A High-Grade Pleomorphic Sarcoma in Left Atrium

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CASE PRESENTATION

A 22-year-old man, with no significant past medical history (without fever, night sweats, and weight loss), presented with a week history of flustered and shortness of breath. He also complained of heaviness and tightness in the chest on exertion. Physical examination showed that the heart rate was arrhythmias (170 beats/min), blood pressure was 100/60 mmHg, and exhibited a systolic blowing murmur (grade 4 of 6). The breathing sounds on both lower lungs were slightly coarse, with a respiratory rate of 18 breaths per minute. His abdomen was soft without rebound tenderness, liver and spleen were not palpable, and without obvious edema in lower extremities. Laboratory investigations revealed a lactate dehydrogenase of 542 IU/liter, \(\alpha\)-hydroxybutyrate dehydrogenase of 419 IU/liter, with the most common values within the normal limits. Electrocardiography showed an evidence of multifocal atrial tachycardia. Echocardiography revealed a 43 × 37 mm heterogeneous mass on the lateral wall of left atrium with floculent echoes, and the mass was very mobile and was oscillating into the mitral valve ostium during diastole, even reached the left ventricle through the mitral annulus (the mitral valve was intact). And there was a small amount of effusion in the pericardial cavity (Fig. 1). The 320-slice computed tomographic (CT) scan revealed a dilated left atrium, with a heterogeneous mass (20 × 55 mm) attached to the lateral wall of left atrium, occupied almost the whole left atrium (Fig. 2). And there was a massive amount of thromboembolism in the left inferior pulmonary vein. The mass blood was mainly composed by circumflex branch of left coronary artery and bronchial artery with minor. And the 320-slice CT also showed an osteolytic bony destruction from the sixth thoracic vertebra (T6) to T10, a compression deformity of the T10 (Fig. 3).

Lung cancer, pulmonary infarction, myocardial infarction, and coronary artery disease were not observed. The soft tissue of left temporal area and right buttock was widespread metastasis through underwent a combined computed positron emission tomography and CT examination by an integrated positron emission tomography/CT system. The histologic sections confirmed the diagnosis of high-grade pleomorphic sarcoma (Fig. 4). Immunohistochemical analysis demonstrated that the tumor cells were strongly positive for vimentin and CD68, and moderately positive for Ki-67(+) and CD34(+), the other immunohistochemicals, such as S-100, desmin, anaplastic lymphoma kinase (ALK), nestin, and CD117 were all negative. Based on the physical and radiologic findings, it is clear that this is a case of cancer with metastasis to multiple organs, the patient was undergone multiple sessions of radiotherapy and chemotherapy, and no recurrence was observed during follow-up.

COMMENTS

Cardiac tumors have an incidence between 0.17% and 0.19%. Primary cardiac sarcomas are more extreme rare tumors with a prevalence of 0.001% to 0.03% in autopsy series.1,2 To the best of our knowledge, approximately 40 cases have been reported worldwide so far, high-grade pleomorphic sarcoma in left atrium was more extremely rare reported. High-grade pleomorphic sarcoma, was also denoted as malignant fibrous...
histiocytoma, which by definition is microscopically composed of atypical elongated fibroblast-like cells with interspersed collagen and the classic herringbone pattern. It is frequently located in the left atrium, mainly in young adult women, the age of patients at diagnosis ranges from 14 to 77 years old, and the median age is in the fourth decade. Patients with high-grade pleomorphic sarcoma typically present with cardiac symptomatology that may include arrhythmias, obstruction to blood flow and valve function, or symptoms of heart failure, but the symptom of metastatic sites was not obvious. The main factors have been postulated for the infrequency of cardiac metastasis: the strong action of the myocardium, metabolic peculiarities of striated muscle, rapid blood flow through the heart, and lymph flow moving away from the heart. Enhanced CT and echocardiogram can highlight the disease, and cardiac magnetic resonance imaging approach is also excellent, especially high-resolution CT has been widely applied and provided accurate clinical diagnosis. The standard procedure therapy for eligible patients with cardiac sarcoma without metastasis at diagnosis is a complete surgical removal of the tumor. When extracardiac metastasis is identified, surgical resection is reserved for palliative indication. Without surgical resection, patients with sarcoma typically live for 9 to 12 months despite any radiotherapy and chemotherapy.

REFERENCES