Cardiac Angiosarcoma with Ruptured Right Atrium Diagnosed by Echocardiography*

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A 41-year-old man was admitted to our hospital complaining of right-sided chest pain. Two-dimensional echocardiographic examination revealed a ruptured right atrium and a localized pericardial effusion. Color Doppler echocardiography and contrast echocardiography showed blood flow from the right atrium into pericardial effusion, which was confirmed at emergency surgery. The pathologic specimen from the right atrium showed a spindle cell angiosarcoma.

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Cardiac Angiosarcoma is a rare tumor. Little is known about its natural clinical course, and it is difficult to make the diagnosis prior to autopsy. However, using echocardiography, it has become easier to diagnose cardiac tumors. Recently, we encountered a patient with right atrial rupture caused by a cardiac angiosarcoma that was demonstrated by echocardiography.

CASE REPORT

A 41-year-old man was admitted to our hospital complaining of the sudden onset of right-sided chest pain. He had been well until four days previously. A chest roentgenogram showed a right pleural effusion, with the liquid level at the fourth rib. The pleural effusion was bloody, containing 7.7 g/dl of hemoglobin and was cytologically negative for malignant cells. About 1,000 ml of pleural fluid was removed at thoracentesis.

A follow-up chest roentgenogram obtained about one week later revealed enlargement of the right side of the cardiac silhouette. On physical examination, his temperature was 37.3°C, the pulse rate was 110/min, and the blood pressure was 94/50 mm Hg. Pulsus paradoxus and distention of the jugular vein were not present. Inspiratory crepitations were not heard. A Levine I/6 systolic ejection murmur was heard along the left sternal border. The abdomen was normal. Laboratory tests gave the following data: the hematocrit was 33.8 percent, the white blood cell count was 6,600/μm³, the platelet count was 214,000/μm³, and the cardiac enzyme values were normal. An electrocardiogram showed sinus tachycardia at a rate of 122 beats/min. Computed tomography of the chest showed a low density mass adjacent to the pericardium at the right atrium. The apical four-chamber view of two-dimensional echocardiography showed a localized moderate pericardial effusion over the right ventricle and right atrium. In this effusion, there was an immobile mass adjacent to the pericardium that was 4 cm in diameter (Fig 1). The left side of the heart was normal except for hyperdynamic function. The view obtained through the right sternal border showed a 1-cm fissure in the right atrial wall. Color Doppler examination revealed blood flow from the right atrium to the pericardial effusion through this fissure. Contrast echocardiography confirmed these findings. Contrast initially filled the right atrium, and later moved through the right atrial fissure into the pericardial effusion (Fig 2). In the anterior view of contrast right atrial radiography, a narrow-width jet from the right atrium into pericardial effusion was noted. The right-sided pressures determined by Swan-Ganz catheter were normal. Right coronary angiography showed stenosing of the contrast medium into the pericardial space.

FIGURE 1. Two-dimensional echocardiogram. The apical four-chamber view showed a localized pericardial effusion (PE) over the right ventricle (RV) and right atrium (RA). An immobile mass (M) was present in the effusion. LV = left ventricle; LA = left atrium.

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In cases of type 3 cor triatrium, however, diagnosis may be difficult because no symptoms or abnormal hemodynam- ics may be present. All four cases reported by Niwayama' were diagnosed by postmortem necropsy. In our patient, as the typical manifestations of pulmonary venous obstruction were absent, diagnosis of the anomaly would not have been made if a CT for examination of the patient's lung cancer had not been performed. The CT revealed the anomaly and, in this case, CT proved to be a very useful diagnostic imaging technique.

Bisset and his coworkers' reported a case in which magnetic resonance imaging (MRI) provided a specific diagnosis of cor triatrium, and they concluded that MRI was effective in diagnosing the anomaly. To the best of our knowledge, however, no case in which CT revealed the anomaly has previously been reported.

Computed tomography and MRI may be helpful tech- niques in the diagnosis of cor triatriatum. Given the increasing number of patients on whom CT and MRI are performed, more cases of cor triatriatum may be diagnosed in adults.

Cardiac Angiosarcoma (Satou et al)
Cardiac rupture of the right atrium was diagnosed, and surgery was performed. Through a median sternotomy, hemopericardium was detected and evacuated. Adhesions between the pericardium and the epicardium had localized the effusion over the right atrium and ventricle. There was brisk venous bleeding through the rupture in the right atrium. The pericardial mass on the pericardium, which had been detected by echocardiography and computed tomography, was partially resected. Repair of the right atrium using a patch was impossible because the wall was too thin, so reconstruction with pericardium was performed.

Light microscopic examination of the resected right atrium specimen revealed a spindle cell sarcoma. The tumor cells were pleomorphic and lined the vascular channels with more than one layer. The patient initially recovered, but about the fourth postoperative week, the chest roentgenogram demonstrated multiple micronodular shadows throughout both lung fields and blunting of the left costophrenic angle. Dyspnea and hemoptysis developed, and the patient died on the 73rd hospital day. Neither chemotherapy nor radiotherapy had been performed because of his poor general condition.

At autopsy, the heart weighed 285 g. No gross evidence of tumor was seen in any of the chambers. The epicardium of the right atrium and the right ventricle were irregular and gray. Histologically, the tumor cell was shown to invade the epicardium of the right atrium and the right ventricle. There was a bloody pleural effusion of approximately 1,500 ml on the left and another of 300 ml on the right. Both lungs were diffusely studded by multiple nodular red-purple metastases that measured up to 1 cm in diameter. No metastasis was found to any other organ.

Immunohistochemically, the tumor cell was positive to Ulex europeus agglutinin I (UEA I) and negative factor VIII antigen.

**DISCUSSION**

The incidence of primary cardiac tumors in a large-unselected autopsy series was less than 0.05 percent (of which angiosarcoma was 10 percent). Angiosarcoma is most commonly located in the right side of the heart, especially in the right atrium, and the patient is usually a middle-aged-man. Clinical symptoms usually arise due to superior vena cava obstruction, hemopericardium, or pleural effusion. Metastasis occurs most frequently to the lungs, liver, thoracic lymph nodes, bone, and adrenal gland. Most patients die within a few months of the onset of symptoms.

Two-dimensional echocardiography generally shows this tumor as a large mural mass that replaces the atrial wall and often protrudes into the chamber, sometimes completely filling it.

Three cases of spontaneous right atrial rupture with angiosarcoma had been reported previously. However, to our knowledge, this is the first report of right atrium rupture diagnosed by color Doppler echocardiography and contrast echocardiography.

**REFERENCES**