





## LETTER TO THE EDITOR

# Surgery of primary malignant cardiac tumors



#### Cirurgia dos tumores cardíacos primários malignos

We read with interest the recent article by Saraiva et al.<sup>1</sup> of the Cardiothoracic Surgery Center, University Medical Center, Coimbra, Portugal, under the direction of Dr MJ Antunes. It is an excellent study on 12 patients treated by surgical excision of a primary malignant cardiac tumor (eight sarcomas, three lymphomas and one epithelioid hemangioendothelioma) in a 20-year period. Complete resection (negative margins) was obtained in five cases and partial resection in seven (five sarcomas and two lymphomas). The operative mortality was zero. Adjuvant therapy was administered in 11 patients. Mean follow-up was 41.7 months: 24.8 months for sarcomas, 70.1 months for lymphomas and 91.9 months for the epithelioid hemangioendothelioma. Among the sarcoma group, the median survival in those with negative surgical margins was 42.2 months, whereas in patients with partial resection and/or positive margins it was 14.1 months. At the end of follow-up 10 patients had died and two were alive: one with lymphoma (206 months) and the one with epithelioid hemangioendothelioma (92 months). The results were very good and similar to other recent data.<sup>2</sup> In agreement with the authors, we conclude that even partial palliative debulking surgery improves the guality of life of these patients and that the establishment of a multicenter database could enhance knowledge and surgical management of these diseases. Between January 1990 and December 2015, we operated on four patients with a primary malignant cardiac tumor, all of them sarcomas. There were three male and one female patients with a mean age of 41.75 years (19-68). The location of the tumor was in the left atrium (LA) for the first three cases and in the right atrium (RA) for the last. The first three patients with an infiltrating LA tumor (undifferentiated sarcoma, leiomyosarcoma and lipomyxosarcoma) were resected via sternotomy incision and under cardiopulmonary bypass. Our youngest patient, a 19-year-old man with an infiltrating RA intimal sarcoma, was considered unresectable and underwent a biopsy through a right thoracotomy. The four patients were operated by the same surgeon (CA). Postoperative chemotherapy was prescribed in all the patients and all four died as a consequence of local tumor recurrence and/or metastasis. The first three cases died at a mean of 19.6 months postoperatively (15, 11 and 33 months, respectively) and the fourth seven months after the biopsy (overall mean survival 16.5 months). On the basis of the small number of reported surgical series, the limited experience of a single surgeon or institution and the complexity of the surgery, we consider that these patients should be referred to a specialized center with an experienced multidisciplinary heart tumor team and no more than 1 or 2 operating surgeons. Radical, extensive and aggressive surgical resection in association with chemotherapy is the mainstay of treatment. Complete excision of the tumor with negative margins leads to a better prognosis and survival. The operation may include reconstruction of the atrial and ventricular walls and/or septum with pericardial or synthetic grafts, valve surgery, coronary bypass, a Fontan-type procedure, partial replacement of the ascending aorta and/or the pulmonary artery, associated lung resection, autotransplantation, mechanical circulatory assistance or replacement, delayed heart transplantation and other surgical strategies. Cardiac transplantation as a first alternative is not contemplated by the majority of heart teams due to concerns about recurrence and the possibility that postoperative immunosuppressive therapy may stimulate further tumor growth or a new neoplasm. There is no general consensus or guidelines on the treatment of patients with primary cardiac sarcoma and other type of primitive malignant heart tumors, and the surgical management of these patients is not well defined. An aggressive, multidisciplinary and multimodality approach with complex, highly specialized and demanding surgery markedly improves survival. Taking into consideration the adverse outcome and dismal prognosis of these diseases, in agreement with Ramlawi et al.,<sup>2</sup> we also consider that new approaches and innovative treatment strategies are required.

### **Conflicts of interest**

The authors have no conflicts of interest to declare.

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