



## EDITORIAL COMMENT

## Primary malignant cardiac tumors: Surgical results<sup>☆</sup>



## Tumores cardíacos primários malignos: resultados cirúrgicos

José Fragata

*Serviço de Cirurgia Cardiorácica, Centro Hospitalar Lisboa Central, EPE, Hospital de Santa Marta, Lisboa, Portugal*

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This issue of the *Journal* contains an article on primary malignant cardiac tumors (PMCTs) by Saraiva et al.<sup>1</sup> This is a series of 12 cases over a 20-year period in a high-volume Portuguese center in which 23 010 patients underwent cardiac surgery; of these, 123 had cardiac tumors, of which 9.8% were PMCTs.

PMCTs are rare, and only long-term multicenter studies obtain significant numbers of patients, making this series particularly important. In one large series of around 550 cases, PMCTs accounted for 0.008% of all malignancies in a registry of over seven million malignant tumors.<sup>2</sup> Most are sarcomas (angiosarcoma, rhabdomyosarcoma and leiomyosarcoma), representing 65–68% of all PMCTs, a figure that is highly consistent in the literature. The next most frequent types are lymphomas (25–27%), mesotheliomas and epithelioid tumors (5–10%); this is similar to the distribution found in the series by Saraiva et al.<sup>1</sup> The slight predominance of females and age around 50 years are also close to the figures in most published series.

PMCTs are usually asymptomatic until the advanced stages of the disease, when symptoms arise due to invasion of the cardiac chambers and resulting hemodynamic disturbances. They thus tend to present late, frequently with dyspnea, chest pain, palpitations, edema or pericardial effusion. In this series, as often found in other studies, some patients present with local invasion or distant metastasization at the time of diagnosis; this is common with the

most frequent type of sarcoma, angiosarcoma, which often metastasizes to the brain, liver, lungs or bone in the initial stages.

Although not specified in the article by Saraiva et al., it can be assumed that the initial diagnosis in most cases was of “unexplained cardiac mass” detected by transthoracic or transesophageal echocardiography. However, these imaging modalities cannot clearly define the invasion margins or characterize the structure of the mass and so cannot distinguish a tumor from a possible thrombus, which can be a differential diagnosis. Nowadays, ECG-gated computed tomography provides better definition of tumor structure and invasion margins and shows its relation with the coronary arteries.<sup>3</sup> However, magnetic resonance imaging (MRI) with late gadolinium enhancement is currently the method of choice to diagnose and stage cardiac tumors, both local and metastatic. In fact, in a few cases of metastasizing cardiac melanoma, the paramagnetic nature of melanin has led to the type of tumor being identified by MRI.<sup>4</sup>

In some other series, the histopathological diagnosis had already been made by endomyocardial biopsy at the time of intervention. Given the short survival time associated with PMCTs, especially sarcoma, imaging studies are particularly valuable in order to select candidates for surgery, which can be extensive. This is particularly true for primary cardiac lymphomas, which, as Saraiva et al. point out, tend to respond well to chemotherapy. The article does not state whether patients were excluded if their tumor was not suitable for resection or had metastasized; presumably aggressive surgery was sometimes performed solely for debulking purposes. In our experience, in such cases local recurrence rates are very high, even with associated chemotherapy.

With regard to techniques, the results obtained by Saraiva et al.<sup>1</sup> clearly demonstrate the high surgical qual-

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E-mail address: [jigfragata@gmail.com](mailto:jigfragata@gmail.com)

ity of the center. In some cases extensive resection was performed, sometimes to the free margin of the lesion, together with reconstruction of the cardiac valves and walls. As the authors point out, it is important to ensure excision with tumor-free margins; in our center we regularly employ extemporaneous examination, which, although increasing surgical time, can help to individualize treatment. Some patients present altered vasoactivity, particularly vasoplegia, which Saraiva et al. do not appear to have encountered, since all their patients recovered without the use of inotropics.

In their discussion, the authors refer to transplantation as a possible, but controversial alternative. However, in selected cases in which the tumor is restricted to the myocardium and there is no metastasization, transplantation can and should be considered. In a case in which this author was involved of cardiac fibrosarcoma subsequent to thoracic radiotherapy, heart transplantation after initial debulking of a right ventricular tumor has resulted in survival for over 15 years.<sup>5</sup>

With regard to outcomes, the low survival rate is not surprising; it is, in fact, gratifyingly higher than reported in the literature (41.7% at two years for all PMCTs and 37.5% for sarcomas<sup>1</sup>). In a series of around 550 malignant cardiac tumors, five-year survival in recent years was 19% for all PMCTs, 11% for sarcomas and 34% for lymphomas.<sup>2</sup> Advances in adjuvant chemotherapy may lead to improvements in these figures, at least for lymphomas, since the prognosis for sarcomas remains bleak even after surgical excision with negative margins combined with chemotherapy or radiotherapy.

To summarize, the rarity of PMCTs appears to justify the creation of a global registry with a sufficiently large number of patients to assess the effects of different therapeutic

options. There is also a need for further improvements in diagnosis, since ideally the histological type should be determined before intervention; MRI and positron emission tomography appear to be best suited for this purpose. Detection of circulating tumor markers may also be useful. If such techniques can be combined, together with the establishment of a wide-ranging registry and early histological diagnosis, our understanding of the biological behavior of these tumors will improve, leading to individualized and integrated treatment plans like those available for other malignancies, resulting in better survival. The article by Saraiva et al.<sup>1</sup> is a significant contribution to achieving this.

## Conflicts of interest

The author has no conflicts of interest to declare.

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