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Short Communication

# Angiosarcomas of the Heart: What we Learned and what can we Improve

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Heart tumors are rare lesions with an atopic incidence of 0.0017 to 0.03% [1,2]. About 75% of primary cardiac tumors are benign with cardiac myxoma being the most common [3]; of the remaining 25% that are malignant, approximately up to 75% are sarcomas. Metastatic tumors are 40–100 times more frequent than primary tumors and have been frequently reported in literature [4]. Heart tumors can present with dyspnea, chest pain appear obstruction to inlet or outlet of the heart chambers or with embolism. However, a significant number of patients are asymptomatic and the cardiac tumor is incidentally diagnosed on routine echocardiogram or other imaging modalities.

Angiosarcomas, well differentiated rhabdomyosarcomas, leiomyomas, malignant fibrous histiocytoma and undifferentiated sarcomas are most frequent in the adult life while undifferentiated rhabdomyosarcomas are most frequent in the pediatric population [5,6].

Angiosarcomas are the most common malignant differentiated cardiac neoplasms that occurs with equal frequency in men and women and can affect a wide age range (from 36 months to 80 years) with a peak incidence between the fourth/fifth decade [7]. Most often angiosarcomas arises in the right atrium near the atrioventricular groove, but has been reported affecting all the others cardiac chambers as well as the pericardium. This predominant location could allow for diagnosis by endo-myocardial biopsy, generally out of reach from other types of heart malignancy.

ElBardissi et al. [8] report the experience at Boston Harvard of surgical resection for primary heart tumors. Overall survival of patients with cardiac sarcomas was dismal than the benign group especially patients were younger.

This dismal outcome was also demonstrate in a series from Simpson and colleagues [9] at the Mayo Clinic, demonstrate that the median survival for patients who underwent complete surgical excision was higher (17 months) compared to those in whom complete surgical remission could not be achieved (6 moths). For that reason an aggressive surgical treatment with a larger excision was hypnotized.

We recently reported our combined experience in treatment of primary malign heart tumors in a multicenter study [6]. In this series we report all cases operated between January 1979 and December 2012 for primary heart malignancy. Fourteen patients selected from our institution's surgical series were identified and the most common histological type was angiosarcoma (28.6%). The treatment outcome for those patients was, unfortunately, poor even if, in some case, an aggressive surgery was performed. Actually, a new multidisciplinary approach befitted our opinion especially for malignant tumors types to attempting to improve long-term survival.

For that reason a combined chemotherapy (adjuvant of neoadjuvant), together with surgical excision should be the treatment option that will be developed in the immediate future. In some case with larger involvement of cardiac chamber the cardiac auto-transplant should lead to a larger resection of the mass with "free margin" excision that perhaps could improve patient's survival [10].

Cardiac transplant for malignancy has been advocate in the past years. Even if is clear the role in case of benign tumors that cannot be resected for a large myocardial involvement, the role in case of malignancy is still debated and nowadays should be weighed case-by-case. In a recent review, Gowdamarajan et al. try to solve the dilemma without success, moreover quite all the cases of angiosarcomas reported in this review died in the first year after transplant [11].

Cardiac angiosarcomas are unfortunately silently fatal with a high risk for surgical only treatment. A new multidisciplinary approach has to be pursued in the next years able to combine chemotherapies, radiotherapies and surgery to achieve a better long term outcome for patients with cardiac angiosarcomas.

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