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EDITORIAL COMMENT

Measure Twice, Cut Once*

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The management of primary cardiac sarcomas remains a vexing problem as patients present infrequently and often unexpectedly. In this issue of *JACC: Case Reports*, we appreciate the case report by Diamond et al. (1), who nicely highlighted some of the challenges associated with these lesions and, in particular, the complexity of complete surgical resection with margins. Unfortunately, too many of these patients are treated at centers without multidisciplinary expertise and undergo inadequate surgical resection or debulking, reducing even further the small possibility of cure.

Primary cardiac tumors as a group are infrequent, with an incidence ranging between 0.001% and 0.030% (2). Fortunately, the majority of these lesions are benign, with myxomas comprising approximately one-half. Surgical resection of myxomas is relatively routine, and minimal margins achieve good long-term results with only a small risk of local recurrence. The problem is that not all primary cardiac tumors are benign. Approximately one-quarter of primary cardiac lesions are malignant, with sarcomas being most common. For these malignancies, the usual resection techniques are not sufficient, and local recurrence is more the rule than the exception. A recent series of sarcoma patients from an experienced center reported a median survival of 20 months following surgical resection (3). One would surmise that aggregate results of patients treated by less-experienced surgeons without multidisciplinary collaboration will be even less favorable. This remains conjecture as this data is not available.

Primary cardiac tumors are typically diagnosed on routine imaging and are classified based on their locations and morphology. Myxoma resections have become a fairly routine cardiac operation with a broad but dilute experience within the surgical community. Pre-operative imaging, however, can be misleading, and even typical-appearing lesions can cloak malignancies (4). Although one could easily argue that atypical lesions in the heart deserve intense scrutiny and are best managed at centers with multidisciplinary expertise, one could also reasonably advise that all patients with cardiac masses deserve such consideration. As these authors point out, there are no available guidelines for best practices in the management of patients with primary cardiac malignancies. We feel strongly that it is time to focus our collective experience and to identify regional centers of expertise.

Part of the challenge in managing these patients is that they frequently present late with hemodynamic compromise, as this case typifies. A patient presenting in shock or who is otherwise unstable cannot always afford the luxury of a multidisciplinary evaluation and optimal imaging to assist surgical planning. Unfortunately, these same patients are the ones that most benefit from pre-operative scrutiny, intensive imaging, and operative planning. Surgical resection remains the gold-standard treatment, and as the authors point out, success with this modality most closely correlates with long-term survival. Although post-operative adjuvant therapy has no proven benefit for cure (5), recent studies suggest the pre-operative neoadjuvant treatment may contribute to increased survival (6). This stresses the importance of having a pre-operative diagnosis. As with this case, sarcomatous lesions often involve or reside in close proximity to vital cardiac structures, making curative resection extremely challenging if not impossible. A

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surgeon must balance the risk of the operation due to damage of vital structures with that of recurrent disease, and we are inherently and understandably biased to avoid the former. One option that having a pre-operative diagnosis affords is the ability to mark close or positive margins with radiopaque fiducial markers to permit post-operative highly focused radiation therapy, including application of proton beam therapy. Surgeons who are the most poised to both minimize and accept operative risk and offer a full range of multidisciplinary providers are those with the most experience in the management of this disease.

Survival of patients presenting with primary cardiac malignancies remains dismal. To make meaningful clinical progress in the management of a disease that occurs at very low frequency, we must support regional centers of expertise. This requires not only institutions willing to commit the necessary resources, but also the willingness of patients and referrers to seek out these resources. When the margin of error is slim, to succeed it is imperative to measure twice and cut once.

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