especially if the RA is not massively dilated, with surgical procedures being reserved for patients with mechanical complications (severe TR, grossly dilated RA, and so forth) or uncontrolled, symptomatic dysrhythmias. There are some, however, who recommend surgical reduction atriplasty even for asymptomatic patients to prevent the risk of future thromboembolic complications and dysrhythmias [1].

References


Myxoid Fibrosarcoma of the Left Atrium and Ventricle in Pregnancy

Wei Zhang, MD, Xiangxiang Zheng, MD, Buqing Ni, MD, Weidong Gu, MD, Hao liang Sun, MD, Yongfeng Shao, MD, and Shijiang Zhang, MD

Department of Thoracic and Cardiovascular Surgery, the First Affiliated Hospital, Nanjing Medical University, Nanjing, China

Myxoid fibrosarcoma is rare in adults, and presentation during pregnancy is extremely rare. We report a woman who presented at 20 +1 weeks of gestation with a 1-month history of swollen legs, exertional dyspnea, and cough with frothy sputum. Dual-source computed tomography and transthoracic echocardiography showed a left atrial mass. The dead fetus was delivered by caesarean section. Two weeks later, she underwent resection of the intracardiac mass and mitral valve replacement. Histopathologic examination showed myxoid fibrosarcoma. The patient refused postoperative adjuvant chemotherapy and radiotherapy. After 1 year, she had no apparent complications, and echocardiography showed no evidence of recurrence.


Myxoid fibrosarcoma is a rare type of cardiac fibrosarcoma that arises from cardiac myxoma. Presentation of myxoid fibrosarcoma during pregnancy is extremely rare.

A 25-year-old woman (gravida 1, para 0) was admitted to hospital at 20 +1 weeks of gestation with a 1-month history of swollen legs, exertional dyspnea, and cough with frothy sputum (New York Heart Association class III heart failure). She had no significant medical or family history. Physical examination revealed a blood pressure of 110/90 mm Hg and a heart rate of 120 beats/min. There was a palpable precordial thrill. Cardiac auscultation revealed an apical pansystolic murmur (grade 4/6) and an early diastolic murmur at the left sternal edge. Roentgenology of the chest showed cardiomegaly, with a cardiothoracic ratio of 0.63. Echocardiography revealed an echogenic mass, 27 × 17 mm, in the left atrium, attached to the middle of the anterior mitral valve leaflet (Fig 1A). The mass was observed to prolapse through the mitral valve, causing severe eccentric mitral regurgitation and severe mitral stenosis. The patient had severe pulmonary hypertension, with a pulmonary artery pres sure of 94 mm Hg. There was also an echogenic mass, 58 × 23 mm, in the left ventricle, extending from the side wall to the subaortic position, which caused outflow tract obstruction resulting from the rapid blood flow in the systolic period (Fig 1A). The left ventricular ejection fraction was 50%. Dual-source computed tomography also showed masses in the left atrium and ventricle (Fig 1B).

After giving written consent, the patient underwent caesarean section to deliver the fetus, which had died because of placental infarction. Two weeks later, she underwent an operation for excision of the intracardiac mass. Intraoperative exploration revealed a mass originating from the left ventricular wall that extended to the mitral valve and into the left atrium. The ventricular part of the mass measured 40 × 40 × 40 mm, and it blocked the ventricular outflow tract. The atrial part of the mass measured 20 × 20 × 30 mm. Only partial resection of the mass was possible (Fig 2A). The mitral valve was replaced with a biologic prosthetic valve (Edwards 25). Post-operative histopathologic examination of the resected mass showed myxoid fibrosarcoma (Fig 2B). Immuno-histochemical staining was negative for actin, desmin, S-100, and CD34; it was positive for vimentin, smooth muscle actin (SMA), and Ki-67.

The patient was discharged on the 20th day after the operation. She refused adjuvant chemotherapy and radiotherapy. Surprisingly, she was still alive after 1 year without any particular complications, and echocardiography did not show recurrence of the mass.

Comment

Primary fibrosarcoma of the heart is a malignant mesenchymal tumor that accounts for only about 3%
of malignant cardiac tumors. This tumor is differentiated from fibrosarcoma by the presence of giant cells and a storiform or whorled pattern of spindle cells [1].

The clinical findings associated with fibrosarcoma are nonspecific, and they vary according to the location, size, mobility, and surface characteristics of the tumor. In our patient, the mass obstructed the mitral valve and the left ventricular outflow tract, causing congestive heart failure. Cardiac myxoid fibrosarcoma may also cause embolic events. Although our patient had no symptoms of embolism, there was placental infarction secondary to tumor emboli. The elevated central venous pressure and presumed low cardiac output may also have played a role in fetal demise.

Management should be decided on an individualized basis. Surgical excision is the gold standard for the treatment of symptomatic or life-threatening cardiac tumors. In pregnant women, such a procedure carries high risks for the fetus, and the pregnancy is usually terminated early. In this case, we performed cesarean section to deliver the dead fetus. There is controversy regarding the appropriateness of a cardiac surgical procedure when the fetus is viable. Given the potential complications of cardiac tumors, surgical excision should be considered in all pregnant patients. However, treatment decisions should be individualized according to the clinical context for each patient [2]. Cardiac sarcomas generally have a worse prognosis than sarcomas in other locations because complete resection is not always possible and because of the importance of the adjacent structures. The reported median survival time is 6 to 9 months [3]. Our patient remains alive without any particular complications 1 year after the operation, and she continues to be followed up.

References