Commentary: Cardiac paraganglioma: The surgical challenge

Michael J. Reardon, MD

From the Department of Cardiothoracic Surgery, Houston Methodist Hospital, Houston, Tex.

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Address for reprints: Michael J. Reardon, MD, Department of Cardiothoracic Surgery, Houston Methodist Hospital, 6550 Fannin, Suite 1401, Houston, TX 77030 (E-mail: mreardon@houstonmethodist.org).

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Paragangliomas are chromaffin tumors from the neural crest cells originating from the parasympathetic and sympathetic chains and are considered neuroendocrine tumors. Only 1% to 2% originate in the chest, mainly in the posterior mediastinum.¹ Cardiac paragangliomas are a subset of these and very rare. Cardiac paragangliomas can occur anywhere in the heart, but in our experience are most common in the roof of the left atrium and base of the pulmonary artery. These tumors have a soft fleshy consistency, draw large parasitic blood vessels from the coronary arteries (characteristic of paragangliomas when seen on coronary angiography), and have no capsule. They occur in both hormonally active and nonhormonally active forms similar to paragangliomas occurring elsewhere. Hormonally active cardiac paragangliomas are not referred to as pheochromocytomas because this term is limited to adrenal hormone-active paragangliomas. Patients with cardiac paraganglioma and symptoms suggestive of hormone activity should be tested and if positive have appropriate hormone blockage before surgery. Surgical resection when possible remains the primary treatment for these tumors. Cardiac paragangliomas are interesting not only for the unique challenges they present to a cardiac surgeon during resection, but also for their interesting genetic associations. About three-fourths arise spontaneously, whereas one-quarter have a genetic basis.¹ Seventeen genes have been linked to extra-adrenal paragangliomas, but only succinate dehydrogenase mutations were found to be associated with cardiac paragangliomas.¹ A small series of 13 patients with a cardiac paraganglioma were genetically tested and 10 patients (76.9%) were positive for mutations in succinate dehydrogenase subunits B, C, or D. Our group believes that patients with a cardiac paraganglioma should be genetically screened.² Finally, about 10% of cardiac paragangliomas are malignant. This cannot be determined by the pathology of the surgical specimen but is defined by distant spread rather than histology. We discuss the possibility of late malignancy with all of our patients and suggest lifelong follow-up.

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Central Message

Cardiac paragangliomas are rare tumors that require complex cardiac resections.

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My interest in cardiac paraganglioma and cardiac tumors in general began as a cardiothoracic resident working with Denton Cooley when he used the technique of cardiac autotransplant to approach and resect a large and complex left atrial paraganglioma.³ This instigated a career-long interest in both the technique of cardiac autotransplant⁴ and cardiac paragangliomas⁵ with our current series at 19 patients (unpublished data). With this long interest, it was with great pleasure that I read the article by Lui and colleagues⁶ describing their 13-year experience with 17 patients undergoing surgical resection for hormonally active cardiac paraganglioma. Their large series is common in that the patients are young with a mean age of 35 years. It is uncommon in that all of the patients have hormonally active tumors. Whether this is a referral bias based on these patients likely having more symptoms prompting referral or just the luck of the draw is unknown. They clearly outline their evaluation for this tumor and describe the surgery with its necessary commonly needed adjunctive procedures. All patients had complete resection despite the complexity of these tumors with only 1 death (5.8%), a highly commendable achievement. The mean follow-up was 6.5 years with all patients other than the operative death remaining alive with 2 local tumor recurrences. The article by Lui and colleagues⁶ is well worth reading for anyone interested in complex cardiac tumors and I salute the authors for both their outstanding outcomes as well as their meticulous follow-up of their patients.

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