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Total artificial heart implantation after undifferentiated high-grade sarcoma excision

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CASE REPORT

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Total Artificial Heart Implantation After Undifferentiated High-Grade Sarcoma Excision

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Funds Collection G

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Background: Total artificial heart (TAH) implantation in patients with aggressive tumor infiltration of the heart can be challenging.

Case Report: We report on a patient with a rare primary undifferentiated high-grade spindle cell sarcoma of the mitral valve and in the left atrium, first diagnosed in 2014. The referring center did a first resection in 2014. In the course of 17 months, computer tomography (CT) scan again showed massive invasion of the mitral valve and left atrium. Partial resection and mitral valve replacement was not an option. We did a subtotal heart excision with total artificial heart implantation. In this report we discuss complications, risk factors, and perioperative management of this patient.

Conclusions: Patients with aggressive tumors of the heart can be considered for TAH implantation.

MeSH Keywords: Adenosarcoma • Heart-Assist Devices • Stroke

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Background

Only up to 25% of all cardiac primary tumors are malignant. Most malignant heart tumors are sarcomas: myxosarcoma, liposarcoma, angiosarcoma, fibrosarcoma, leiomyosarcoma, osteosarcoma, synovial sarcoma, rhabdomyosarcoma, undifferentiated sarcoma, reticulum cell sarcoma, neurofibrosarcoma, and malignant fibrous histiocytoma [1,2]. Symptoms from cardiac tumors primarily result from blood flow or valve obstruction. Arrhythmias can develop from intraatrial invasion, which can also lead to massive pericardial effusion and tamponade. If the tumor mass is located in the left atrium or ventricle, embolic infarction might occur [2].

Case Report

A 55-year-old man presented signs of progressive cardiac decompensation during the last 4 months due to tumorous obstruction of the mitral valve. In 2014 the patient underwent resection of a suspected myxoma (82×41×24 mm) in the left atrium. However, in the histological workup, the tumor mass, resected from the posterior mitral annulus, was diagnosed as chondrosarcoma. In 2014, our colleagues in the referring center noticed nodal calcification in the myocardium and the mitral valve. It was not possible to achieve a complete R 0 resection of the sarcoma. The Tumor Board in 2014 decided on palliative care with poor sensitivity to chemotherapy and radiation and high risk for organ toxicity. CT scan and magnetic resonance imaging (MRI) showed no signs of malignancy outside the cardiac tissue, and the chondrosarcoma found was the primary tumor.

During the course of 12 months, the patient was in a clinically stable state with combined mild mitral valve regurgitation and stenosis and good left ventricular pump function, with ejection fraction (EF) 63%. In January 2016, the patient first presented with recurring tumor mass on the posterior mitral leaflet (PML) max. 25 mm and on the anterior mitral leaflet (AML) max. 23 mm, with moderately decreasing pump function (EF 58%). In April 2016, the patient showed increasing peripheral edema and orthopnea, with no signs of angina pectoris, and NYHA III-IV. The patient presented absolute arrhythmia with atrial fibrillation, pulse 109/min, and blood pressure 82/52 mmHg. Echocardiography showed pleural effusions on both sides and distended vena cava inferior and hepatic veins. There was distinct progress of the tumor mass on the mitral valve.

Upon admission to our center, the patient presented a 40×35×32 mm mass, originating from the PML (Figure 1). There were signs of right ventricular failure due to severe mitral valve stenosis. Pulmonary arterial pressure was elevated (65 mmHg). The right ventricle was dilated, with severely reduced pump function.

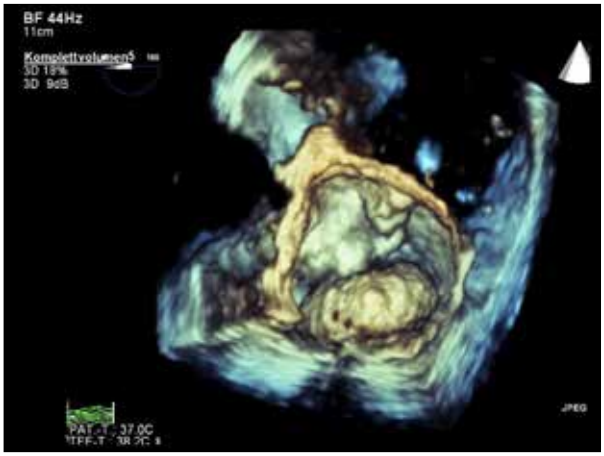


Figure 1. 3D TOE reconstruction clearly demonstrating the tumor mass originating from the PML and infiltrating the left atrial wall.

Mild aortic and tricuspid regurgitations were detected. A cardio MRI could not be performed, as the patient was unable to lie down flat due to major orthopnea. Instead, we decided to perform a cardio CT, which showed atrial roof and interatrial septum invasion. It was not perfectly clear on the CT scan whether the tumor was infiltrating the ascending aorta. A cerebral scan showed no signs of cerebral embolisms or metastasis. Due to clinical instability, the patient was transferred to our CCU after the CT. Serum creatinine levels at that point were 1.56 mg/dl and bilirubin 1.10 mg/dl. Our interdisciplinary team agreed on a repeated surgical approach to remove the sarcoma. After careful review of the scan images, we decided on the implantation of a total artificial heart, as the sarcoma had already invaded the whole atrium and the complete mitral valve annulus. A mitral valve replacement was not feasible because there would be no myocardium or endocardium to fixate the stitches in. Implantation of a self-expanding stent valve would also require an intact mitral valve annulus. Furthermore, autotransplantation was rejected for the same reasons. The severe mitral valve stenosis precluded use of a left ventricular assist device (LVAD) or biventricular assist device (BiVAD). The new approach introduced by Bruckner et al. [3] has never been performed in our center and needs experience in the surgical technique as the construction of the new atria presents major challenges because they easily collapse when not prepared properly.

We decided on the SynCardia Total Artificial Heart (TAH) (SynCardia, Tucson, AZ). As the CT scan showed no metastases and the sarcoma presented as a recurrence in the left atrium, we hoped that the patient could be listed for HTX following recovery after TAH implantation. In a similar setting, Reich et al. successfully transplanted a patient after TAH implantation, with an occult intracardial malignancy, after a cancer-free period of 14 months [4].

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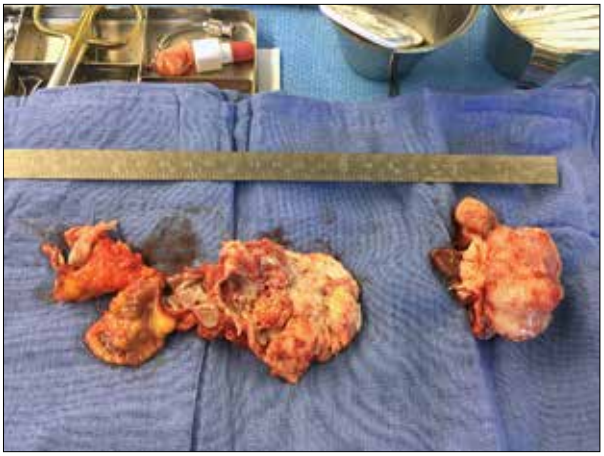


Figure 2. Intraoperative image showing the resected large sarcoma mass together with invaded parts from the left atrium and mitral annulus.

Before the operation, the patient and next of kin were informed that it would be an ultimo ratio intervention because we could not 100% foresee the invasion of the myocardium and the intraoperative circumstances. Zhang et al. demonstrated a 50% survival rate after initial surgical treatment of primary cardiac sarcomas, depending on histological grade of the tumor [5]. Surgical treatment is the most common approach to improve survival. Even as a palliative measure, reducing the tumor mass through an operation relieves orthopnea and ameliorates signs of decompensation if the patient stabilizes after the intervention.

Operative technique

After re-sternotomy, the left lung showed morphological changes and calcifications. Cardiopulmonary bypass was established via the ascending aorta and venae cavae cannulation.

Following aortic cross-clamping, the apex, ascending aorta, and pulmonary trunk were distally resected, preserving both the aortic and pulmonary valve. The right and left ventricles were cut out while maintaining functional tricuspid valves. The left atrium showed infiltration of the intraatrial septum by the sarcoma after resection of the mitral valve (Figure 2). The atrial cuffs were prepared and broad felt strips for suture support were placed. Due to extensive left atrial resection, a bovine pericardial patch was implanted to expand the pulmonary vein orifice with a 4-0 Prolene suture. Aortic anastomosis was also completed with a 4-0 Prolene suture. Anastomosis of the pulmonary artery and prosthesis was sutured with 5-0 Prolene. Functional valve movements were tested and TAH chambers were put in the AV-annuli, followed by de-aeration of both TAH chambers. Weaning of cardiopulmonary bypass was performed without complications and TAH was started. To allow postoperative tissue edema, we decided to leave the thorax open. After sufficient volume substitution, the patient was transferred to the ICU under moderate catecholamine support and NO ventilation.

Postoperative course

Postoperatively, acute-on-chronic renal failure demanded continuous hemofiltration. Five days after the initial operation, we planned a secondary thorax closure. During the intervention, substantial hematoma was evacuated from around the TAH. Extensive bleeding control was needed under continuous heparinization.

After weaning from sedation, the patient had a generalized seizure on day 10. Cerebral CT showed vast media infarction with hyper-dense blood appositions and perifocal edema with elapsed grey/white matter differentiation and early compression of the left posterior horn, with ongoing hemorrhagic transformation. Neurological examination showed the patient was comatose, with slight head movement to the left side. A right-sided facial paralysis was diagnosed.

Under continuous anticoagulation for TAH, there was a major risk for secondary bleeding due to the size of the media infarction.

Renal function did not recover during ICU hospitalization. The patient showed signs of prolonged low-output syndrome with beginning multiorgan failure. Neurological status was deemed unfavorable and after consultation with the family, the patient died on postoperative day 16 after withdrawal of support.

Histological workup showed a malignant mesenchymal neoplasm with spindle cell formations and heterologous chondroid and osseous components, with dystrophic calcifications. There were elongated, pleomorphic cell nuclei, some with high chromatin density. High mitotic activity was detected. Immunohistochemistry detected cytoplasmic CD56-expression with mild bcl2 expression. Proliferation rate was elevated in spindle cell formations up to max. 15–20% and up to 5% in the heterologous chondroid component. Chromogenic *in situ* hybridization could not confirm an amplification of MDM2-gene, and translocation of SYT-gene locus was ruled out. Hence, the classification was undifferentiated high-grade spindle cell sarcoma, grade 2 using the *French Fédération Nationale des Centres de Lutte Contre le Cancer* (FNCLCC) grading. Chondroid and osseous components are considered heterologous parts.

Discussion

Chondrosarcomas are cartilaginous tissue sarcomas. Our patient was first diagnosed with primary cardiac chondrosarcoma.

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After initial resection, relapse of the cardiac mass in the left atrium was diagnosed as early as 14 months later. After excision of the heart for TAH implantation, our pathologists found an undifferentiated high-grade spindle cell sarcoma, FNCLCC G2. With histological grading through the FNCLCC classification, survival, by major correlation, can be estimated. Cell differentiation, mitotic rate, and percentage of necrosis are evaluated in a systematic score. Undifferentiated cardiac sarcomas are most likely to be found in the left atrium. Since the 4th edition of the WHO Classification of Tumors of Soft Tissue and Bone, there is a new category of tumors that cannot be classified in earlier-defined categories. Spindle cell sarcomas fall into such a subset of sarcomas [6].

Despite left atrial infiltration, we were able to remodel a neo-atrium for TAH implantation. There were no signs of assist malfunction during ICU stay. With atrial infiltration and major excision of healthy tissue, implantation of TAH becomes more difficult. Traditional BiVAD implantation as bridge-to-transplant as a short-term solution was not an option due to severe mitral valve stenosis from the atrial mass and mitral valve infiltration. The left atrial tumor was successfully removed. Like Bruckner et al., we acknowledge that TAH implantation as bridge-to-transplantation is surgically possible even with atrial wall involvement [7].

Surgical treatment is still the criterion standard for localized cardiac sarcomas, with various perioperative survival rates, ranging from 6 to 17 months. With complete resection of the tumor mass, surgery can facilitate palliative care and even improve survival [8–11]. There is no valid data on the benefits of adjuvant chemotherapy or radiation therapy on malignant cardiac soft-tissue sarcomas. Limitations of radiotherapy are cardiac sensitivity to radiation injury and adverse effects on global cardiac functions [8]. Chemotherapy generates more promising results when applied as combination therapy [9,11]. Our patient showed good recovery after initial gross resection in 2014 for 14 months without any adjuvant therapy.

Bleeding complications due to continuous anticoagulation for device functioning remains a major risk factor in circulatory assist device programs. In a study in 13 patients with TAH, Ramirez et al. found that bleeding complications occur generally between postoperative days 1 and 3 [12]. Patients with mechanical assist devices are prone to suffer from thrombotic events [13–16], device malfunctioning, wound infections, and bleeding complications.

Conclusions

This intervention was the second attempt to help the patient with a long-term solution after first recurrence of an atrial tumor. The surgical options and possibilities were discussed at length with the patient and his family before the TAH implantation. We aimed at a potential cure and long-term survival until possible transplantation. A multidisciplinary team and truthfully informed next of kin are hugely important when taking decisions in such a high-risk case.

Competing interests

The authors report no competing interests.

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