Multiregional Embolizations and Takotsubo Cardiomyopathy Associated with Left Atrial Myxoma

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Cardiac myxoma represents the most common primary cardiac neoplasm, accounting for nearly 75% of benign cardiac tumor. Tumor embolisms occur in 30% to 40% of patients with myxoma, and half of these are cerebral arteries. To our knowledge, this is the first published report of multiregional tumor embolizations except for cerebral artery and Takotsubo cardiomyopathy in patients with left atrial myxoma.

Keywords: cardiac myxoma, tumor embolism, Takotsubo cardiomyopathy

Introduction

Cardiac myxoma is a rare disease with a variety of nonspecific clinical manifestations depending on its size, location and mobility. The clinical course of the left atrial myxoma is characterized by classic triad of obstructive, embolic, or constitutional effects of the tumor.1–3)

Tumor located in the left side of the heart may embolize to various systemic organs, leading to cerebral artery, upper and lower extremity artery, splenic and renal arteries.4)

Takotsubo cardiomyopathy is a type of non-ischemic cardiomyopathy, in which there is a sudden, temporary weakening of the myocardium. It can be provoked by a stressful or emotional situation or exposure to high doses of catecholamines.5,6)

We report the first case of multiregional tumor embolizations except for cerebral artery and Takotsubo cardiomyopathy in patients with left atrial myxoma.

Case

A 53-year-old woman was brought into our hospital’s emergency department by ambulance and had complained of sudden onset resting pain, pallor, parenthesis, and paralysis in right leg during 4 hours. She had also complained of dyspnea and palpitation. She had no medical history such as hypertension and diabetes mellitus.

She had complained hot flushing, palpitation, fatigue, depressive mood, sleeping trouble, and myalgia since about one year. She was diagnosed with perimenopausal syndrome, so she had had hormone replacement treatment for two months.

Her blood pressure was 120/80 mmHg, and her pulse rate was 120 beats per minute. She was afebrile. Laboratory findings showed elevated cardiac enzyme (troponin T 2.24 ng/mL, creatine kinase-MB 154 ng/mL, pro-BNP 4687 pg/mL), leukocytosis (WBC count 16400 /microL), creatine phosphokinase / lactic dehydrogenase 203 / 492 U/L and normal renal function (blood urea nitrogen / creatinine 15.5 / 0.9 mg/dL). Her chest radiography demonstrates pulmonary edema.
Her electrocardiography shows tachycardia and ST-segment elevation in the precordial leads (V3 through V6, Fig. 1). Echocardiographic findings were decreased left ventricular (LV) systolic function (ejection fraction = 35%), mild pulmonary hypertension (tricuspid regurgitant maximal velocity = 3.2 m/sec), akinesia and ballooning of apical LV wall, and large left atrial myxoma (Fig. 2A–2D). Computed tomography (CT) of the chest and abdomen showed splenic and both renal infarctions. Computed tomography angiography (CTA) of lower extremities showed acute occlusions of lower extremity arteries (Fig. 3).

Emergent vascular surgery was performed because she complained progressive pain, weakness, and foot drop on right leg. Embolectomy was performed with the Fogarty catheter through the right common femoral artery under local anesthesia, and gelatinous materials were removed (Fig. 4B). Her pain was relieved after surgery. After vascular surgery, massive anti-platelets (aspirin 300 mg and clopidogrel 75 mg daily), anticoagulant (heparin) and thrombolytic (urokinase, 1000000 units) were administered.

On day 2 after admission, follow up laboratory findings showed decreased renal function (blood urea nitrogen / creatinine = 28.1 / 1.8 mg/dL) and marked elevation of muscle enzyme (creatine phosphokinase / lactic dehydrogenase = 32258 / 5388 U/L).

On day 7 after admission, follow-up echocardiography showed findings similar to those of the previous examination. Coronary angiographic findings were normal. Normalization or decreasing trends of laboratory findings are shown (troponin T 0.184 ng/mL, creatine kinase-MB 4.94 ng/mL, blood urea nitrogen / creatinine = 23.3 / 0.9 mg/dL, creatine phosphokinase / lactic dehydrogenase = 10430 / 4179 U/L, WBC 10,200 /microL). On day 10 after admission, cardiac surgery was performed, in which a 3 × 5 cm, polypoid left atrial myxoma was removed (Fig. 4A). On day 3 after cardiac surgery, follow-up echocardiography showed that the LV ejection fraction had increased to 59% and previously akinetic regions of myocardium had improved (Fig. 2E and 2F). On day 6 after cardiac surgery, CTA of lower extremities showed that occluded lesions of lower extremity arteries had been resolved.

On day 7 after cardiac surgery, the patient was discharged from the hospital. She had no complaints of dyspnea, palpitation, or pain in the lower extremities.
Fig. 2  Echocardiography. A: Magnification of Left atrium shows a nodular and mobile 3.25 × 3.15 cm sized mass. B: Apical 4-chamber views at the patient’s initial evaluation show apical ballooning in end-diastolic. C: in end-systolic. D: Trans-esophageal echocardiography view in 71 degree shows a huge polypoid mass in the left atrium. E: Apical 4-chamber views at hospital discharge show improved apical wall motion in end-diastolic. F: in end-systolic.

Fig. 3  Computed tomography angiography (CT). Initial CTA shows acute occlusion in lower extremity artery. Arrow indicates acute occlusion in the right superficial femoral artery.
Discussion

Cardiac myxomas, although rare, are the most common, primary benign tumors of the heart. They occur more frequently in women and most commonly between the ages of 30 and 60 years. About 75% of cardiac myxomas present in the left atrium. The rest of the myxomas are right atrium (20%), right ventricle (3%), and left ventricle (3%).

Clinical manifestations depend on location, size, and mobility of tumor. Obstructive symptoms may be due to obstruction of the ventricle. It results in dyspnea, pulmonary edema, syncope, and even sudden cardiac death. Embolization occurs in 30% to 40% of cases due to tumor fragments or thrombi from the surface of the myxoma, with the brain being the most common destination. Involvement of extremity, visceral, coronary, splenic, and renal arteries are rare. Constitutional symptoms such as fever, skin rash, joint pain, and weight loss, as a result of the production of cytokines and growth factors by the myxoma.

Takotsubo cardiomyopathy (TCM) is a sudden temporal cardiac syndrome that involves left ventricular apical akinesia and mimics acute coronary syndrome. Although the exact etiology is still unknown, the syndrome appears to be triggered by an emotional or physical stressor. Complications such as heart failure, cardiogenic shock, mitral regurgitation, ventricular arrhythmia, LV mural thrombus formation, LV free-wall rupture, and even sudden cardiac death occur in 20% of TCM cases, but nearly 95% of patients experience complete recovery within 4–8 weeks.

In our case, multiregional embolizations, such as those of both lower extremities, splenic, and both renal arteries, and Takotsubo cardiomyopathy simultaneously occur in patients with LA myxoma. The middle-aged woman in our case had constitutional symptoms mimicking perimenopausal syndrome, so she had had hormone replacement therapy (HRT). HRT may accelerate the thrombus formation on the surface of the myxoma.

Lower extremity embolizations result in acute limb ischemia, which presents as progressive pain, motor weakness, and foot drop. Physical and emotional stress from acute limb ischemia may cause TCM, which represents dyspnea and pulmonary edema.

Treatment of embolizations from cardiac myxoma may be organ specific. In general, resection of cardiac myxoma is the only efficient treatment to ensure recovery of the patients. Conservative therapy, such as anti-platelet and anticoagulant therapy, and surgical embolectomy may be treatment options, depending on the severity of organ damage. Thrombolytic treatment is not usually recommended because of the risk of embolism and hemorrhage.

Cardiac myxomas occur more frequently in middle-aged women, and its constitutional symptoms may be confused with perimenopausal symptoms. Cardiac myxoma should be suspected before HRT is started. Cardiac tumors are an unusual cause of acute limb ischemia, but embolic myxoma should be suspected in younger, previously healthy patients presenting with acute arterial ischemia. Treatment of arterial embolization from cardiac
myxoma does not be understood. Although cardiac myxomas have benign nature, emboli from cardiac myxomas have the ability to implant, grow, and lead to life-threatening complications. Therefore, closed observation of organ function may be needed during follow up periods. The decision to resect the tumor embolus must be weighed against existing risks.

Conclusion

We reported Takotsubo cardiomyopathy associated with acute, lower extremity ischemia due to embolic events in patients with left atrial myxoma, which we treated by surgical embolectomy and open heart surgery.

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Disclosure Statement

The authors declare no conflicts of interest associated with this study.

References