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Commentary: Management of cardiac paraganglioma is a team sport

Kevin L. Greason, MD

No one is big enough to be independent of others.

—Dr William Worrall Mayo¹

Readers involved in the treatment of patients with complex cardiac tumors will be interested in this nicely written article on the rare condition of cardiac paraganglioma by Chan and colleagues.² Although the focus is on patient and operative characteristics, the article also provides a general overview of the disease and the institutional multidisciplinary heart team's philosophy on evaluation and management.

The study provides a review of a single institution and surgeon experience (MJR) in 19 patients undergoing surgery from 2004 to 2020. No patient received embolization. The operation was often complex, requiring aortic root replacement in 4 patients (21%) and autotransplant in 8 (42%). All patients had complete gross tumor resection. Operative mortality occurred in 2 patients (11%; 1 intraoperative). No patient experienced local recurrence of disease, but 1 died of metastatic disease. Survival was 72% at 3 years.

Additional data and analysis provide several important observations. Two patients had previous emergency sternotomy for control of needle biopsy related bleeding and cardiac tamponade. Metabolically active tumors were present in 6 patients (32%). Complications were frequent and included blood transfusion (16 patients, median of 5 U), renal failure, cardiac tamponade, need for mechanical circulatory support, prolonged ventilation, tracheostomy, and vocal cord paralysis. Median length of stay in the intensive care unit was 204 hours (my calculation from the authors' Table 2) and hospital was 14 days.

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CENTRAL MESSAGE

Cardiac paraganglioma is a rare and complex disease that often requires complex operative techniques and mechanical circulatory support. A multidisciplinary heart team approach is required.

The main strength of this article is the focused analysis of a rare disease. It is kind of a military postdeployment briefing on lessons learned in the management of 19 patients over 16 years by a single surgeon. The discussion section gives an overview of the pathology, imaging, perioperative management, genetics/malignancy, and multidisciplinary heart team approach. The surgical takeaway is that operations should be tailored to tumor location, size, and structures involved to achieve complete resection.²

The limitations of the study are appropriately outlined by the authors. The small number of patients, retrospective methodology, and referral bias are reflective of the rarity of the disease. Similar confines have been present in other recently published focused institutional experiences.^{3,4} A perceived weakness of these studies is an apparent lack of address on what to do about bleeding.²⁻⁴ Safe and successful embolization has been reported.⁵ Why not do preoperative embolization? My review of the authors' associated video leads me to believe the patient would have been an ideal candidate for preoperative embolization.

Chan and colleagues² have added to our understanding of this rare disease. I believe a consensus is present that mandates a multidisciplinary heart team approach to these patients' condition. A magnetic resonance imaging scan should be done before biopsy of any heart tumor. All patients should be genetically screened and medically managed. Preoperative embolization should be considered. Effective surgical resection, complex reconstruction, and need for mechanical circulatory

support should be thoroughly planned for to optimize success. Complete surgical resection is the treatment.

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Commentary: What lessons can a surgeon learn from something rare?

Kenza Rahmouni, MDCM, and
Fraser D. Rubens, MD, MSc, FACS, FRCSC

Primary cardiac paragangliomas (PCPs) are rare neuroendocrine tumors of the heart. In this issue of the *Journal*, Chan and colleagues¹ share their center's experience in managing patients with PCP, presenting a cohort of 19 patients with a perioperative mortality of 10.6%. The surgical steps are complex yet through experience and wise application of oncologic principles, this center is now a site of established excellence.

So how does the average cardiac surgeon interpret this type of presentation? Even in academic centers, one anticipates that these cases will be few and far between, and it should be recognized that we should support focusing this cohort into the hands of a few experts in each country. A better lesson from this manuscript is a template of how to address rare surgical diseases in general, be they from cancer or from other uncommon cardiac mechanical problems.

The authors have identified several principles that are critical to success. First, they have emphasized the importance of a large volume to efficiently turn experience into

Kenza Rahmouni, MDCM (left), and Fraser D. Rubens, MD, MSc, FACS, FRCSC (right)

CENTRAL MESSAGE

Surgical outcomes are intimately related to surgeon and institution expertise. We therefore must further centralize the care of rare cardiac tumors and refer patients to specialists earlier.

reproducible surgical strategies. We know well the intimate relationship of surgical outcomes with institution expertise. A concrete example of this correlation is the experience in San Diego in managing chronic thromboembolic pulmonary hypertension with pulmonary endarterectomy.² Over 50 years, their surgical outcomes for this highly subspecialized operation have consistently improved despite growing complexity of their cases. They are the center of excellence, frequently managing patients with chronic thromboembolic pulmonary hypertension deemed non-surgical in other centers.

The second important facet of success involves contribution from a multidisciplinary team, a strategy that has been recommended for more than 25 years for patients with

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