

Left Atrial Myxoma with Neovascularization Presenting as a Sick Sinus Syndrome

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ABSTRACT

Myxoma is benign tumor of the heart. It is mostly located in the left atrium and revascularized by the left and right coronary artery in 30% to 40% of cases. Symptoms of these neovascularized cardiac myxomas are typically quite variable, from obstruction of mitral valve to coronary embolism resulting in acute myocardial infarction. In this case, left atrial myxoma that is revascularized by nodal branches of the right coronary artery presented as a sick sinus syndrome, which is rare in literature.

INTRODUCTION

Intracardiac myxoma is the most frequent benign tumor of the heart. Approximately 75% to 80% of myxomas are located in the left atrium, 10% to 20% are in the right atrium, and 5% to 10% are in both atria or either ventricle. By angiography, neovascularization of cardiac myxomas can be seen in 30% to 40% of cases. These vessels arise from the left circumflex or right coronary arteries in equal distribution. The symptoms of these neovascularized cardiac myxomas are typically quite variable, ranging from obstruction of the mitral valve, coronary embolism resulting in acute myocardial infarction, cerebral emboli, and constitutional symptoms, and in many patients diagnosis is found late [Yamanaka 1990]. In this paper, a case of left atrial tumor that is revascularized by the atrial branches of the right coronary artery (RCA) is presented as a sick sinus syndrome.

Sick sinus syndrome is commonly seen in the elderly and, in most cases, does not cause any symptoms. Symptoms are often intermittent, changeable, and unpredictable. The sinus node is supplied by the RCA in about 65% of cases, by the left circumflex branch in 25% of cases, and by both in 10% of cases, although some may originate from the aorta and left main artery [Mangrum 2000]. Etiologies include intrinsic and extrinsic factors affecting the sinus node. One of the extrinsic factors affecting the sinus node is cardiac tumor. In the literature, a right atrial tumor, which is lymphoma, can cause

sick sinus syndrome by the way of interruption of sinus nodal artery or invasive effect of tumor to the sinus node, and a few previous cases of right atrial myxoma have been shown to have first atrioventricular block, but there is no information about the left-sided tumor [Roberts 1990; Motto 2008].

CASE REPORT

A 63-year-old female patient was presented to a hospital with a clinical presentation of intermittent palpitation and presyncope that had been occurring for a year. Two years ago, she had coronary arterial angioplasty with stents on the left anterior descending and circumflex arteries. Parameters other than echocardiography were normal on the last control. After tachycardia following bradycardia, the patient was admitted to the hospital, and the prediagnosis was sick sinus syndrome. Once sinus rhythm had been restored, physical examination showed normal heart sounds and blood pressure of 120/80 mmHg. All laboratory tests and her chest x-ray were normal. Level of serum creatine kinase and its MB-fraction were

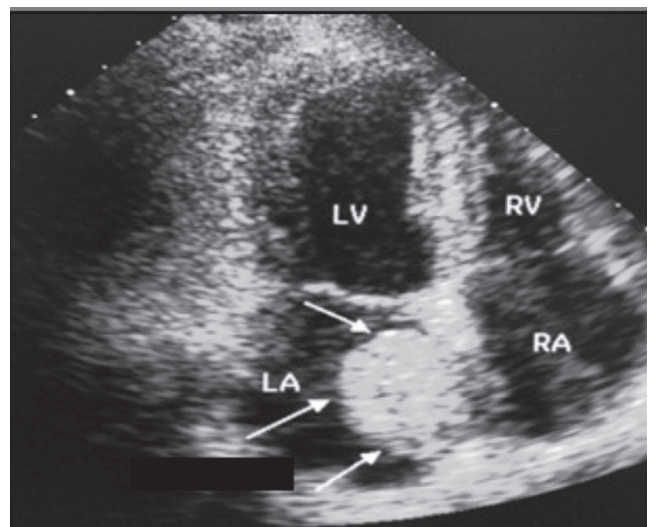


Figure 1. Preoperative transthoracic echocardiography demonstrates a round, mobile, pedunculated mass in the left atrium near the superior pulmonary veins originating from interatrial septum. LV indicates left ventricle; RV, right ventricle; LA, left atrium; RA, right atrium.

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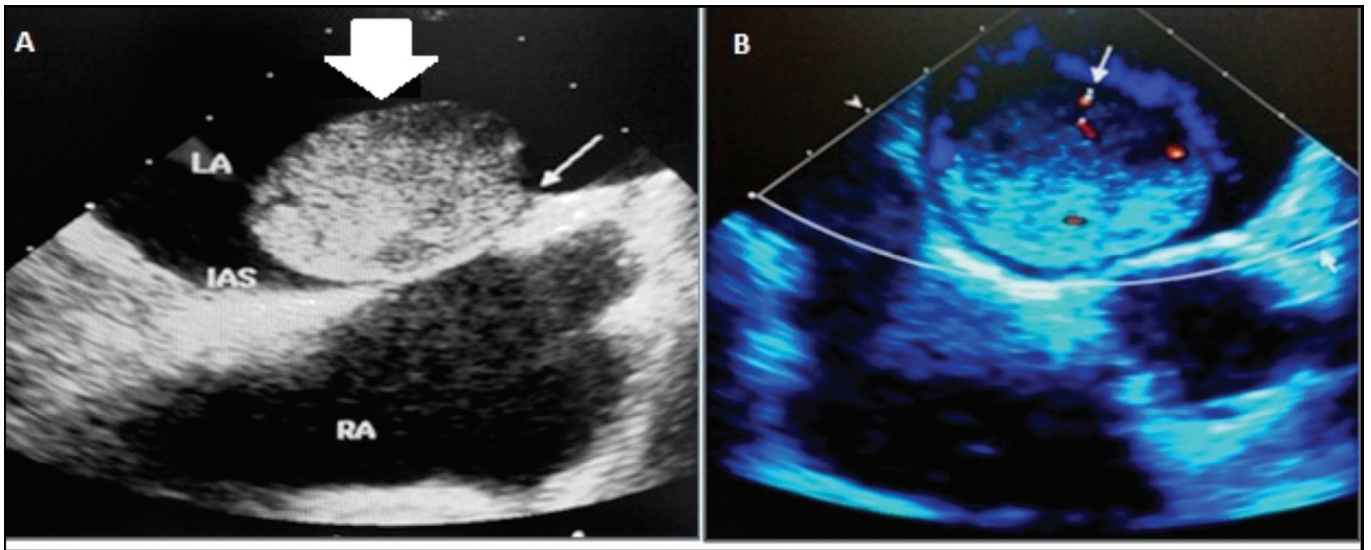


Figure 2. Preoperative transesoophageal echocardiography revealed the mass (A) and its vascularization with color doppler (B). LA indicates left atrium; RA, right atrium; IAS, interatrial septum.

normal, and troponin T was negative. Two-dimensional echocardiography demonstrated a mass in the left atrium.

After demonstration of left atrial mass, the patient was transferred to our hospital. By repetition of 2-dimensional echocardiography, a round, mobile, pedunculated mass was seen in the left atrium near the superior pulmonary veins originating from the interatrial septum that seemed to be an

atrial myxoma with dimensions of 3.9×2.9 cm (Figure 1). Other cardiac chambers and both great vessels were of normal size and clear. All cardiac valves were normal. Color doppler imaging of transesoophageal echocardiography revealed vascularized architecture of the mass (Figure 2).

Preoperative selective coronary angiography showed that coronary artery stents on the left anterior descending and

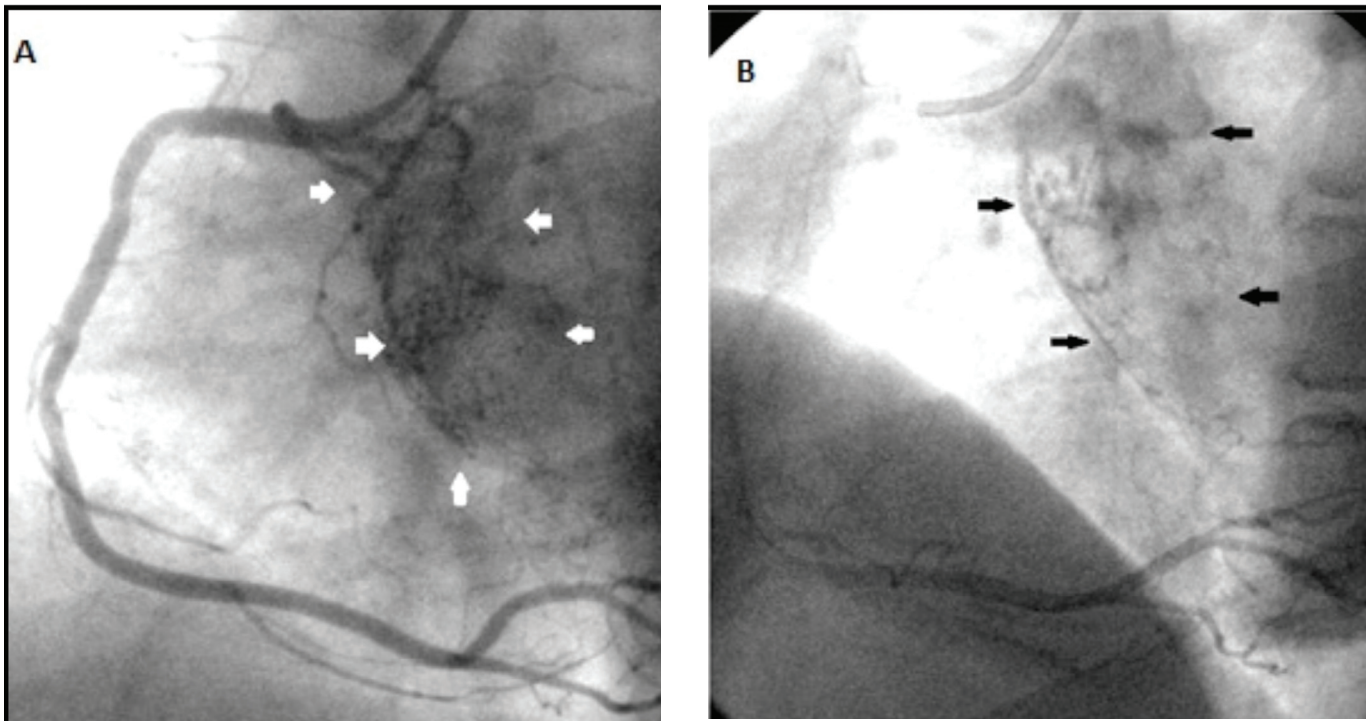


Figure 3. Coronary angiogram shows neovascularization of left atrial myxoma with arterial blood supply from the right coronary artery (A-B).

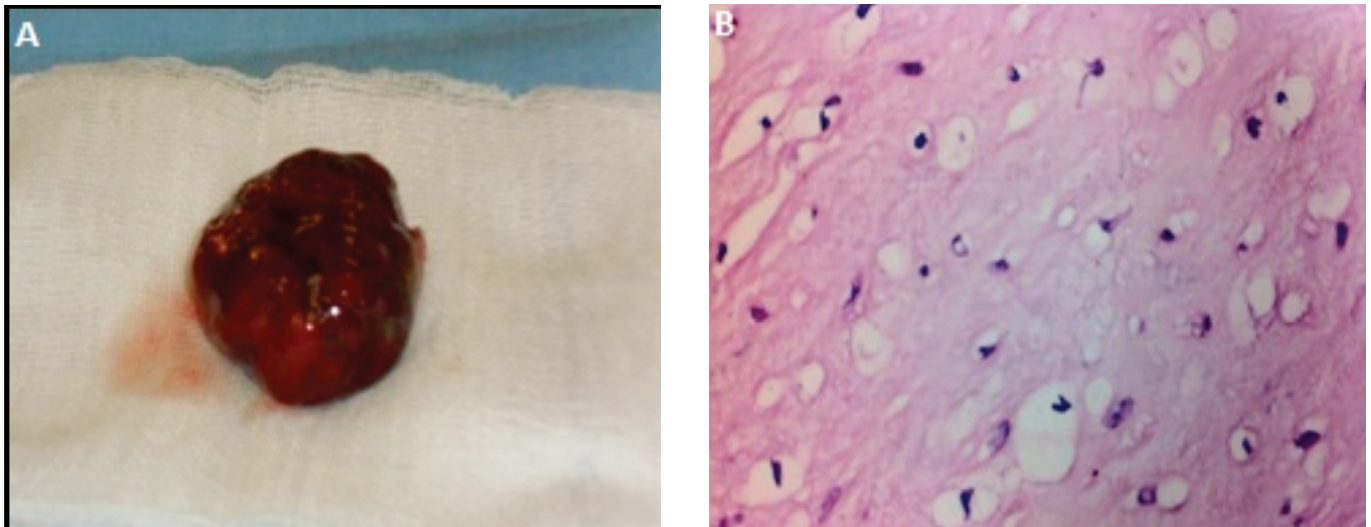


Figure 4. A, Macroscopic view of left atrial myxoma measuring 3.9 × 2.9 cm and B, its histologic appearance diagnosed as myxoma (H-E ×400).

circumflex arteries were normal with right-dominant coronary circulation, and it demonstrated marked neovascularization of the left atrial mass supplied by a cluster of new vessels that originate from the atrial branches of right coronary artery (Figure 3). This configuration of right coronary artery was not seen on coronary angiography 2 years ago.

During the preoperative period, the patient had arrhythmias intermittently. The patient underwent cardiac surgery, and the mass in the left atrium was resected via left atriotomy. Histologic diagnosis was atrial myxoma (Figure 4). Cardiac dysrhythmias and symptoms of presyncope were sustained postoperatively. In the 48-hour period of holter monitoring, sick sinus syndrome was diagnosed.

The treatment of choice for the symptomatic sick sinus syndrome was implantation of permanent pacemaker (dual-chamber pacemaker). Afterward, the course of postoperative period was uneventful.

DISCUSSION

Myxomas are more common among women and can affect the atrium, ventricles, or mitral valve, with the left atrium being most commonly involved (70% to 80%). The gelatinous, lobulated tumor arises from a short stalk in the atrial septum in 50% of patients, but myxomas may have multiple foci and can involve valvular tissue. Because the symptoms are atypical and vary greatly, the diagnosis was elusive until the advent of echocardiography [MacGowan 1993].

Transthoracic echocardiography and transeosophageal echocardiography are the noninvasive methods for diagnosis. Two-dimensional echocardiography was used for diagnosis of cardiac tumor, and transeosophageal echocardiography was used as a second step for demonstration of tumor place and to show tumor vascularization by color doppler.

Coronary angiography is usually performed to exclude coronary artery disease. By angiography, neovascularization of the tumors

can be seen in 40% of the patients. In the literature, tumor neovascularization from branches of the left circumflex or right coronary arteries was observed; both left and right atrial myxomas and a right ventricular myxoma have been demonstrated in this manner [Sharma 1993; Van Cleemput 1993]. In this patient, we performed selective coronary angiography because of the history of coronary artery disease. Coronary artery stents were normal, and the left atrial mass that is supplied by a cluster of new vessels that originate from the atrial branches of the right coronary artery was seen.

Presentations of neovascularized myxomas are quite variable. In the literature, the left atrial myxoma supplied by the circumflex coronary artery arising from the right sinus of valsalva presented with acute inferior myocardial infarction secondary to embolism [Oğuzhan 2008]. Left atrial myxoma supplied by the left and right coronary arteries presented with paroxysmal dyspnea due to obstruction of the left ventricular inflow tract [Bauer 2002]. Nonspecific constitutional symptoms were dominant in a case of left atrial myxoma supplied by the right coronary artery [Tay 2002]. Arrhythmias are not a common symptom related to left atrial myxoma.

Sinus node dysfunction, also known as sick sinus syndrome, is a common disease. Most causes of sick sinus syndrome are idiopathic, and the cause can be multifactorial. The prevalence of coronary artery disease in chronic conduction disorders has been reported to be 30% to 70% [Hsueh 2001]. Coronary artery disease may coexist with sick sinus syndrome in a significant number of patients, although it is not considered a major cause of the syndrome. Nodal dysfunction was noted only to be transient and recovered after ischemia was removed. Degenerative fibrosis of nodal tissue is actually considered the most common cause of bradyarrhythmias.

Normal sinus node generator function in the presence of arteriosclerotic involvement of the sinus node artery could be explained by the extensive collateral supply to the sinus node. Kyriakidis et al also found that even though the sinus node arteries were atheromatous, and no sinus node dysfunction

was noted [Kyriakidis 1988]. In a study by Shaw et al, chronic sinoatrial disorder was present even when blood supply to the sinus node was unobstructed [Shaw 1987]. Uncommonly, chronic ischemia may cause fibrosis and lead to symptoms of sick sinus syndrome for months to years after myocardial infarction. In this patient, coronary artery disease on the left anterior descending and circumflex arteries was present without history of myocardial infarction, and the right coronary artery was normal on coronary angiography.

The pathophysiological basis is unclear, but in this left atrial tumor sick sinus syndrome probably occurred transiently by embolism of sinus nodal artery or a steal phenomenon through the atrial collateral vessels or just by coincidence. Such theories should be supported by surgical removal of the cardiac tumor. In this case, dysrhythmia was sustained, probably because of conduction disturbances following cardiac operation for removal of the left atrial myxoma.

Treatment should be aimed at controlling complications and relieving symptoms. Cardiac pacemaker (dual-chamber) implantation is the most powerful therapy for sick sinus syndrome. In our patient, a pacemaker was implanted, and she was discharged smoothly.

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