Cardiac rhabdomyosarcoma of the left atrium

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Contemp Oncol (Pozn) 2014; 18 (1): 73–75
DOI: 10.5114/wo.2014.40588

Primary cardiac tumours are quite rare. Only 7 cases were reported in a series of 12,000 autopsies, and its prevalence is less than 0.1% [1]. Malignant tumours account for approximately 15% of primary cardiac tumours, and sarcomas are the most common [2]. Cardiac sarcomas are quite rare and have been published as isolated case reports. Rhabdomyosarcomas account for 20% of overall cardiac sarcomas and are more prevalent in adults. Rhabdomyosarcomas most commonly originate from the ventricles. Echocardiographic (ECHO) findings can provide diagnosis, while electrocardiography and X-ray findings can be nonspecific [2]. In addition to cardiac magnetic resonance imaging (MRI), computed tomography (CT) is used for diagnostic purposes. Primary cardiac sarcomas cause death due to obstruction of blood flow through the heart. Surgical resection is the treatment of choice for all primary cardiac tumours. Additional treatment options include chemotherapy and radiotherapy. In the present report, a case of rhabdomyosarcoma in the left atrium is described.

Posteroanterior (PA) chest radiograph was performed in a 44-year-old female patient with congenital kyphosis, complaining of cough, shortness of breath and weakness. On subsequent ECHO, a 4.3 cm × 2.8 cm left atrial mass was detected beginning from the annulus of the anterior mitral leaflet and extending onto the anterior leaflet. The mass extended towards to the left ventricle inlet during each systole so that 2–3 degrees of mitral insufficiency occurred (Fig. 1A). On 19th January, 2011, the patient was operated on and a tumour was found in the left atrium with extensive infiltration into the atrial septum and adhesion of the anterior leaflet of the mitral valve, which was slightly thickened. The tumour was resected together with part of the atrial septum, and the anterior mitral leaflet was preserved. The atrial septum was reconstructed, and the mitral valve was repaired. A safety margin was confirmed by frozen section technique. Histopathological examination of the surgical specimen revealed pleomorphic sarcoma, while immunohistochemical examination showed negative staining of the neoplastic cells with caldesmon, s-100 and cd-34. Additionally, desmin was focal positive, and smooth muscle actin was positive. The histopathological and the immunohistochemical findings were consistent with rhabdomyosarcoma (Fig. 2). There were no malignant cells on the surgical margins (R0 resection). No systemic involvement was found in thoracic MRI and abdomen CT imaging. On 28th February, 2011, an adjuvant chemotherapy regimen including ifosfamide 2500 mg/m^2/day on days 1–3, mesna 2500 mg/m^2/day on days 1–3 and doxorubicin 60 mg/m^2/day on day 1 was administrated after complete resection. The patient received six cycles of chemotherapy under echocardiographic monitoring, and left ventricle ejection fraction (EF) was 60% on 18th January, 2011. Following the detection of left ventricle EF drop below 40% on the echocardiographic examination prior to the chemotherapy, doxorubicin was excluded from the combination chemotherapy due to the decrease of ejection fraction to the level of 40% on 25th February, 2011. Ifosfamide was commenced as a single agent after 28th February, 2011. Interval evaluation after six-cycles of chemotherapy with thoracic MRI, whole abdomen CT and ECHO showed no sign of disease. Fifteen months after diagnosis a newly appearing mass was seen in the ECHO, and palliative radiotherapy (total dose 6000 cGy) was administrated to the recurrent mass on 24th April, 2012 (Fig. 1B). Control ECHO was carried out after radiotherapy on 10th August, 2012, and a 44 cm × 14 cm mass-like appearance was found in the left atrium. In two-dimensional mode of ECHO, the mass was seen to be formed on the edge of the posterior leaflet and reached the base of the left atrium. Because the duration of relapse-free interval was longer than 6 months, the same chemotherapy regimen was planned. However, the patient did not accept chemotherapy treatment. Afterwards, it was decided to re-excite the recurrent tumour by the cardiovascular surgeons in the oncology council of the hospital. She was then operated on 5th August, 2012. Unfortunately, she died because of postoperative complications including renal and hepatic failure on 8th September, 2012.

The treatment options for cardiac sarcoma are summarised in Table 1. Surgery is recommended, if possible, for the treatment of tumours originating from the heart. The median survival has been reported as 6 to 12 months [3]. Although longer median survival times have been reported in patients that underwent complete surgical resection, recurrence and death are seen in the majority of patients. Low mitotic activity and receiving treatment are determined as two important factors that affect the survival rates in multivariate analyses [4]. There has been

Table 1. Treatment options of cardiac sarcoma

Treatment	Median survival	
surgery	6–12 months [3, 11]	low grade may have a better prognosis and long-term survival has been reported with complete resection [4]
adjuvant chemotherapy vs. alone surgery	17 vs. 6 months [12]	no randomized trials and anecdotal case rhabdomyosarcomas may have a better outcome with chemotherapy VAC (vincristine,dactinomycin plus cyclophosphamide), irinotecan,ifosfamide and doxorubucin ifosfamide and etoposide [8, 13–15]
radiofrequency ablation or radiation treatment	25 month [7]	for tumor recurrence (patients who underwent surgical resection)
cardiac transplantation	12 months [9]	poor results with surgical resection (not metastases) most patients have undergone treatment
cardiac autotransplantation	18.5 months [16]	increases the likelihood of major resection

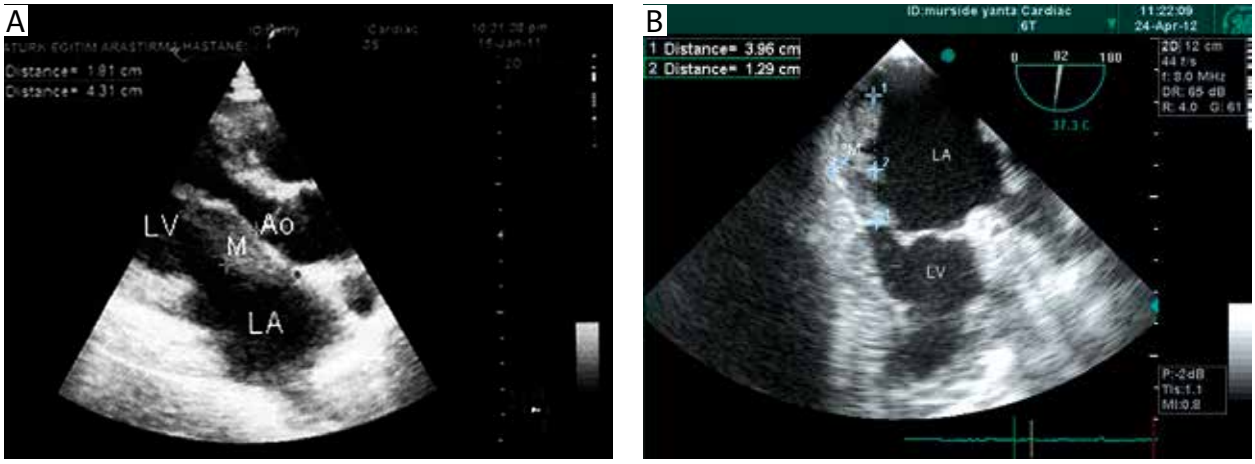

Fig. 1. A) Preoperative ECHO revealed a large tissue mass in the left atrium. **B**) TEE (transoesophageal echocardiography) test showing recurrence of the mass lesion in the left atrium

Ao – aorta, LV – left ventricle, LA – left atrium, M – tumour mass

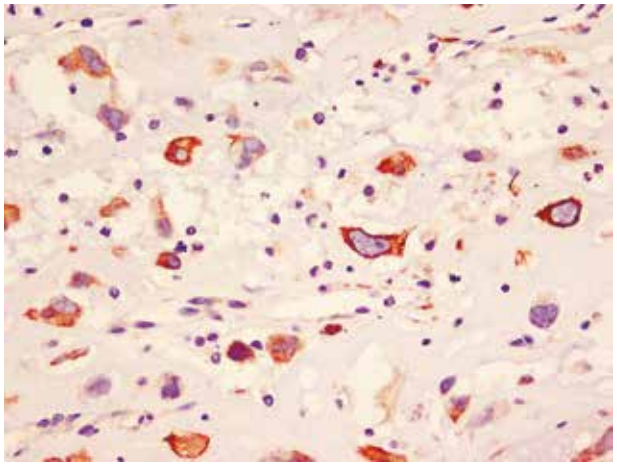

Fig. 2. There is focal, strong positivity for desmin within pleomorphic large cells. Desmin expression confirms skeletal muscle differentiation (original magnification 20×)

no randomised study conducted regarding adjuvant chemotherapy. Two retrospective series, in which anthracycline-based combination chemotherapies (cyclophosphamide, ifosfamide, vincristine, dacarbazine) were used, failed to demonstrate a significant contribution of adjuvant chemotherapy to the survival [5]. The contribution of adjuvant chemotherapy to the survival has been reported in studies with limited numbers of patients, of whom the majority had leiomyosarcoma [6]. Radiotherapy was rarely used, and the data is inadequate [7]. Non-cardiac adult rhabdomyosarcomas may have a better outcome with chemotherapy [8]. Cardiac transplantation may be attempted in patients with local disease, in whom surgical resection, chemotherapy and radiotherapy have been unsuccessful. Median survival was reported as 12 months in patients that underwent cardiac transplantation. In this series, recurrence was reported in only seven patients after the median of 27 months follow-up period [9]. Therefore, the importance of cardiac transplantation for the treatment of patients with malignant cardiac tumour remains unclear. In the present case, contrary to the ventricle-originated cases in the literature, rhabdomyosarcoma that originated from the left atrium has been reported. Complete surgical resection was performed. To date, there has not been any randomised prospective trial that supports the efficacy of adjuvant chemotherapy. However, the effectiveness of chemotherapy has been shown in two large retrospective

series in non-cardiac adult rhabdomyosarcoma. Six cycles of adjuvant chemotherapy was administered due to good performance status, younger age and the history of effective results from previous studies in the treatment of rhabdomyosarcoma originating from somewhere other than the heart. Although the efficacy of radiotherapy for treatment of cardiac rhabdomyosarcoma is not clear, similarly to the results of chemotherapy, the efficacy of radiotherapy for sites other than the heart were previously shown [10]. Thus, radiotherapy seems to be an alternative treatment option for cardiac rhabdomyosarcoma. The median overall survival of our patient was 20 months, which was superior to the survival of similar patients in the literature.

Surgical resection, when possible, is the treatment of choice for all primary cardiac tumours in which effective palliation is possible with resection of malignant tumours. There is no consensus on adjuvant chemotherapy or radiotherapy. Case reports indicate that certain histological subtypes may benefit from adjuvant therapy, although there is no standard chemotherapy regimen recommended. Studies on larger series are needed to illuminate these issues.

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Submitted: 28.02.2013
Accepted: 17.12.2013