A needle in a haystack

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Central Message

Primary cardiac sarcoma is rare and very aggressive. Treatment protocols remain to be fully defined. Specialized cardiac tumor referral centers help to define the best treatment strategies.

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Wu and colleagues¹ present a series of 12 primary cardiac sarcomas operated on over a 16-year period at a large institution. There are a number of important findings in this small series that are important in understanding this rare disease. The first is that it is very unusual with this large institution seeing less than 1 case per year. These are young patients with a median age of 43 years. They had 58% of their surgeries ending with macroscopic (R2) margins. Median survival was only 12 months, but negative (R0) or microscopically positive only (R1) margins had better survival than R2 margins (21.8 vs 7.2 months). This better survival should not surprise us, but more than half having macroscopic disease left behind is problematic, and ways to avoid this should be sought. The authors specifically note that the completeness of surgical resection is key. They note that 75% of their cases had postoperative chemotherapy or radiation therapy because surgery alone was not curative and that these patients had a 6 times longer survival than surgery alone. The authors note that in their patients presenting with metastatic, no patient survived longer than 7 months. All of these findings show how deadly and aggressive this orphan disease really is.

Our group developed an interest in primary cardiac sarcoma stemming from an initial left atrial sarcoma operated on by the senior author (MJR).² We developed a multi-institutional, multidisciplinary team and wrote a protocol approved by our institutional review board to standardize our approach to primary cardiac sarcoma. Currently, we have treated more than 200 primary cardiac sarcomas and operated on 116 at last count. From this experience, we have gleaned several interesting observations that have helped us in our approach to this disease. We divide primary cardiac sarcoma into 3 groups depending on their location: right heart, left heart, or pulmonary artery. We find location more than histology controls how these tumors present and the treatment options available. Right heart tumors tend to be bulky and grow exophytically such that obstruction of flow and heart failure is a very late finding. They also

tend to be more infiltrative, making R0 resection more difficult. We always seek tissue diagnosis before surgery both to R/O tumors such as lymphoma that do not need surgery and to allow neoadjuvant therapy before surgery. We now give this neoadjuvant therapy for as long as the patient can tolerate it, and we continue to get a response. This had led to increased R0 resections and survival.³ Left heart tumors tend to be more solid and obstruct blood flow earlier. In fact, all but 1 of the left atrial sarcomas we have operated on were first removed in patients at other institutions where they were thought to be myxoma and inadequate resection led to local recurrence. The anatomy of left heart sarcomas can lead to extreme difficulty in achieving complete resection with specialized surgical techniques, such as cardiac autotransplantation.⁴ We fully agree with the authors that these complex tumors are best handled at centers with access to these specialized procedures. Pulmonary artery sarcoma generally presents late with near total obstruction of pulmonary blood flow, making both histologic diagnosis and neoadjuvant therapy difficult because of the urgent need for surgery.⁵ In addition, we have treated both left heart tumors and pulmonary artery tumors that require both a large cardiac resection and a pneumonectomy for complete removal. These cases also require a specialized surgical team approach performing the cardiac resection first, allowing the patient to recover, and then proceeding with pneumonectomy: the “Texas Two-Step.”⁶

Finally, we agree with the authors that not only specialized surgical procedures are needed but also overall specialized care is needed from experienced oncologists

and cardiology cardio-oncologists. Treatment planning for these patients is crucial to success and is best done with an established heart tumor team. Adriamycin and ifosfamide are the standard front-line drugs, but we find that many referring oncologists may limit the doses required for this aggressive disease because of worry about cardiotoxicity. This problem can be managed well by the experienced oncology and cardio-oncology cardiologist. We have established a monthly cardiac tumor conference that is web cast so that we can include a number of institutions both nationally and internationally. The American College of Cardiology cardio-oncology committee chaired by Dr Leja has taken on primary cardiac sarcoma as a cardio-oncology endeavor, and we are discussing the formation of an international registry for these rare tumors. These tumors are rare and difficult to treat effectively. Finding the best treatment approach when the cases have

been so spread thin is like finding a needle in a haystack. We thank the authors for adding cases to the haystack and welcome help in this search.

References

1. Wu Y, Million L, Moding EJ, Scott G, Berry M, Ganjoo KN. The impact of postoperative therapy on primary cardiac sarcoma. *J Thorac Cardiovasc Surg.* 2018;156:2194-203.
2. Reardon MJ, DeFelice CA, Sheinbaum R, Baldwin JC. Cardiac autotransplant for surgical treatment of a malignant neoplasm. *Ann Thorac Surg.* 1999;67:1793-5.
3. Abu Saleh WK, Ramlawi B, Shapira OM, Al Jabbari O, Ravi V, Benjamin R, et al. Improved outcomes with the evolution of a neoadjuvant chemotherapy approach to right heart sarcoma. *Ann Thorac Surg.* 2017;104:90-6.
4. Ramlawi B, Al-Jabbari O, Blau LN, Davies MG, Bruckner BA, Blackmon SH, et al. Autotransplantation for the resection of complex left heart tumors. *Ann Thorac Surg.* 2014;98:863-8.
5. Blackmon SH, Rice DC, Correa AM, Mehran R, Putnam JB, Smythe WR, et al. Management of primary pulmonary artery sarcomas. *Ann Thorac Surg.* 2009; 87:977-84.
6. Chan EY, Reul RM, Kim MP, Reardon MJ. The "Texas Two-Step" procedure. *J Thorac Cardiovasc Surg.* 2018;155:285-7.