Asymptomatic Cor Triatriatum Incidentally Revealed by Computed Tomography*

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We report a case of asymptomatic cor triatriatum in a 75-year-old man in whom the anomaly was incidentally revealed by computed tomography (CT). To our knowledge,

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this patient is the oldest case reported and the only such case in which the anomaly was demonstrated by CT.

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Cor triatriatum is a rare congenital cardiac anomaly, usually seen in childhood, and only rarely seen in adulthood, in which a fibromuscular membrane divides the left atrium into two chambers, one receiving pulmonary venous return and the other containing the left atrial appendage and the mitral valve. In its classic form, the two chambers communicate through a small opening, which results in obstruction of pulmonary venous return. Usually, diagnosis is made when two-dimensional (2-D) echocardiogram or angiography demonstrate the subdividing membrane. We report an unusual case of asymptomatic cor triatriatum in a 75-year-old man in whom this anomaly was incidentally revealed by computed tomography (CT) during an examination for lung cancer.

CASE REPORT

A 75-year-old man presented for investigation of abnormal chest roentgenographic findings. He was asymptomatic, and results of physical examination were normal. Chest roentgenogram showed a mass in the left upper lobe, but no evidence of cardiomegaly or pulmonary congestion was demonstrated. A transbronchial lung biopsy was performed, and the histologic section revealed an adenocarcinoma. Chest CT confirmed this tumor and, at the same time, incidentally revealed an abnormal chamber behind the left atrium (Fig 1). This abnormal chamber appeared to communicate with the left atrium through an opening in an anomalous septum. A 2-D echocardiogram demonstrated an echo-dense membrane that divided the left atrium into two chambers (Fig 2). The venous return phase of pulmonary arterial angiogram demonstrated the subdividing membrane, and the diagnosis of cor triatriatum was established. No complicating cardiac anomaly was demonstrated. Cardiac catheterization revealed normal pulmonary arterial pressure (25/12 mm Hg; mean, 16 mm Hg) and capillary wedge pressure (8 mm Hg).

The chest was surgically explored through the left fifth intercostal space on July 7, 1989, and a left upper lobectomy and lymph node dissection for lung cancer were performed. However, in view of the patient's age and the presence of normal pulmonary arterial pressure and capillary wedge pressure, surgical correction of the cor triatriatum was not performed. Postoperative course was uneventful.

DISCUSSION

In cor triatriatum, the onset and severity of symptoms depend on the size of the opening between the accessory chamber and the true left atrial chamber. Loeffer and Gasul et al classified variations of the anomaly into the following three types according to the size of the opening: type 1, with no opening; type 2, with a small opening; and type 3, with a large opening.

Usually, the opening is small and symptoms produced by the obstruction of pulmonary venous return first appear in childhood. Only a few patients in whom the opening is large survive into adulthood. Niwayama reviewed 37 patients with cor triatriatum and found only four patients who could be classified as having type 3. The oldest patient with the anomaly whose case has ever been reported was a 70-year-old woman.

The reason that our patient presented no symptoms and his hemodynamics were normal, even when he had reached the age of 75 years, may be that the opening was too large to obstruct pulmonary venous return. Thus, our patient is a case of type 3 cor triatriatum, and the oldest patient who has been reported in the world literature.

Previously, cor triatriatum has been difficult to diagnose and, in most cases, the anomaly was revealed either by necropsy or during operation. Recently, there have been many reports of cases in which the classic form of the anomaly was diagnosed before operation; in these cases, 2-D echocardiogram and pulmonary arterial angiography were useful diagnostic techniques.
In cases of type 3 cor triatriatum, however, diagnosis may be difficult because no symptoms or abnormal hemodynamics 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 triatriatum, 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 techniques 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.

REFERENCES
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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 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 1/6 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/μl mm, the platelet count was 214,000/cu mm, 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 staining of the contrast medium into the pericardial space.

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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.