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Case report

Robotic approach to a subcarinal functional paraganglioma

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A B S T R A C T

Intro: Functional mediastinal paragangliomas arise from extra-adrenal tissues and are rare. These cases create challenges related to diagnosis, peri-operative management, and surgical management. We present a case that demonstrates a planned robot-assisted thoracoscopic resection of a mediastinal paraganglioma that ultimately required a *trans*-sternal resection of the tumor off the left atrium.

Case report: Our patient is a 42-year-old male with a prolonged history of refractory hypertension, palpitations, headaches, and diaphoresis, which led to the discovery of a subcarinal functional mediastinal paraganglioma. The patient was brought to the operating room for a right robotic-assisted thoracoscopic subcarinal dissection with attempted resection of the mass. Subsequently, the patient's paraganglioma was successfully resected off the left atrium using a *trans*-sternal approach, cardiopulmonary bypass, and cardioplegic arrest. He was successfully transitioned to minimal anti-hypertensive medication post-operatively.

Discussion: Pheochromocytomas are neural-crest derived tumors that typically arise from the adrenal medulla. Rarely, paragangliomas arise in the thoracic cavity, at an approximate incidence of 2%. Our sequential approach offered the potential for a minimally invasive resection, and though initially unsuccessful, safely elucidated the feasibility of resection using cardiopulmonary bypass after confirming no invasion of the airway, esophagus, or other mediastinal structures.

1. Introduction

Pheochromocytomas are neural-crest derived, tumors that typically originate in the adrenal medulla [1]. Approximately 10% of these tumors arise from extra-adrenal tissues and are termed paragangliomas. Approximately 10–15% of pheochromocytomas will exhibit malignant behavior, defined as the presence of paraganglioma in non-chromaffin tissues [2]. Mediastinal paragangliomas are uncommon and usually non-functional, with an approximate incidence of 2% of all reported cases [3]. Functional mediastinal paragangliomas are extremely rare, with approximately fifty cases reported in the literature and require resection for symptomatic relief. Previous case reports have illustrated the complexity and multi-disciplinary approach to functional mediastinal paragangliomas. However, most of these reports utilized an open surgical approach to attempt resection, with some tumors unresectable.

We present a case that demonstrates a planned robotic-assisted thoracoscopic resection to stage a mediastinal paraganglioma, which ultimately required cardiopulmonary bypass for resection and reconstruction of the posterior wall of the left atrium.

2. Case

Our patient is a 42-year-old male with a prolonged history of refractory malignant hypertension, palpitations, headaches, and diaphoresis. Confirmatory diagnostic testing included metanephrines levels consistent with pheochromocytoma. The patient underwent an MIBG (metaiodobenzylguanidine) scan and cross-sectional imaging which localized a 6 cm sub-carinal mass with no evidence of metastatic disease. Figures 1 and 2 based on preoperative *trans*-thoracic echocardiography and cross-sectional imaging, there appeared to be a well-defined plane between the mass and the left atrial wall.

The patient was pre-operatively alpha blocked with doxazosin for several weeks prior to resection, then brought to the operating room for a right robotic-assisted thoracoscopic resection and lymphadenectomy. Despite thorough dissection of the subcarinal lymphatics, resection proved challenging. A complete subcarinal lymph node dissection was performed, and the trachea, mainstem bronchi, and esophagus were noted to be completely free of tumor invasion. However, there was significant tumor invasion of the posterior atrial wall. Intra-operative *trans*-esophageal echocardiography confirmed involvement with the left atrium, which precluded a minimally invasive resection (Figs. 3 and

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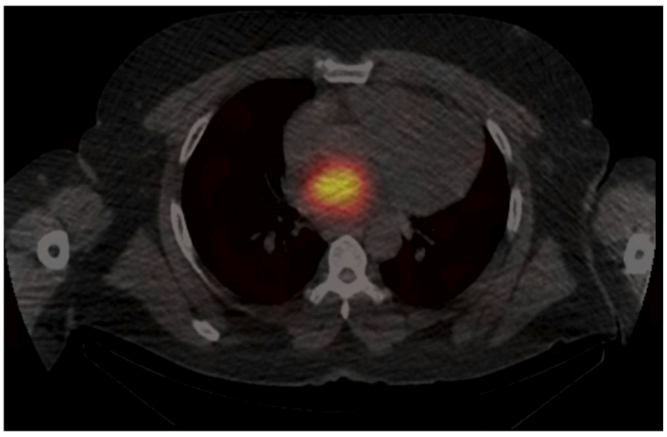

Fig. 1. 10-18-18 MIBG/CT combined scan demonstrating the subcarinal paraganglioma.

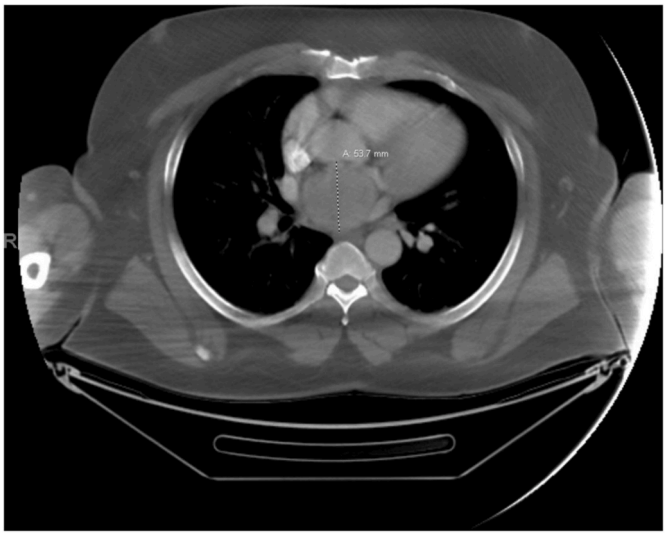

Fig. 2. 10/12/18 preoperative CT with contrast.

4).

Two weeks later, the patient was re-admitted for a *trans*-sternal resection of the mass. The mass was visible upon retracting the ascending aorta to the left. The mass extended rightward underneath the superior vena cava, and this was dissected free from its attachments to the mass. Manipulation of the mass variably resulted in sustained tachycardia, associated with forceful cardiac contractions. The mass was dissected posteriorly from the peri-carinal tissue, but the mass was firmly adhered to the posterior atrial wall. Cardiopulmonary bypass and cardioplegic arrest were required to complete the resection safely. The oncologic resection resulted in a left atrial 2 cm × 2 cm defect, which was repaired with a bovine pericardium patch.

The patient remained in the Intensive Care Unit for three days but had an unremarkable post-operative course and was discharged home on post-operative day five.

The pathology report demonstrated a 6.1 × 4.5 cm mass, staining "strongly and diffusely" positive for synaptophysin and chromogranin without evidence of capsular or vascular invasion. In addition, there were no atypical features such as tumor necrosis or high cellularity.

At his post-operative follow up one month later, he was recovering well, and had dramatically reduced his anti-hypertensive medication regimen to a calcium channel blocker.

3. Discussion

Mediastinal paragangliomas encompass 2% of all catecholamine-secreting tumors and on rare occasion present as typical pheochromocytomas, with refractory hypertension, headaches, diaphoresis, and palpitations [4]. Management for benign disease is relatively routine: pre-operative alpha blockade and complete R0 resection, which confers a significant survival benefit. Malignant disease, defined as extensive local invasion with presence of tumor in nonchromaffin tissues on pathological evaluation, can be managed with adjuvant 131I meta-iodobenzylguanidine. At the time of surgical resection, there was concern for a locally advanced tumor, but subsequent pathological evaluation revealed a defined capsule with complete R0 resection.

Although previous case series suggest an open approach, ideal access of the sub-carinal space is through the right chest and presently this is accomplished via thoracoscopy including robotic VATS. Preoperative imaging showed no evidence of metastatic spread. Complete oncologic resection would include both resection of the paraganglioma and lymph

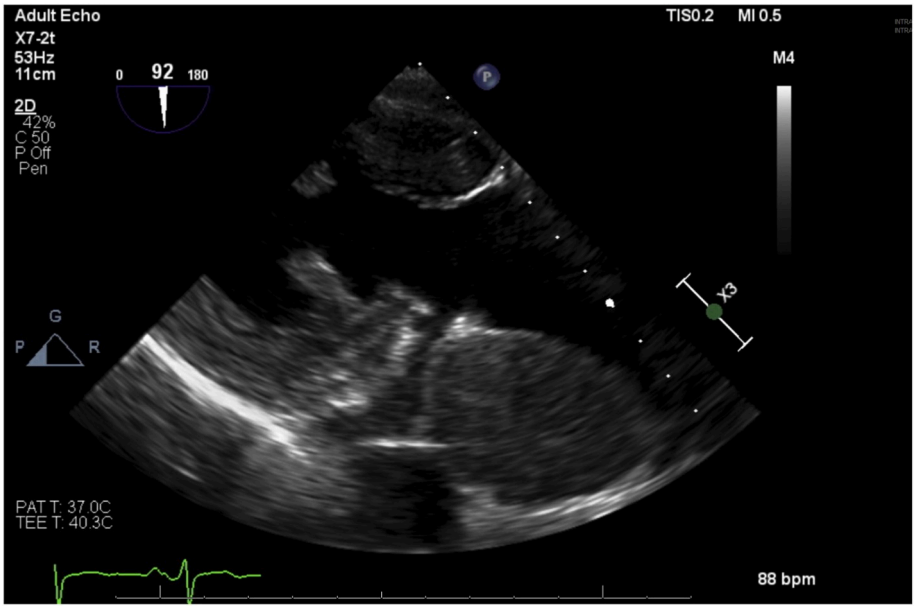

Fig. 3. 1/9/19 Intraoperative *Trans*-Esophageal Echocardiography (TEE): (long axis, showing pulmonary vein compressed by mass and mitral valve in near vicinity).

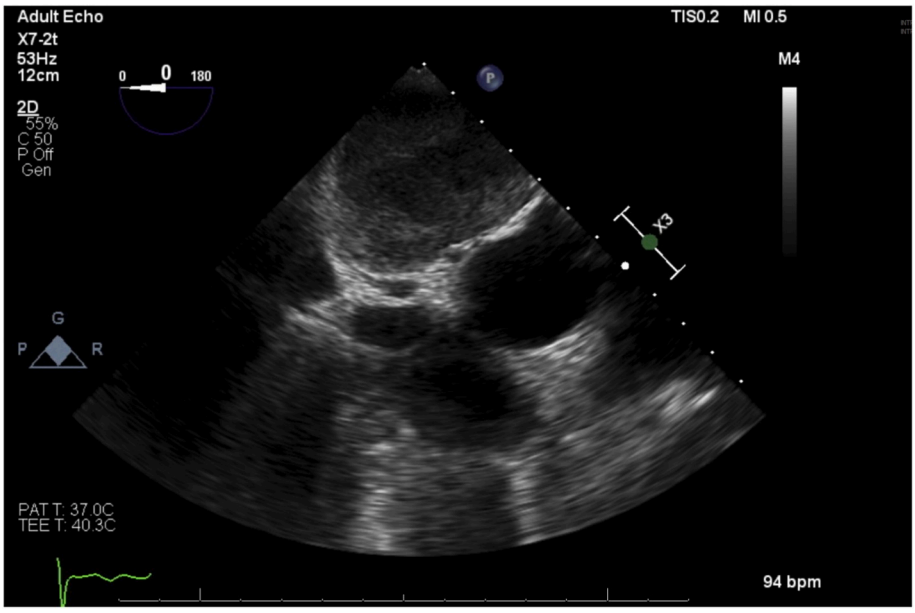

Fig. 4. Intraoperative TEE cont'd: now showing the aortic valve (and right coronary) with mass abutting the area immediately adjacent.

node dissection, due to size, both of which could be accomplished with robotic VATs [5]. The robotic VATs served as a staging procedure with negative lymph nodes found on pathology.

Surveillance follow up is controversial and potentially differs between malignant and benign disease. As reported by Elder et al., in a review of 85 patients, 71 patients had tumors considered benign, and none of these patients had recurrence at a median follow-up of 144 months [6]. The authors offered a reasonable suggestion that patients with benign tumors, required no further surveillance following resection of their paraganglioma. However according to the NCCN (National Comprehensive Cancer Network) guidelines, patients should have surveillance every three months for the first year with blood pressure and serum markers; biannually for three years, and then annually for 10 years thereafter. Malignant tumors should be followed every 3–12 months. For metastatic disease, systemic chemotherapy is utilized in addition to cytoreductive resection. Patients with malignant pheochromocytomas should have surveillance every three months with blood pressure screenings and plasma markers [7]. In addition, the European Society of Endocrinology recommends ten year follow-up with annual surveillance laboratory testing with no distinction between benign and metastatic disease [8].

There is a paucity of case reports in the literature highlighting a mediastinal mass with local invasion into the left atrium, with successful R0 resection. Our sequential approach provided a more detailed evaluation of the mass thoracoscopically with minimal morbidity, confirmed the potential resectability of the mass with no invasion of the airway or esophagus, and allowed a safe and thoughtful plan to completely resect the patient's tumor.

4. Declaration

The primary author noted above and the relevant authors noted in this manuscript to have no undue conflict of interest to report. In addition, there is no inappropriate alternative funding source or study sponsor to disclose.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.rmcr.2020.101092.

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