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Giant Right Atrial Myxoma Mimicking Hepatic Cirrhosis: A Case Report

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ABSTRACT

Cardiac myxomas are rare benign tumors of the heart. The growth rate of these tumors remains unknown. Right atrial myxoma can simulate nonspecific constitutional symptoms, such as remittent or lasting fever, weight loss, and chronic anemia, and may escape timely diagnosis until the development of severe complications such as pulmonary hypertension due to embolism from fragments originating from the tumor mass or blockage of the right atrioventricular ostium or Budd-Chiari syndrome with acute abdominal pain. We present a case of a giant right atrial myxoma mimicking hepatic cirrhosis in a 52-year-old man.

BACKGROUND

Primary tumors of the heart are rare in reported autopsy series. Considering all age groups, the most common cardiac tumor is the myxoma. Myxomas may arise in any of the 4 chambers or, rarely, on the heart valves. Because of nonspecific symptoms, early diagnosis may be a challenge. About 90% are located in the atria, and right atrial location is seen less often than left atrial location, with a left-to-right ratio of 4:1. The majority of patients are usually in the age group of 30 to 60 years [Akyildiz 2006].

Signs and symptoms range from nonspecific and constitutional to sudden cardiac death, according to the location of the myxoma in the cardiac chambers. In about 20% of cases, myxoma may be asymptomatic and discovered as an incidental finding. Atrial myxoma can simulate mitral stenosis, endocarditis, mitral regurgitation, and collagen vascular disease [Reynen 1955]. Budd-Chiari syndrome characterized with hepatic venous outflow obstruction caused by large right atrial myxomas is a relatively uncommon clinical condition reported in the literature [Anagnostopoulos 2004]. It has been emphasized that myxomas present with signs and symptoms of right heart failure and pulmonary embolism [Murayama 2001; Grebenc 2002; Anagnostopoulos 2004]. We report a case of giant right atrial myxoma mimicking hepatic cirrhosis.

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CASE REPORT

A 52-year-old man was admitted to our clinic with constitutional symptoms of palpitation, fatigue, shortness of breath, and myalgia. He had been hospitalized at another clinic 4 years earlier with a diagnosis of cirrhosis caused by chronic alcohol usage. The hepatic enzymes had been mildly elevated at that time. Upper gastrointestinal endoscopy showed esophageal mild-grade varices and ulcers at the duodenum. Because of elevated hepatic enzymes and esophageal varices, the patient's treatment was based on a diagnosis of hepatic cirrhosis. Detailed echocardiographic evaluation had not been performed at that time. He was discharged with the diagnosis of hepatic cirrhosis caused by alcohol use. He had no specific medical history except for an operation for a proximal femur fracture after a traffic accident.

At admission, the patient's physical examination revealed hepatomegaly and bilateral rales at the bases of the lungs. Blood analysis revealed elevated gamma-glutamyl transpeptidase and alkaline phosphatase levels, which were 2381 U/L and 3481 U/L, respectively. The erythrocyte sedimentation rate and complete blood count were normal except for mild leukocytosis (white blood cell count, 12,700/mm^3). On chest x-ray there was cardiomegaly, and electrocardiography demonstrated right atrial enlargement and incomplete right bundle-branch block. Transthoracic echocardiography showed a large atrial mass (82 × 61 mm), which was moving toward the tricuspid orifice during diastole. Cardiac catheterization showed an arterial branch arising from the circumflex artery and extending to the tumor, and the coronary arteries were shown to be normal. Right heart catheterization was not able to be performed because of the mass, which was entirely filling the right atrium. Cardiac magnetic resonance imaging (MRI) was performed; a large solid polypoid mass in the right atrium was demonstrated (Figure 1). The patient underwent open-heart surgery. Selective superior vena cava and left femoral venous cannulation was performed. A right atriotomy was done with cardiopulmonary bypass (CPB). A large,

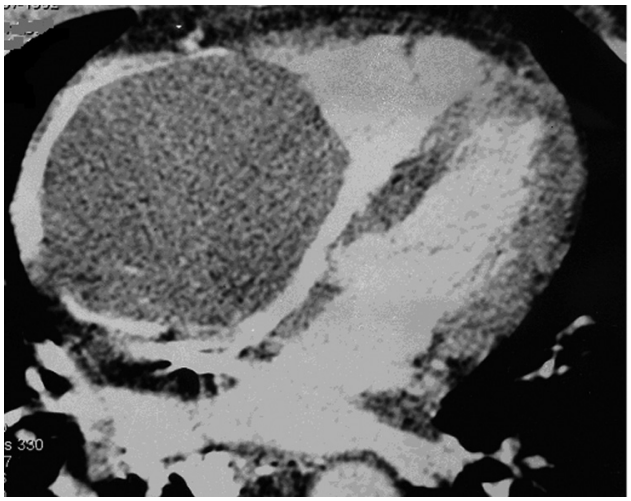

Figure 1. Magnetic resonance image of the giant myxoma, which was filling the entire right atrium.

encapsulated and soft lobulated tumor with a size of 100 × 80 mm almost totally filling the right atrium was seen (Figures 2 and 3). It had a broad-based attachment to the dorsal free wall of the right atrium and interatrial septum. With mild systemic hypothermia, the aorta was cross clamped and cold blood cardioplegia was induced. The tumor with its pedicle was completely excised from the atrial wall and the interatrial septum. The defect in the interatrial septum was repaired with polyprolene stitches. The tricuspid valve function was spared. Right atriotomy was primarily closed with continuous polyprolene stitches. Weaning from CPB was uneventful. The pathological examination revealed the tumor had the typical appearance of a myxoma, with spindle cells widely spaced by an abundant myxoid matrix, a size of 9 × 6 × 6 cm, and a pedicle with a diameter of 1.2 cm. The patient underwent gastrointestinal system evaluation with the cooperation of the gastroenterology department after surgery to clarify the hepatic cirrhosis symptoms. Hepatomegaly disappeared. The patient's hepatic enzymes returned to nearly normal ranges; gamma-glutamyl transpeptidase was 259 U/L and alkaline phosphatase was 301 U/L, and his upper endoscopic evaluation showed mild duodenitis and gastritis. There were no esophageal varices. The diagnosis of hepatic cirrhosis was wrong. The postoperative period was uneventful, and the patient was discharged on the sixth postoperative day. One month later, patient reports from another university hospital showed that there were no signs or symptoms of cirrhosis.

DISCUSSION

Primary cardiac tumors are uncommon, with an incidence rate between 0.001% and 0.28% reported in an autopsy series [Anagnostopoulos 2004]. Cardiac myxoma is the most common, constituting approximately 50% of all benign cardiac neoplasms in adults. Three fourths of these tumors are situated in the left atrium, and 18% in the right atrium [Murayama 2001; Anagnostopoulos 2004]. The diagnosis is based on histopathological and immunohistochemical findings. Although atrial myxomas are typically benign, local recurrence due to inadequate resection or malignant change has been reported [Hemachandran 2003; Yu 2006]. Sudden death may occur in patients with atrial myxoma. Death is caused by coronary or systemic embolization or by obstruction of blood flow at the mitral or tricuspid valve. Clinical manifestations may vary from no symptoms to various manifestations, including congestive heart failure, syncope, and arrhythmias with or without systemic findings such as high erythrocyte sedimentation rate, anemia, leukocytosis, elevated gamma globulin, thrombocytopenia, or low-grade fever. Left atrial myxomas produce symptoms when they reach about 70 g. Right atrial myxomas grow to approximately twice this size before becoming symptomatic. Tumors vary widely in size, ranging from 1 to 15 cm in diameter. They can mimic not only every cardiac disease but also infective, immunologic, and malignant processes. Myxomas must therefore be included in the differential diagnosis of valvular heart disease, cardiac insufficiency, cardiomegaly, bacterial endocarditis, disturbances of ventricular and supraventricular rhythm, syncope, and systemic or pulmonary embolism. The symptoms depend on the size, mobility, and location of the tumor [Reynen 1995]. Symptoms of left-sided heart failure include dyspnea on exertion (75%) that may progress to orthopnea, paroxysmal nocturnal dyspnea, and pulmonary edema. Symptoms are caused by obstruction at the mitral valve orifice. Valve damage may result in mitral regurgitation. Symptoms of right-sided heart failure include fatigue and peripheral edema. Abdominal distension due to ascites is rare; however, it is more common in slowly growing right-sided tumors. In this case, due to high levels of hepatic enzymes, esophageal varices, and hepatomegaly, the patient had been treated in another clinic for hepatic cirrhosis. The patient was admitted to our clinic with the worsening of

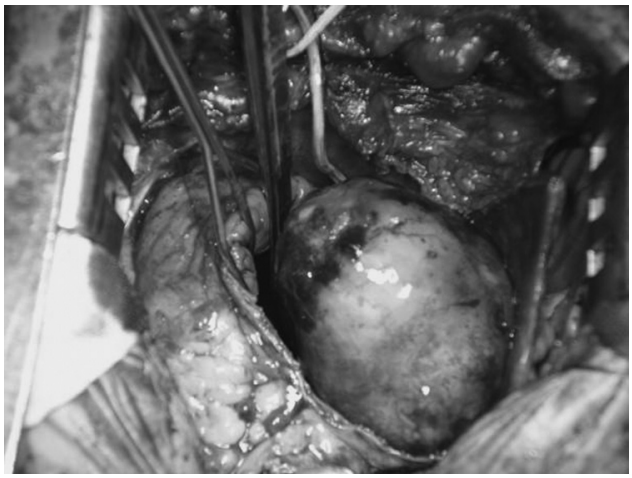

Figure 2. The giant myxoma in an intra-atrial position during the operation.

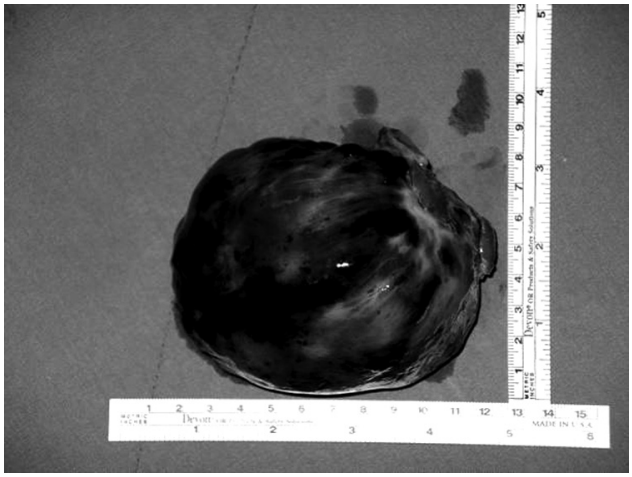

Figure 3. Diameter of the myxoma.

constitutional symptoms of palpitation, fatigue, shortness of breath and myalgia and for further evaluation. These symptoms are also observed in the later stage of progressive heart failure associated with left atrial myxomas. Constitutional symptoms that include fever, weight loss, arthralgias, and Raynaud phenomenon are observed in 50% of patients. These symptoms may be related to the overproduction of interleukin 6. The motion of the tumor can damage the atrioventricular valve and rupture the chordae [Reynen 1995].

When present, anemia, thrombocytopenia, and high erythrocyte sedimentation rate can be explained by continuous destruction of blood elements due to the "ball-valve" movement of the mass [Reynen 1995], but our patient had only mild leukocytosis in the complete blood count. Jelic et al reported a delay ranging from 10 days to 25 months from the onset of symptoms to diagnosis [Jelic 1996]. Our patient was hospitalized with the diagnosis of cirrhosis four years before the operation. Afterward, the patient was unable to be followed-up.

Surgical removal of the tumor should be performed as soon as possible; the long-term prognosis is excellent, and recurrences are rare and are prevented by resection of the tumor as well as normal tissue surrounding the base [Hanson 1985; Reynen 1996]. Recurrence is usually attributed to incomplete excision of the tumor, growth from a second focus, or intracardiac implantation from the primary tumor. As in this case, it is very rare to see cirrhotic changes that were was caused by a giant right atrial myxoma. In such a case, all cirrhosis etiologies including the rarer ones such as cardiac masses must be recognized and a detailed investigation should be done.

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