right heart sarcoma protocol. In this particular case, the patient with past medical history of multiple myeloma presented with a large right atrial mass that was not suggestive of a benign mass. Therefore, our diagnostic algorithm dictated that he undergo preoperative endomyocardial biopsy.

Transvenous endomyocardial biopsy has been reported as a safe procedure that can be easily performed to obtain tissue with less morbidity and mortality than a thoracotomy [3–5]. Guidance with transesophageal echocardiography is currently advised by the American Heart Association, the American College of Cardiology, and the European Society of Cardiology [6]. For our patient, adequate tissue sampling through a transvenous approach did allow for accurate pathologic diagnosis and the formulation of a tailored therapeutic regimen. Clinical complications from endomyocardial biopsy are rare and are estimated to be around 1% to 2% with death occurring 0.2% of the time [7]. The most common complication from endomyocardial biopsy is sampling error and missing pathology that is focal in nature or potentially limited to the left ventricle [8]. Thus, it is important to obtain multiple tissue fragments and strongly consider the clinicopathologic presentation and disease entity.

Once biopsy confirmed the mass was not a sarcoma but instead a plasmacytoma, we began radiation and ruled out the need for chemotherapy or surgery. The patient’s radiation regimen included 5 fractions of 180 cGy during his brief stay in the hospital. He showed prompt improvement clinically and even had a 1-cm reduction in tumor size in about a week. He was breathing well and walking around without any problems, and was discharged with appointments for further radiation.

Transvenous endomyocardial biopsy serves as a safe and effective modality to diagnose cardiac tumor type. Pathologic tumor diagnosis allows for implementation of the most efficacious therapy going forward. In this case, the utility of transvenous endomyocardial biopsy allowed for the diagnosis and optimal treatment of a patient with a large right atrial plasmacytoma.

References

Left Atrial High-Grade Undifferentiated Pleomorphic Sarcoma Protruding Through the Mitral Valve

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Primary cardiac tumors are uncommon. Malignant neoplasms account for 25%, including 75% of cardiac...
A 53-year-old female complained of exertional dyspnea and orthopnea. Chest computed tomography revealed a mass within the left atrium. Echocardiography confirmed a bilobed left atrial mass protruding through the mitral valve orifice. The tumor was completely resected and was histologically diagnosed as a high-grade pleomorphic sarcoma. A 13-month follow-up was achieved without any recurrence on magnetic resonance imaging. (Ann Thorac Surg 2014;98:2227–9) © 2014 by The Society of Thoracic Surgeons

Primary cardiac tumors have an incidence ranging from 0.001% to 0.03% in autopsy series [1]. Only 25% are malignant, with sarcomas as the most common subtype (75%). Complete surgical resection is the standard treatment. The benefit of adjuvant chemotherapy, radiotherapy, and other strategies is unknown. Patients with cardiac sarcomas have an unfavorable prognosis with a mean survival of 11 months [2]. Although malignant tumors are usually detectable with echocardiography, the diagnosis of sarcoma is rarely made until after surgical intervention. We describe a case of high grade pleomorphic sarcoma.

A 53-year-old Caucasian woman presented to her general practitioner with a 3-week history of gradual onset of dyspnea on exertion, orthopnea, palpitations, fatigability, and presyncope. He prescribed a chest scan when he saw bilateral pleural effusion on the chest radiograph. She also complained of lower limb edema 4 months before, a visual blurring, and a transient lateral homonymous hemianopsia 2 months before admission. She had no past medical history and was not taking any medications or using tobacco, alcohol, or drugs.

Systemic examination revealed a temperature of 102°F, a preserved hemodynamic status, respiratory rate of 30 breaths/minute, and oxygen saturation of 93%. Her examination was notable for jugular venous distension, bilateral lower-extremity edema, bibasal crepitations, and a grade 2/6 mid-diastolic heart murmur. Her electrocardiogram showed sinus rhythm of 90 beats/minute, without any significant abnormalities.

Chest scan with intravenous contrast media injection confirmed bilateral pleural effusions with pulmonary atelectasis and revealed a mass within the left atrium (Fig 1). Cerebral scan showed small cerebral embolisms. A transthoracic echocardiogram showed a large mobile bilobed mass arising from the lateral wall of left atrium, measuring 60 mm and 30 mm (Fig 2). The tumor was prolapsing across the mitral valve orifice into the left ventricle in diastole. There was a mild mitral regurgitation and moderate tricuspid regurgitation with a mild pulmonary hypertension. The systolic function was preserved.

The patient was immediately evaluated by the cardiovascular surgery service for tumor excision. The preoperative coronary angiogram was normal.
The patient had the tumor excised through median sternotomy and mitral annuloplasty (Carpentier-Edwards Physio II; Edwards Lifesciences SAS, Irvine, CA) was performed because of a mitral annulus enlargement leading to mitral regurgitation upon intraoperative testing. The mitral valve was not involved with the lesion and postresection transesophageal echocardiogram showed no significant mitral valve abnormality, a good left ventricular function, and no evidence of any remaining tumor. The lesion was macroscopically described as a bilobed tumor measuring 6 and 3 cm in their longer dimensions and weighing 52 g (Fig 3). The histologic analysis revealed a neoplasm characterized by dense areas of mainly spindle cells in a myxoid background (Fig 4A). The tumor cells showed nuclear atypia and mitotic activity (Fig 4B). There were foci of necrosis. The histologic diagnosis was high-grade pleomorphic sarcoma. Surgical excision appeared complete because the implantation base was not affected. This form is very rare, difficult to classify, and could be an equivalent of cardiac aortic intimal sarcomas.

The patient recovered without complications and was discharged to a convalescent home on postoperative day 7 with recommendation for follow-up. She refused adjuvant chemotherapy.

No evidence of tumor dissemination was found on extension workup, except for the small cerebral embolisms. No recurrence of neurologic symptoms occurred after discharge. Despite successful management and favorable evolution of the tumor implantation base and detected metastases, only palliative surgery can be performed. We present a rare case of high-grade pleomorphic cardiac sarcoma successfully removed, without invasion of the tumor implantation base and detected metastases. Despite successful management and favorable evolution after 13 months, close follow-up is mandatory because of the overall poor prognosis of cardiac sarcoma.

Comment

Primary malignant cardiac tumors are uncommon with angiosarcoma as the most common type [3]. Undifferentiated pleomorphic sarcomas are rare (only 6%) and usually found in the left atrium with an infiltrative pattern [4, 5]. Primary cardiac tumors are locally aggressive, proliferate rapidly, and up to 80% are metastatic at diagnosis. The prognosis is poor with a median survival of patients who underwent surgery with and without metastasis of about 5 and 15 months, respectively [1, 4]. Because there are only a few limited studies on the subject, no standard treatment strategy has been defined. The cause of death is, in most cases, local recurrence and progressive tumor enlargement. Although aggressive resection of cardiac sarcoma is challenging, some studies demonstrate that complete tumor removal is the best method for improving survival [1, 6, 7].

When the tumor infiltrates the left atrium more deeply and to a greater extent, resection and reconstruction is relatively difficult. In such cases, autotransplantation is an option allowing complete resection and repair [8]. In many cases, cardiac sarcoma is so widespread that only palliative surgery can be performed.

The effectiveness of chemotherapy and radiotherapy for cardiac sarcoma remains controversial. However, combining several chemotherapies seems more effective than single-agent therapy, and postoperative radiotherapy may improve survival [1, 3–4].

In our patient case, the implantation base of the tumor was not affected and there were no confirmed metastases, so we can think the complete tumor removal will significantly improve the chances of survival, despite the lack of adjuvant therapy.

References