Minimally invasive approach in the setting of a malignant primary cardiac tumor

Emre Polat, Sina Stock, Tamer Owais, Evaldas Girdauskas.

Affiliations below.

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Abstract:
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Corresponding Author:
Dr.med. Emre Polat, University Hospital Augsburg, Department for Cardiothoracic Surgery, Augsburg, Germany, emre.polat.64@icloud.com

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Design of the study: E.Polat, S. Stock, T. Owais, E. Girdauskas
Analysis and interpretation of the data: E.Polat, S. Stock, T. Owais, E. Girdauskas
Drafting the manuscript: E.Polat, S. Stock, T. Owais, E. Girdauskas
Critical revision of the manuscript: E. Girdauskas, T. Owais

Affiliations:
Emre Polat, University Hospital Augsburg, Department for Cardiothoracic Surgery, Augsburg, Germany
Sina Stock, University Hospital Augsburg, Department for Cardiothoracic Surgery, Augsburg, Germany
Tamer Owais, University Hospital Augsburg, Department for Cardiothoracic Surgery, Augsburg, Germany
Evaldas Girdauskas, University Hospital Augsburg, Department for Cardiothoracic Surgery, Augsburg, Germany
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Abstract

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Introduction

Undifferentiated high-grade pleomorphic cardiac sarcomas (also named as malignant fibrous histiocytomas) are the second most type of primary malignant cardiac sarcomas and in general widely known to be associated with a poor prognosis owing to rapidly size progression, aggressive invasion and high recurrence rate after surgical resection. (1) The early detection of the sarcoma mass is critical for planning the best surgical approach, and thus the proposal to combine surgery and histologic evaluation is a relevant strategy for planning the best treatment of UPCS.
Case Description

A 71-year-old man with known dilated cardiomyopathy and left ventricular ejection fraction of 40% without any clinical complaints (NYHA I) revealed an incidentally suspicious finding in annual cardiovascular check-up. At current state on physical examination, he was afebrile, blood pressure of 117/79 mm Hg, heart rate of 81 beats/min, respiratory rate of 13 breaths/min with oxygen saturation 97% without the need for additional oxygen supplementation. Chest radiograph did not show pulmonary venous congestion or infiltrates. An electrocardiogram showed sinus rhythm without higher-degree blockages. Transthoracic echocardiography showed a 2.8cm x 3cm discrete echo dense mass in the moderately dilated left ventricle with defined margins that are distinct from the endocardium. Due to clinical suspicion of an apical left ventricular thrombus, an oral anticoagulation therapy with Edoxaban was initiated and a follow-up echocardiography after three months was rescheduled.

Investigations

After three months the patient developed progressive dyspnea on exertion with NYHA class III and presented to our emergency department. At follow-up echocardiography, serious progression of the LV mass was revealed (5.9cm x 4.5cm) despite anticoagulation therapy. (Figure 1 A) Computed tomographic angiography revealed a sessile mass with deeply invasion of the myocardium and ruled out extra-cardiac tumors. (Figure 2, B) Abnormal vascularization of the tumor mass in the left ventricle could be ruled out by contrast echocardiography and diagnostic coronary angiography. The Positrone Emission Tomography scan detected elevated fluorodeoxyglucose
uptake of the tumor mass, which is strongly suspicious for malignancy. Cardiac magnetic resonance imaging demonstrated irregular borders of the tumor without infiltration of the adjacent myocardial wall. (Figure 2, A and C)

After a multi-modality cardiac imaging the suspicion of a primary cardiac tumor in the left ventricle was substantiated and the patient was prepared for minimally invasive cardiac surgery for tumor resection.

**Operative Management**

The patient was positioned supine with the right hemithorax tilted up to 30°. A right anterolateral mini-thoracotomy was performed through a 4-5 cm peri-areolar incision. Another port in the 4th intercostal space mid-axillary line was used for the insertion of 3D endoscope. Percutaneous femoral cannulation for extracorporeal circulation was performed by transesophageal guidance. Body temperature is maintained at around 34°C. A third port in the 2nd right intercostal space in the anterior axillary line was used for cross-clamping the ascending aorta and antegrade Del-Nido cardioplegia was delivered directly into the aortic root.

After a left atrial incision, the inspection of left ventricular cavity revealed a huge whitish yellow tumor mass with nearly occluding the entire cavum and affecting the papillary muscles which made a mitral valve replacement mandatory. After resection of the anterior mitral leaflet and both papillary muscles, the broad base tumor was peeled off en-bloc thoroughly from the anteroapical wall and placed safely in a bag to prevent the seeding of highly malignant cells into the thoracic cavity, as best as possible. (Figure 1 B) Especially, because intraoperatively frozen section analyses
revealed undifferentiated pleomorphic cardiac sarcoma, we strived copiously for achieving a R0-resection. Then, a straightforward, biological mitral valve replacement was performed. Additionally, we implemented a spine plane block to provide effective pain relief while minimizing opioid use during the perioperative period.

In accordance with our enhanced recovery after surgery program the hemodynamically stable patient was then extubated in the operating room and transferred to ICU for one night and discharge at home after 5 days without any perioperative events. Postoperative chemotherapy by multiple cycles of liposomal doxorubicin was established and further supported by protracted proton beam radiotherapy. At 2-year follow-up, no tumor recurrence could be revealed at multimodal imaging studies. (Figure 1 C)

**Discussion**

Primary cardiac neoplasms, especially undifferentiated high-grade pleomorphic cardiac sarcomas, are extremely rare and accompanied with an unfavorable prognosis owing to their aggressive biological behavior and poor overall survival. (1-2) An early diagnosis and a definition of individualized treatment concept are required to improve the prognosis of patients with primary cardiac tumors. Furthermore, histological differentiation is the key to provide insights into prognosis and survival of these patients. Characteristically, the cytology reveals heterogeneous neoplasms with various plump spindle lesions and giant features, which makes the definite diagnosis difficult. (3) Such cells often display frequent mitotic activity and nuclear polymorphism, while others undergo late programmed cell death like apoptosis. (Figure 3) (4)
Cardiac surgery aimed at radical resection is the cornerstone of the treatment. (5) Instead of the old-fashioned median sternotomy or ventriculotomy, the minimally-invasive surgery is increasingly moving to the fore in cardiac surgery and cardiac tumors are no exception, considering all the advantages like less trauma and pain, less blood loss, less wound infection, shorter rehabilitations and especially way easier redo-surgeries in case of tumor recurrence. Furthermore, in terms of tumor clearance, minimally invasive techniques with 3D endoscopes provide better visualization of the intra-cardiac structures which results in more specific excision, which in turn can be crucial for the life expectancy of the patient. If the tumor location is suitable for a minimally invasive approach, it should be the preferred method of access. However, certain other locations might not allow for thorough removal of a ventricular lesion. For experienced minimal-invasive surgeons, the access and exposure of subvalvular apparatus in cardiac tumor surgeries is a straightforward procedure, despite the rarity of these tumors. (6)

Conclusions

Primary cardiac sarcomas are neoplasms with a dismal prognosis due to their aggressive biological nature, their primary localization, and the difficulty in achieving complete surgical and pharmacological eradication. Surgery remains the gold standard technique, above all when succeeding in removing cancer cells even from the margins of the lesion. In fact, despite innovative surgical techniques and achievements of modern chemo-/radiotherapies, the survival of these patients is still poor, for what
reason minimally invasive surgical techniques with reduced trauma and way easier redo-surgeries play an important role.

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**Ethical statement:** Approval of the study by the Ethics Committee of on July the 28th, 2023 (ID number 23-0656) and also written patient informed consent was obtained.

(2) **References**


Figure 1. (A) Apical three chamber view of transthoracic echocardiography at initial cardiovascular checkup with a suspicious 28mm x 30mm echo dense mass in the left ventricle (B) Cropped apical four chamber view of transthoracic echocardiography three months later, showing inhomogeneous left ventricular apical mass (59mm x 45mm diameter), almost adherent to the mitral ring and irregular borders, occupying most of the ventricular cavity. (C) Intraoperative photograph after en-bloc resection of a huge whitish yellow tumor mass out of the left ventricle. (D) Apical four chamber view of transthoracic echocardiography two years after minimally-invasive extraction of the sarcoma showing no recurrence or rather progression.

Figure 2. (A) Giant left ventricular mass on fluorodeoxyglucosepositron emission tomography (B) Cardiac computed tomography, axial reconstruction of the the
sarcoma in the left ventricular apex through the mitral valve (C) Cardiac magnetic resonance imaging demonstrated irregular borders of the tumor mass.

**Figure 3.** A Hematoxylin and eosin staining displayed atypical spindle shaped tumor cells with cartilage formation and ossification (x 5 – x 20).