Case Reports

Catastrophic Systemic Embolization from a Left Atrial Myxoma

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We report the fatal course of a left atrial myxoma: its systemic embolization to the coronary, cerebral, renal, and peripheral vascular beds in a 39-year-old woman resulted in rapid clinical deterioration, multiorgan failure, and death. Among reported cases of left atrial myxoma, this degree of embolic burden is exceedingly rare. In addition to reporting the patient's case, we discuss the presentation and diagnosis of possible intracardiac sources of systemic emboli. ***(Tex Heart Inst J 2014;41(1):64-6)***

When a patient presents with simultaneous vascular insults that involve multiple organ systems, a catastrophic clinical outcome can result. We report the case of a patient who had symptoms of systemic embolization, and we discuss the presentation, recognition, and treatment of the left atrial myxoma that was responsible.

Case Report

A 39-year-old black woman was found unresponsive and in respiratory distress outside her home. According to her family, she had felt fatigued over the past 2 months and had occasionally reported fevers and night sweats. She had been taking oral contraceptive pills and smoked half a pack of cigarettes per day. Her personal and family medical histories yielded nothing else of note.

Upon evaluation by emergency medical personnel, the patient was unconscious, tachypneic, and displaying possible seizure activity. In the emergency department, the patient was comatose with a Glasgow coma scale of 5, a temperature of 37 °C, a blood pressure of 117/77 mmHg, a heart rate of 133 beats/min, a respiratory rate of 30 breaths/min, and an oxygen saturation of 76% on room air. She was immediately intubated. A 12-lead electrocardiogram revealed 2- to 4-mm ST-segment elevation in the lateral leads, consistent with acute ST-elevation myocardial infarction (STEMI). Pertinent laboratory values included a cardiac troponin I level of 1.6 ng/mL, a white blood cell count of 13,800/μL, and an international normalized ratio of 1.21. The patient was anuric. A chest radiograph showed a normal cardiac silhouette with bilateral pulmonary edema. Aspirin (325 mg) was administered through a nasogastric tube, and an intravenous heparin drip was initiated. The diagnosis was cardiac arrest secondary to STEMI. A noncontrast computed tomographic scan of the patient's head was performed, and she was sent for emergent cardiac catheterization.

Femoral arterial access was difficult to achieve in either groin because of the aspiration of thrombotic material. A 4F sheath was finally placed in the right femoral artery; the access sheath thrombosed twice during the procedure, necessitating the aspiration of large amounts of dark-red thrombus. Coronary angiograms obtained with use of 4F catheters showed abrupt thrombotic occlusion of the proximal-to-mid left anterior descending coronary artery, first diagonal branch, and proximal-to-mid left circumflex coronary artery (Fig. 1). The right coronary artery was angiographically normal. The occlusions were not suitable for catheter aspiration, balloon angioplasty, or stenting. A left ventriculogram showed a left ventricular ejection fraction (LVEF) of 0.30 and akinesis of the anterolateral, inferolateral, and apical walls. The computed tomographic report, received during the cardiac catheterization procedure, noted a large, acute embolic infarct in the region of the left middle cerebral artery. In view of the substantial thrombotic burden in multiple vascular beds, we suspected either a cardiac source of emboli or catastrophic systemic antiphospholipid antibody

Key words: *Cerebral arteries/pathology; embolism/diagnosis; fatal outcome; heart atria/pathology; heart neoplasms/complications; myocardial infarction/etiology; myxoma/complications/diagnosis/epidemiology/pathology*

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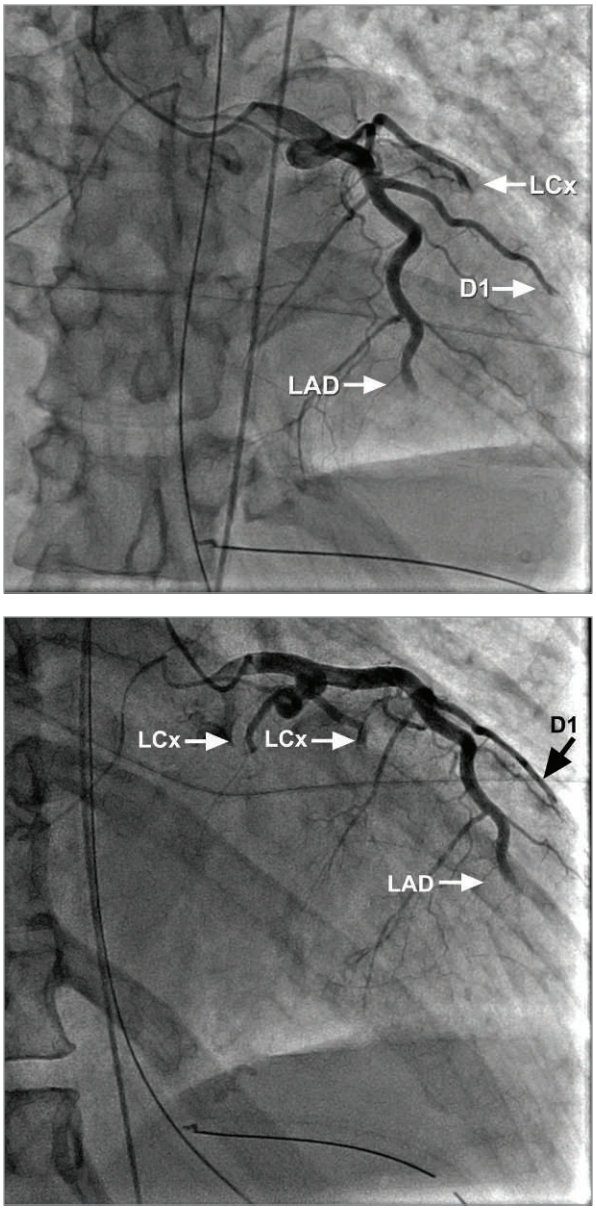

Fig. 1 *Coronary angiograms in **A**) left and **B**) right anterior oblique cranial projections show abrupt thrombotic occlusions (arrows) in the left anterior descending coronary artery (LAD), first diagonal branch of the LAD (D1), and left circumflex coronary artery (LCx).*

syndrome. While in the catheterization laboratory, the patient became hypotensive and progressed to fulminant cardiogenic shock. We decided to administer intravenous thrombolytic therapy to treat the coronary thrombi despite the increased risk of cerebral hemorrhage, given that her last known normal mental status was 9 hours earlier.

Vasopressor and inotropic support was initiated, and the patient was transferred to the cardiac critical care unit. A transthoracic echocardiogram, obtained immediately after left-sided heart catheterization, revealed an LVEF of 0.15, akinesis of the anterior and lateral left ventricular walls, and a 1.8 × 2.4-cm left atrial mass arising from the interatrial septum (Fig. 2A). An extensive hypercoagulability evaluation revealed nothing unusual. During the next 48 hours, the patient deteriorated clinically, from multiorgan failure and signs of severe anoxic brain injury to death.

Autopsy results revealed a 2 × 2.5-cm pedunculated cardiac mass arising from a short stalk attached to the left atrial side of the interatrial septum, 2 cm superior to the mitral annulus at the level of the fossa ovalis (Fig. 2B). Microscopic evaluation of the mass showed thin-walled vasculature dispersed within a mucinous, myxoid background with perivascular eosinophilic cells, consistent with myxoma cells. Calretinin staining of the mass was strongly positive, confirming the presence of cardiac myxoma cells. Arterial occlusions were identified within each common iliac artery, the renal arteries, the left middle cerebral artery, and the epicardial branches of the left main coronary artery. Each embolus had a homogeneous myxoid background, similar in appearance to the left atrial mass. Calretinin staining of

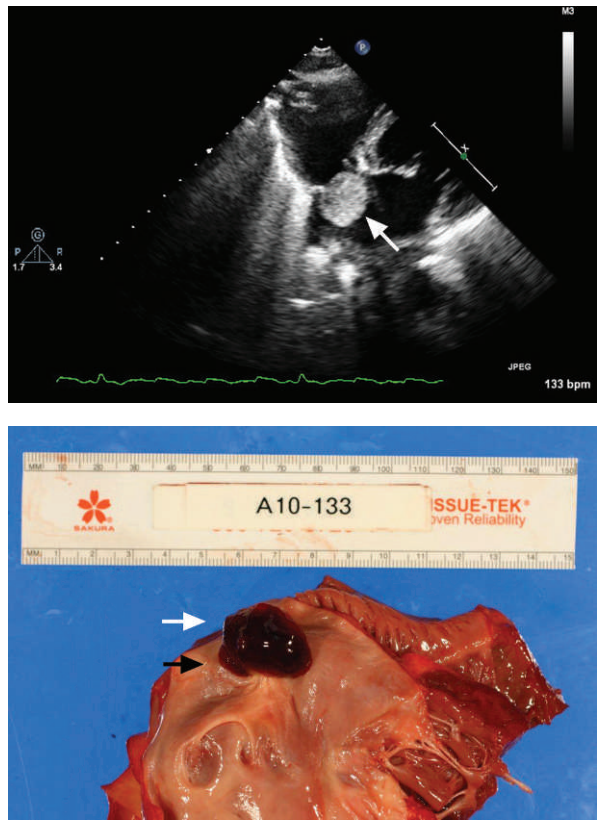

Fig. 2 ***A**) Transthoracic echocardiogram (subcostal view) shows a left atrial mass (arrow) arising from the interatrial septum, suspicious for myxoma. **B**) Photograph from autopsy shows a left atrial myxoma with a polyploid gelatinous appearance (white arrow), and a broad-based stalk attached to the interatrial septum (black arrow).*

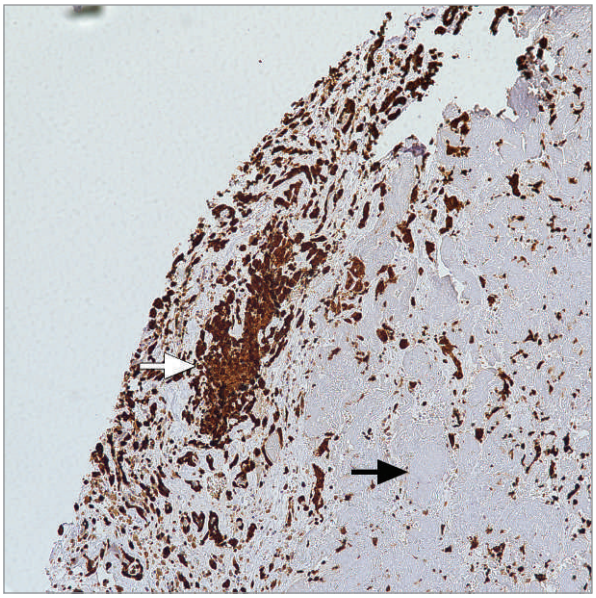

Fig. 3 *Photomicrograph of left middle cerebral artery embolus shows strong, diffusely positive calretinin staining of myxoma cells (brown color). White arrow points to a large nest of myxoma cells, staining positive. Pale white tissue is the myxoid matrix (black arrow), staining negative. The entire image represents the myxoma tumor (orig. ×10).*

the left middle cerebral artery occlusion confirmed the presence of myxoma cells (Fig. 3).

Discussion

As an embolic complication of left atrial myxoma, acute myocardial infarction is rare: in one large case series,[1] it occurred in only 2% to 3% of patients. In 50% of patients with left atrial myxoma, mitral valve obstruction is the typical presenting symptom. Systemic embolic events occur in 30% to 40% of patients, and constitutional symptoms (fever, fatigue, myalgias, and weight loss) 20% of the time. Up to 20% of patients are asymptomatic.[2,3] Sudden cardiac death, an infrequent consequence of left atrial myxoma,[4] is thought to result from coronary embolization or severe mitral valve obstruction (ball-valve syndrome). The infrequency with which coronary embolization occurs can be explained by the nearly perpendicular relationship of the coronary ostia and the aortic root, filling of the coronary arteries during ventricular diastole, and shielding of the coronary ostia by the aortic valve leaflets during ventricular systole.[5,6] As reported by her family, our patient had experienced constitutional symptoms (fever, chills, and mild weight loss), probably related to the myxoma, in the months before presentation. These systemic symptoms are mediated through increased interleukin-6 release from myxoma cells.[7,8] There was no reported syncope or presyncope to suggest obstruction of the mitral valve.

The large embolic burden in this case is perplexing. One possibility is an unidentified hypercoagulable disorder: the patient was an active smoker and was taking oral contraceptive pills, which would increase the risk of thrombosis and probably contribute to the thrombotic burden associated with the myxoma. The patient's myxoma was polyploid, which also increased the embolic risk. Her rapid clinical deterioration was unfortunate but unavoidable. Given the large systemic burden of emboli and the few therapeutic options, intravenous thrombolytic therapy was the best remaining treatment; however, it was unsuccessful. Successful intracoronary thrombolysis, catheter aspiration of thrombus, or both have sometimes been successful in cases of embolization to the coronary arteries.[9] Catheter-directed thrombolysis of cerebral emboli from myxomas has also been described, as has successful surgical thrombectomy of peripheral emboli.[10-13]

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