Ex vivo Tumor Resection for Primary Cardiac Sarcoma

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Cardiac sarcoma is a rare and aggressive cancer with atypical clinical manifestations. It is often detected in an advanced stage. The median survival of patients treated conservatively ranges from 6 to 12 months. Therefore, complete (R0) resection of the tumor remains an important component of a multimodality treatment for this rare but devastating malignancy [1-5]. Complete resection is technically challenging due to difficult exposure, the need for extensive resection, and complex reconstruction to preserve function. We describe a patient with a locally invasive left atrial sarcoma successfully resected using the cardiac autotransplantation technique.

PATIENT DESCRIPTION

A 36 year old previously healthy male presented to the emergency department after an effort-induced 30 minute episode of diaphoresis, palpitations, chest pain and extreme shortness of breath. He reported fever, productive cough and dyspnea on exertion and a 5 kg weight loss in the preceding 3 weeks. He was treated with antibiotics and bronchodilators with a presumptive diagnosis of acute bronchitis with little if any response. Physical examination upon admission was remarkable for a pulse of 99/minute, blood pressure 101/73 mmHg, temperature 36.6°C, room air oxygen saturation 100%, and intense pallor. Laboratory workup revealed normocytic normochromic anemia and elevated lactate dehydrogenase. Trans-thoracic echocardiogram showed a large broad-based mass filling the left atrium, prolapsing through the mitral valve during diastole, resulting in severe functional mitral stenosis. The mean pressure gradient across the mitral valve was 28 mmHg. Pulmonary arterial pressure was 95 mmHg.

The patient was taken to the operating room with a tentative diagnosis of left atrial myxoma. Exploration revealed a multilobulated broad-based 4.2 x 3 x 2 cm^3 mass invading a large portion of the left atrial wall and left pulmonary veins. Partial resection of the mass was performed. Frozen sections during surgery were interpreted as myxoma. Final pathology confirmed the tumor to be a high grade unspecified pleomorphic sarcoma.

The patient was referred to another center specializing in the management of primary cardiac tumors for further evaluation. Repeat imaging showed a locally advanced tumor with functional mitral stenosis and no evidence of distant metastasis. Due to the hemodynamic compromise, a second operation was performed: using the cardiac autotransplantation technique we completely excised the tumor [Figure 1]. The left atrium was reconstructed with a bovine pericardial patch and the left pulmonary veins with two 12 mm dacron interposition grafts. The postoperative course was unremarkable and the patient was discharged home on the 10th postoperative day. He declined adjuvant chemotherapy and, therefore, only a focused course of adjuvant radiotherapy was administered. At 3 years follow-up, the patient is alive, has a good quality of life, is in New York Heart Association Functional Class II, and was recently diagnosed with bone metastases and no local recurrence.

COMMENT

The management of primary cardiac sarcoma is challenging. The challenges include establishing an accurate diagnosis in a timely fashion, designing a patient-specific treatment plan, achieving complete resection often requiring extensive surgery,

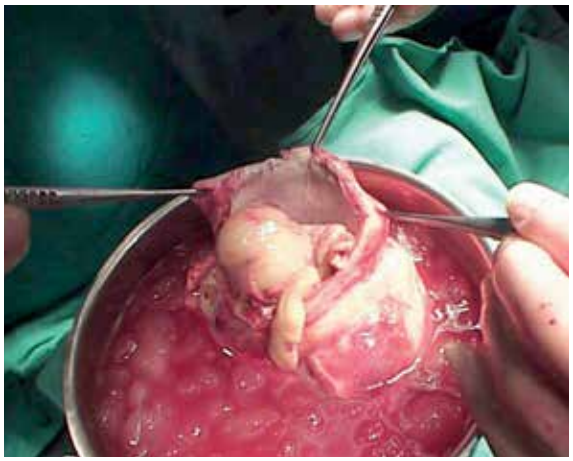

Figure 1. In surgery, the aorta, vena cava, pulmonary artery and left atrium are incised. The excised heart is immersed in ice-cold solution to minimize ischemic injury. The tumor, seen here as a multilobulated mass in the left atrium adjacent to the anterior leaflet of the mitral valve, is resected. Next, reconstruction of the left atrium and pulmonary veins is performed and the heart is autotransplanted to gain reperfusion

and integrating neoadjuvant and adjuvant therapy [1-5]. The best and most efficient way of achieving these goals is to refer the patient to a specialized center for evaluation and treatment by a multidisciplinary team including radiologists, cardiologists, medical and radiation oncologists, pathologists and surgeons [3].

Echocardiography is the most useful, first in-line, non-invasive diagnostic test for intracardiac masses [1-5]. Seventy-five percent of primary cardiac tumors are benign (mostly myxomas) with rather typical echocardiographic features [1,5]. A high index of suspicion for malignancy should arise when a patient presents with constitutional signs (e.g., weight loss) and the echocardiographic finding of a large, broad-based infiltrative immobile mass. In such cases additional imaging, particularly cardiac computed tomography (CT) and magnetic resonance imaging, are very useful. Radiologic features that are highly suggestive of malignancy include tumor enhancement and local invasion. Positron emission tomography (PET)/CT is only useful to assess metastatic disease, and with relatively low sensitivity. Coronary angiography is indicated only if surgery is contemplated [1,3,5]. Preoperative percutaneous biopsy is rarely performed because of the risk of systemic tumor embolization. Trans-esophageal or trans-thoracic CT-guided needle biopsies may be used in cases of mediastinal invasion. Intraoperative frozen section analysis can be misleading since these tumors can comprise histological features of myxomatous changes [1,5]. The most common histology is unspecified sarcoma, found in up to 35% of cardiac sarcoma cases [1,4-5]. However, prognosis is not affected by tumor histology and is related more to the anatomic site [1-3]. Most commonly, primary cardiac sarcomas are classified as pulmonary artery, right heart, or left heart tumors. The prognosis is best for pulmonary artery sarcomas, worst for right heart sarcomas, and intermediate for those of the left heart [1-5].

In hemodynamically stable patients a course of neoadjuvant chemotherapy should be the first step. The most effective regimen includes a combination of high dose adriamycine and ifosfamide [3-5]. Repeat imaging to assess local and distant control is mandatory. In hemodynamically unstable patients, such as our patient, surgery must be performed early. Almost invariably adjuvant chemo- and radiation therapy should follow surgery [1-5]. Complete surgical excision with microscopically negative margins of the tumor (R0) is mandatory for an optimal outcome [1-5]. Cardiac explantation, ex vivo tumor resection, cardiac reconstruction, and autotransplantation is an extremely effective technique for primary sarcomas of the left heart [1,3,5]. In experienced centers, these procedures can be performed with an acceptable operative morbidity and mortality. It is important to avoid a concomitant pneumonectomy due to unacceptable procedural mortality [5]. Thus, as in our case, pulmonary vein invasion should be managed by resection and reconstruction. Since primary cardiac sarcoma is a rare disease, data regarding the optimal treatment protocol are inconclusive. However data derived from several single-center series and isolated case reports, including ours, strongly suggest that a patient-tailored multidisciplinary approach, comprising radical (R0) resection, chemotherapy and radiation therapy provided in specialized centers, results in a significant improvement in patient survival and quality of life.

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