Complete resolution of paraneoplastic syndrome of inappropriate antidiuretic hormone secretion following thymic small-cell carcinoma thoracoscopic resection

Pouya Hemmati and Stephen D. Cassivi*

Division of Thoracic Surgery, Department of Surgery, Mayo Clinic, Rochester, MN, USA

* Corresponding author: Division of Thoracic Surgery, Department of Surgery, Mayo Clinic, 200 First St SW, Rochester, MN 55905, USA. Tel: 1-507-266-0911; e-mail: Cassivi.Stephen@mayo.edu (S. D. Cassivi).

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Abstract

Thymic neuroendocrine tumours are rare anterior mediastinal neoplasms often associated with paraneoplastic syndromes. A patient presented with intractable hyponatraemia and a DOTATATE-avid mediastinal mass. Following medical optimization, she underwent thoracoscopic thymectomy with en bloc thymic small-cell carcinoma resection. Her symptoms resolved and her sodium levels normalized. In localized disease, curative-intent, minimally invasive thymic neuroendocrine tumour resection is safe and effective following preoperative staging and paraneoplastic syndrome management.

Keywords: Thymic small-cell carcinoma • Neuroendocrine tumor • Mediastinal mass • VATS thymectomy • Paraneoplastic syndromes

INTRODUCTION

Thymic neuroendocrine tumours (TNET) are rare anterior mediastinal neoplasms that may present with aggressive locoregional and distant metastases. Paraneoplastic syndromes are common, and there is a paucity of literature on surgical treatment. However, with an appropriate preoperative workup and perioperative medical management, minimally invasive resection is safe and effective. We present a case of thoracoscopic resection of a TNET with ectopic antidiuretic hormone (ADH) production.

CASE PRESENTATION

A previously healthy 41-year-old female presented with 3 months of nausea, vomiting, weakness and headaches. Laboratory values showed hyponatraemia (sodium 110 mmol/l), hypochloremia, serum hyposmolality and inappropriately elevated urine sodium levels. Endocrine panel values were normal. She was admitted and treated for syndrome of inappropriate ADH secretion. However, she experienced persistent symptoms and admissions for recalcitrant hyponatraemia.

A positron emission tomography-computed tomography scan showed a DOTATATE-avid anterior mediastinal thymic lesion measuring 2.4 x 1.7 cm (Fig. 1). The results of magnetic resonance imaging of the brain were normal. An encapsulated TNET with ectopic ADH production was the most likely diagnosis. It appeared amenable to a thoracoscopic total thymectomy with en bloc resection. Preoperatively, her sodium increased to 129 mmol/l with intravenous infusions of hypertonic (3%) saline, oral fluid restriction and sodium chloride tablets.

Operative technique

After induction of general anaesthesia, double-lumen endotracheal intubation and right lateral decubitus positioning, a pneumothorax was induced with a Veress needle and carbon dioxide insufflation. Five-mm airtight Thoracoport trocars (Medtronic, Minneapolis, MN, USA) were placed 2 cm anterior to the scapular tip (5th intercostal space) and the axillary hairline base (3rd intercostal space). A 10-mm Thoracoport trocar was placed anteriorly (7th intercostal space) (Fig. 2A). The lesion was visible within the thymic capsule, and dissection was initiated on the left lower thymic pole. We deflected away tissue by pushing instead of grasping the mass to create countertraction for dissection.

The mass was removed from the pericardium posteriorly within its own capsule (Fig. 2B). The left phrenic nerve was identified and preserved. After dissecting the left lateral edge, we crossed the pericardium to reach the right mediastinum. Superiorly, we clipped 2 innominate vein branches draining the thymus. Both upper poles were dissected to their cephalad extent prior to division. We then dissected inferiorly along the right...
lateral border to completely excise the right lower pole. Finally, the anterior retrosternal attachments were divided. The specimen (Fig. 2C) was removed with a surgical bag. No chest tube was required. Frozen sections showed negative margins and a 2.7-cm encapsulated TNET.

Postoperative course

There were no intraoperative complications. She was discharged on postoperative day 1 with normal sodium levels. The final pathological analysis demonstrated a 25-g, 2.7 x 1.1 cm fully encapsulated small-cell carcinoma confined to the thymus with negative margins (Fig. 2D). At the 2-month follow-up, her symptoms had resolved (normal sodium levels). One year later, she had distant recurrence with bony metastases and hyponatraemia; she was treated with chemoradiation therapy (etoposide/cisplatin).

DISCUSSION

TNETs are the least common primary thymic neoplasms (2–5%) [1]. Only 2–8% of patients with multiple endocrine neoplasia-1 develop TNETs, but 25% of patients with TNETs have multiple endocrine neoplasia-1, so genetic testing is warranted [2]. Thymic small-cell carcinomas are high-grade TNETs that present with significant local invasion. Over 50% of patients have mediastinal lymph node involvement, and 20–40% have distant metastases at presentation, but carcinoid syndrome is rare [1]. There is a paucity of reported cases of surgical resection of TNETs, especially with associated paraneoplastic syndromes [3–5].

To our knowledge, this is the first report of minimally invasive resection of a thymic small-cell carcinoma causing inappropriate ADH secretion. The patient presented with intractable hyponatraemia and an anterior mediastinal mass. Multidisciplinary medical optimization and preoperative imaging for accurate localization were critical for thoracoscopic resection. Using carbon dioxide insufflation facilitated meticulous dissection near...
critical mediastinal structures and obviated chest tube placement. Successful en bloc total thymectomy immediately resolved her debilitating symptoms and normalized her sodium levels. However, repeat imaging, clinical follow-up and monitoring laboratory results are essential for detecting likely recurrence.

CONCLUSION

Thymic small-cell carcinomas are rare anterior mediastinal masses and a subset of TNETs with aggressive locoregional invasion and distant spread. They can also present with disabling paraneoplastic syndromes. With rare early detection of localized disease, curative-intent, minimally invasive resection is safe and feasible following appropriate preoperative staging, localization and management of the paraneoplastic syndrome. The benefit of adjuvant local or systemic therapies is unclear, but they are offered due to the often poor prognosis.

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