternal mass extending below the tracheal bifurcation.

surrounding tissues including major vessels and could not be delivered intact. The thyroid mass was found to be separate from another retrosternal mass (fig. 3). Delayed recovery from anesthesia led to clinical suspicion of a myasthenic crisis which was confirmed clinically by response to administration of pyridostigmine. Furthermore, laboratory tests confirmed the presence of acetylcholine receptor antibodies. A tracheostomy was carried out a week later and the ventilator support continued for 10 days. Administration of intravenous immunoglobulin 2 g/kg body weight divided over 5 days led to improvement of the myasthenic symptoms. The patient was weaned from the tracheostomy after 3 weeks.

The final histopathology reported the thyroid tissue as nodular colloid goiter with mediastinal extension. The other mediastinal mass consisted of nests and sheets of cells with dark nuclei and scanty cytoplasm (fig. 4a). In a small focus the cells exhibited poorly formed rosettes but there were no Hassall's corpuscles. There were scattered mitoses. By immunohistochemistry almost all the cells were positive for the T-cell marker CD 3 (fig. 4b) but were

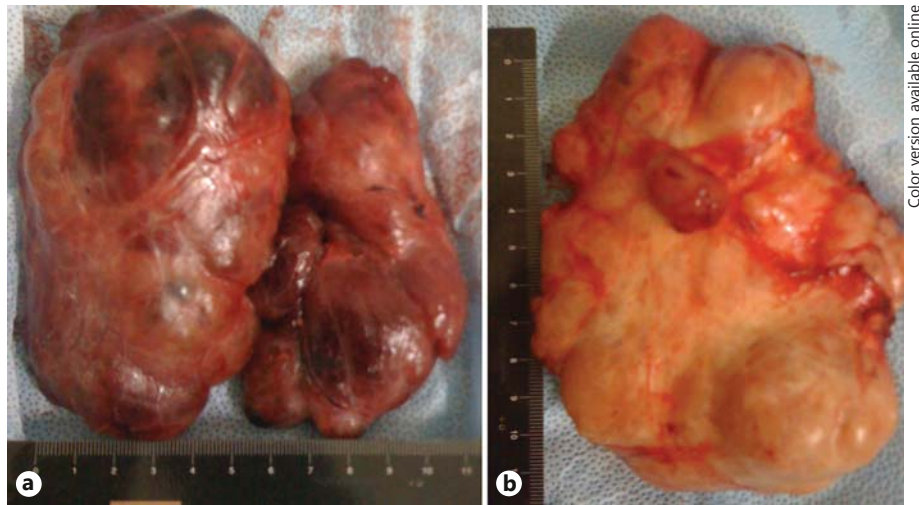
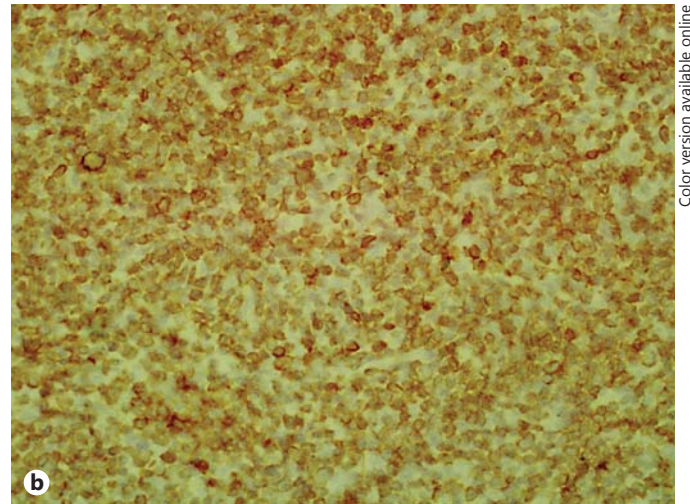
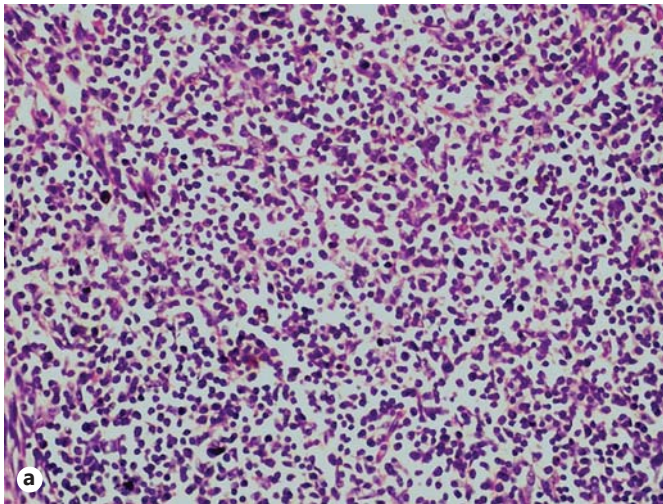


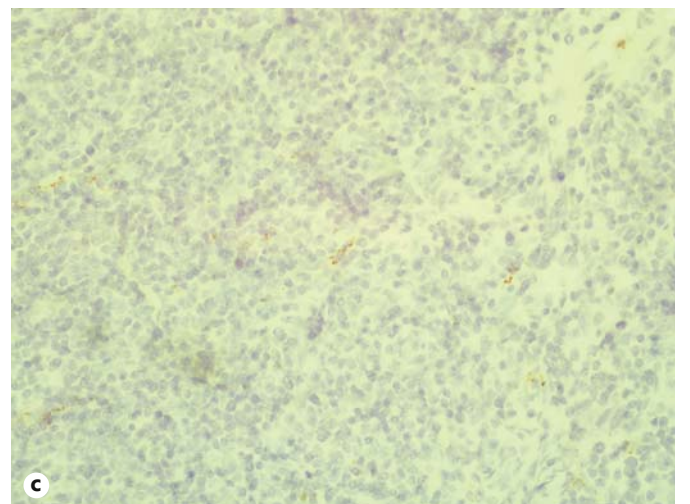
Fig. 3. Coexistence of a retrosternal goiter (a) and a thymoma (b).

Color version available online



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Fig. 4. a Nests and sheets of small cells with dark nuclei and scanty cytoplasm. There are few poorly formed rosettes. HE. $\times 40$. The cells are positive for the T-cell marker CD3. Immunoperoxidase stain (IPS). $\times 40$. **b** Most of the cells are positive for the T-cell marker CD3. IPS. $\times 40$. **c**. Cells are negative for the B-cell marker CD20. IPS. $\times 40$.



negative for the B-cell marker CD20 (fig. 4c), cytokeratin 20 and EMA. Despite the fact that epithelial cells were not seen, the diagnosis of a T-cell-rich thymoma was made. It is known that epithelial cells may be scanty on this type of thymoma. The diagnosis of thymoma was confirmed when the patient went into a thymic crisis and was shown to have acetylcholine receptor antibodies.

Thereafter, the patient received treatment for T-cell lymphoma in six cycles of chemotherapy in the form of cyclophosphamide 1 g, vincristine 2 mg, adriamycin 80 mg, prednisolone 40 mg and allopurinol 300 mg. She had been on regular follow-up with the neurologist and taking pyridostigmine 40 mg/day. After 18 months the patient was still in remission from T-cell-rich thymoma and the associated myasthenia gravis.

Discussion

The cervical swelling with retrosternal extension is a fairly common presentation of goiter. However, the pressure symptoms such as choking, shortness of breath and dysphagia as a result of the goiter masked the early symptoms of myasthenia gravis in the form of fatigability towards the end of the day.

Retrosternal, substernal, intrathoracic or mediastinal are terms that have been used to describe a goiter that extends beyond the thoracic inlet. Retrosternal extensions have been observed in 3–20% of the patients undergoing thyroidectomy [3, 4]. In contrast, primary extranodal sites of lymphoid neoplasms are rare and affect about 5% of patients with Hodgkin's lymphoma. Extranodal development is more common in non-Hodgkin's lymphoma and may reach up to 33% [5].

Myasthenia gravis nowadays includes heterogeneous autoimmune diseases with a postsynaptic defect of neuromuscular transmission as the common feature [6] and very rarely is associated with lymphoblastic malignancies. Thymic epithelial tumors can be challenging to manage because of local invasion of mediastinal structures and a high recurrence rate [7]. Non-cutaneous T-cell lymphomas are rare aggressive tumors. Peripheral T-cell lymphoma, derived from post-thymic T cells, accounts for 10–15% of non-Hodgkin lymphomas [8]. By definition, all patients with a myasthenic crisis are in respiratory failure due to muscle weakness and require ventilatory assistance.

A preoperative prediction score of postoperative myasthenic crisis following thymectomy was introduced and stratification of the patient risk was graded into four groups with grade 4 carrying a 50% risk of developing myasthenic crisis. Our patient in retrospect could be staged in group 1 with 6% risk of postoperative respiratory failure based on Osserman stage IIA (mild myasthenia gravis), BMI <28 and duration of symptoms <1 year [9]. In another study

the risk prediction of postoperative myasthenic crisis was based on the presence of bulbar palsy and an extended surgery, and both were present in our patient [10].

The differential diagnosis of anterior mediastinal masses includes thymoma, teratoma, thyroid disease, and lymphoma [8, 11]. Retrosternal goiter with pressure symptoms needs surgical treatment [12–14]. Assessment of the mediastinal mass will need both CT-guided fine-needle aspiration cytology. However, in our case the presence of a large goiter in a patient coming from an endemic region made the preoperative diagnosis of retrosternal goiter rather obvious. This patient had a thymoma in the posterior mediastinum. Thymomas may affect both the anterior and posterior mediastinum.

During the surgical procedure, intraoperative assessment of the mass indicated the need for a median sternotomy that was carried out and the mass macroscopically appeared different and totally separate from the thyroid gland. A review of the literature revealed a total of 25 cases reported as lymphoma along with myasthenia gravis and only 4 of these were reported as being T-cell lymphoma [13].

The diagnosis of myasthenic crisis should be suspected clinically and patients with impending crisis must be admitted to an intensive care unit for respiratory support. The condition is fully reversible and carries no long-term disability if treated quickly and appropriately [15, 16]. The association of retrosternal goiter with thymoma is rare. Baker et al. [17] reported a case of intrathymic primary intrathoracic goiter in a patient with breast cancer which was removed via median sternotomy.

Finally, while treatment of lymphoid malignancies usually induces improvement or remission of the myasthenia gravis symptoms, myasthenia gravis occasionally emerges as a result of treatment [13].

Conclusion

The coexistence of a large goiter with intrathoracic swelling does not always mean a retrosternal goiter. Careful history-taking and preoperative assessment is mandatory to exclude other potentially malignant conditions. Caution is needed to avoid factors that can trigger myasthenic crisis.

Disclosure Statement

The authors have no conflicts of interest to disclose.

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