Two young women with soft tissue tumours of the heart

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Abstract
Primary cardiac sarcomas often strike young, healthy patients and tend to have a dismal prognosis. Because of limited experience, the heterogeneous nature of cardiac sarcomas and different treatment results of patients with malignant primary tumours of the heart, the role of heart transplantation should be weighed on a case-by-case basis.

Keywords: Cardiac neoplasms • Sarcoma • Treatment • Heart transplantation

CASE REPORT

CASE 1

A 19-year old female presented at the emergency department of another hospital because of episodes of temporary loss of consciousness and chest pain. Her medical history revealed recurrent syncope on rising quickly since the age of 13 years. On clinical examination, vital signs were normal, except for a mild tachycardia (heart rate 110 bpm). Echocardiography revealed a large pericardial effusion, which could be evacuated by pericardiocentesis, resulting in 1 l of haemorrhagic fluid. After relief of symptoms, she was discharged. Additional viral and bacterial work-up of the pericardial fluid was negative. Cytological examination showed only inflammatory cells. Seven months later, she presented again after an episode of an irregular fast heartbeat while swimming. During exercise testing, she showed a non-sustained broad QRS-complex tachycardia, suggestive of a ventricular tachycardia. A computed tomography (CT) scan and magnetic resonance imaging (MRI) revealed a large mass in the infero-posterior wall of the left ventricle protruding in the left ventricular cavity and pericardial space (Fig. 1A). No distant metastases were found on CT and positron emission tomography (PET) scans. Subsequent incisional biopsies of the tumour showed a malignant mesenchymal tumour of moderate cellularity, a low mitotic index, but not otherwise specified (Fig. 1B–D). Radical surgical resection of the tumour was technically not possible because of its infiltrative growth in the left ventricle. Heart transplantation (HTx) was deemed the only therapeutic option for this patient and she was placed on the high-urgency transplantation list. Ten weeks later, an uneventful HTx was performed after a donor heart became available. Post-transplantation, routine immunosuppression therapy was started. During HTx, the surgeon was convinced that the tumour had been completely removed by excision of the native heart with the tumour-adjacent pericardium. Examination of the explanted heart revealed a tumour with a maximum diameter of 7.5 cm in the left ventricle, partly covered by the pericardium, while some parts displayed central cystic degeneration (Fig. 1E). Microscopic examination of the resection margins showed no tumour cells. Four years after HTx, she is doing well under immunosuppressive therapy without local recurrence or distant metastases.

CASE 2

A 26-year old female presented at the emergency department with a fast regular heartbeat and dizziness. Moreover, she experienced facial swelling, a non-productive cough and fatigue. She was otherwise healthy. An electrocardiogram displayed a narrow complex tachycardia of 230/min with an intermediate axis without ST-segment/T-deviations. The patient was diagnosed with an atrioventricular (AV) nodal re-entry tachycardia, and treatment with verapamil was started. A routine transthoracic echocardiogram 3 weeks later showed a large echogenic mass in the right atrium with pericardial effusion and normal left and right ventricular function. CT scanning and MRI confirmed a right atrio-atrial multinodular mass extending into the mediastinum with local thrombus formation, compressing the superior vena cava and a large pleural effusion (Fig. 2A). Paraesophageal mediastinoscopy was performed to obtain diagnostic biopsies of the tumour mass. During the same procedure, pericardial drainage took place combined with a surgical fenestration of the pericardium towards the right pleural cavity. Histopathological examination showed a haemangioendothelioma of the Kaposiform subtype. Human herpesvirus-8 (HHV-8) and human immunodeficiency virus (HIV) status were negative. HTx was considered, but the local superior caval vein obstruction made primary resection necessary. Resection of the total right atrium combined with partial resection of the left atrium and distal part of the superior caval vein was necessary, followed by reconstruction of the defect using the bovine
pericardium. Frozen sections of the left atrial margins were unfortunately positive for tumour. The definitive histopathological diagnosis of the tumour in the resection specimen showed an incomplete removal of a high-graded haemangioendotheliosarcoma, consisting of multiple, partly necrotic tumour noduli of variable size, measuring at least 9.0 cm and with extensive vasoinvasive growth (Fig. 2B–E). This final diagnosis and the obvious aggressive behaviour of the tumour excluded her from HTx. Six weeks later, PET, CT and MRI scans were performed because of numb chin complaints and the presence of a cutaneous mass on the occipital skull. Massive leptomeningeal, osseous and cutaneous metastases were seen, and the patient died 6 months after the operation.

**DISCUSSION**

Primary cardiac sarcomas often strike young and healthy patients and tend to have a dismal prognosis. Left heart sarcomas tend to be more solid and less infiltrative. They usually lead to early heart failure symptoms and metastasize late, whereas right heart sarcomas are generally angiosarcomas and tend to be bulky, show infiltrative growth and metastasize early [1]. Although complete resection is the treatment of choice for sarcomas, for many of these cardiac malignant tumours, this is not an option either because of the limited amount of the remaining myocardium and expansion of the tumour at the time of diagnosis or because of the presence of distant metastases [2]. A recent paper evaluated surgery for primary cardiac tumours in 89 children. Surgery consisted of complete resection in 70% of patients with a large benign tumour mass invading the left ventricular cavity, partial resection in 24% and HTx in 5% [3].

The prognosis of primary cardiac sarcomas is poor, while the role of chemotherapy and radiotherapy remains unclear. In general, cardiac sarcomas proliferate rapidly and cause death through widespread infiltration of the myocardium, dysfunction and obstruction of blood flow through the heart and/or distant metastases. In a recent report, survival of 18 patients with angiosarcoma was 19.5 months for localized advanced angiosarcoma, whereas for metastatic disease survival was only 6 months [4].

If local resection of the tumour is not feasible, complete excision in the setting of HTx might be a consideration. Yet, drawbacks include the lack of organ availability and the unknown effect of immunosuppression on tumour growth and spread. HTx in cardiac-tumour patients has been reported to portend a poor survival rate, with a mean survival time of 12 months [5], even after complete resection of the tumour and in the absence of extracardiac disease. However, as a minority of these patients may be long-term survivors after HTx as...
demonstrated by our first patient, this should not be categorically denied to all patients.

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