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CASE REPORT
Other

A case report of atrial myxoma presenting with systemic embolization and myocardial infarction

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Background Cardiac myxomas are the most common benign primary tumour of the heart. Clinical presentation is variable and ranges from constitutional symptoms to clinical features due to intracardiac obstruction, such as mitral stenosis, coronary embolization, or systemic embolization. Surgical resection is the only effective treatment to prevent its debilitating and catastrophic complication.

Case summary A 61-year-old woman presented with an-hour history of bilateral leg pain, numbness, lightheadedness, dyspnoea, and diaphoresis. Physical exam was remarkable for pale and cold lower extremities. Arterial pulse was not palpable in the right femoral, popliteal, and posterior tibial and dorsalis pedis arteries bilaterally. Electrocardiogram demonstrated normal sinus rhythm with T-wave inversion in lead I, V2, V3, and V4. Laboratory investigations were remarkable for leucocytosis and elevated troponin. Computed tomography angiogram showed emboli with acute infarcts involving the spleen and kidneys, acute embolic occlusion of right external and internal iliac arteries, and left distal common femoral artery. She underwent emergent bilateral cut-down and femoral artery thrombectomies. Transthoracic echocardiogram demonstrated wall motion abnormalities. Computed tomography angiography of the chest revealed an atrial mass and transoesophageal echocardiography was obtained which confirmed an atrial myxoma. Coronary angiography demonstrated no significant coronary artery disease, raising the possibility of myxoma embolization to the coronary arteries as the cause of her troponin elevation and wall motion abnormality. Subsequently she underwent successful resection of the atrial myxoma.

Discussion The majority of cardiac myxomas are sporadic and arise from the left atrium as an isolated lesion in middle-aged women. Echocardiography is the diagnostic procedure of choice. The long-term survival after surgical resection is excellent and recurrence is rare.

Keywords Atrial myxoma • Myocardial infarction • Systemic embolization • Echocardiography • Case report

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Learning points

- Atrial myxoma is the most common primary cardiac tumour and can present with vague constitutional symptoms, or symptoms due to systemic embolization or intra-cardiac obstruction.
- Coronary embolism due to atrial myxoma is a very rare but a well-known cause of acute myocardial infarction, and coronary angiography can be normal.
- Echocardiography is the method of choice to make diagnosis and prompt surgical resection should be done to prevent devastating embolic complications or sudden death.

Introduction

Myxomas are the most common benign primary tumour of the heart. It is commonly located in the left atrium and mainly originates from an area in the atrial septum near the fossa ovalis. Patients with atrial myxoma present with clinical features due to intracardiac obstruction, such as mitral stenosis, coronary embolization, systemic embolization, or constitutional symptoms. Echocardiography is the method of choice for the diagnosis of myxoma and surgical resection is the only effective treatment to prevent its debilitating and catastrophic complications. Herein, we report a rare case of a large atrial myxoma presenting with extensive systemic embolization and coronary embolization causing myocardial infarction (MI).

Timeline

Case presentation

A 61-year-old white woman with a history of hypertension and hyperlipidaemia presented with 1-h duration of sudden onset pain and numbness in both legs, associated with lightheadedness, dyspnoea, and diaphoresis. Upon evaluation patient was afebrile with a temperature of 98.6°F, blood pressure of 160/110 mmHg, regular heart rate of 70 b.p.m., respiratory rate of 24, and oxygen saturation of 95% on ambient air. On physical examination, she was in acute distress due to pain in both legs. There was no jugular venous distention. Lungs were clear to auscultation and cardiac examination revealed a normal rate, regular rhythm, and normal heart sounds without a murmur. Abdomen and neurological examination were unremarkable. Her lower extremities were mottled, pale and cold to touch from mid-thigh down. Right femoral, bilateral popliteal, dorsalis pedis, and posterior tibial pulses could not be palpated. Electrocardiogram (EKG) showed normal sinus rhythm with T-wave inversion in Lead I, V2, V3, and V4 (*Figure 1*). Laboratory investigations were significant for leucocytosis of 20.7 × 103/μL and peak troponin of 2.08 ng/mL (<0.03 ng/mL). Basic metabolic panel and liver function test were normal.

Doppler ultrasonography of the lower extremity arteries was done immediately, which demonstrated thrombosis of the right iliac and bilateral common femoral arteries. To further characterize the extent of thrombosis, computed tomography angiography of abdomen and pelvis was obtained which showed emboli with acute infarcts involving the spleen and kidneys, acute embolic occlusion of right external and internal iliac arteries, and the left distal common femoral artery (*Figures 2 and 3*). Computed tomography angiography of the chest did not show pulmonary emboli but revealed a 3.4-cm solid ovoid filling defect in the left atrium concerning for thrombus vs. myxoma (*Figure 4*). The patient was taken to the operating room for emergent bilateral femoral cut-down and thrombectomy.

Time	Events
Day 1	A 61-year-old woman presented with sudden onset lower leg pain and numbness and was diagnosed with extensive systemic thromboembolism including acute bilateral lower leg arterial thrombosis. The patient underwent emergent bilateral femoral cut-down and thrombectomy. She was also diagnosed with non-ST-elevation myocardial infarction and was managed with heparin infusion.
Day 2	Transthoracic echocardiogram was obtained which showed low normal left ventricular ejection fraction (LVEF) estimated at 50–55%. The apex was dyskinetic and the mid to apical septum and apical inferior segments were akinetic. It also revealed a large echogenic mass attached to the interatrial septum which was thought to represent a myxoma vs. clot in transit. Heparin infusion was continued, and patient was monitored closely.
Day 3	Transoesophageal echocardiogram was done, which showed normal ejection fraction and wall motion abnormalities were no longer seen. A large protruding irregular mass present attached to the interatrial septum. Location of the mass on the interatrial septum further raised the suspicion for a large myxoma. Heparin infusion was continued and patient was monitored closely.
Day 4	Coronary angiography was performed which showed normal coronaries.
Day 5	Patient underwent resection of left atrial myxoma with autologous pericardial patch repair atrial septal defect.
Day 10	Patient was discharged from the hospital.
1 year follow-up	Transthoracic echocardiogram: normal LVEF and no evidence of atrial myxoma recurrence.

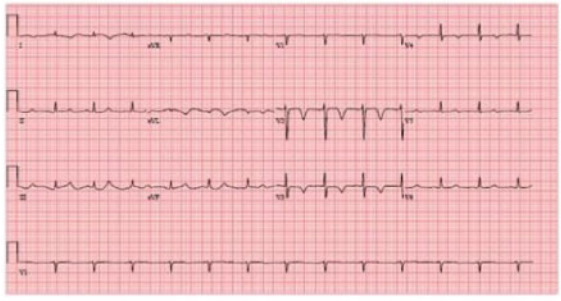

Figure 1 Electrocardiogram shows normal sinus rhythm with T-wave inversion in Lead I, V2, V3, and V4.

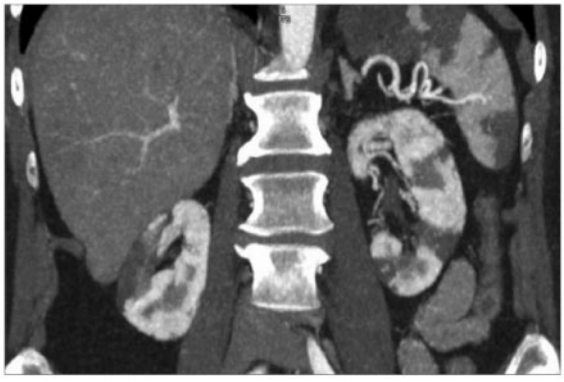

Figure 2 Computed tomography scan coronal section showing infarction of spleen and both kidneys.

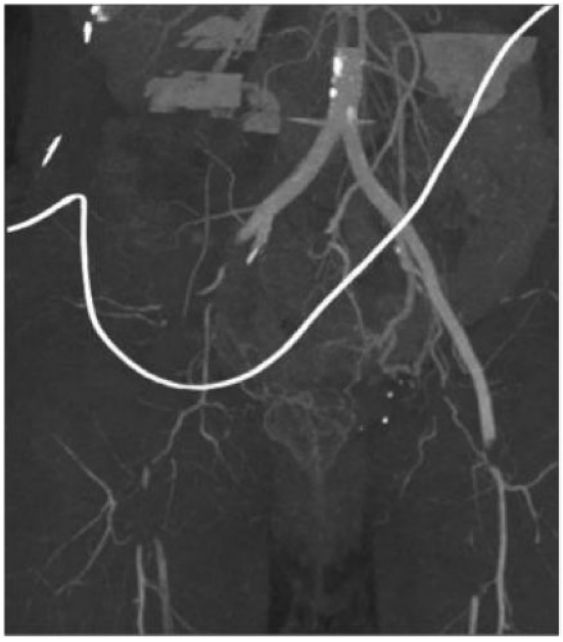

Figure 3 Computed tomography angiography coronal section showing embolic occlusion of external and internal iliac arteries on the right, and distal common femoral artery on the left just above the femoral bifurcation.

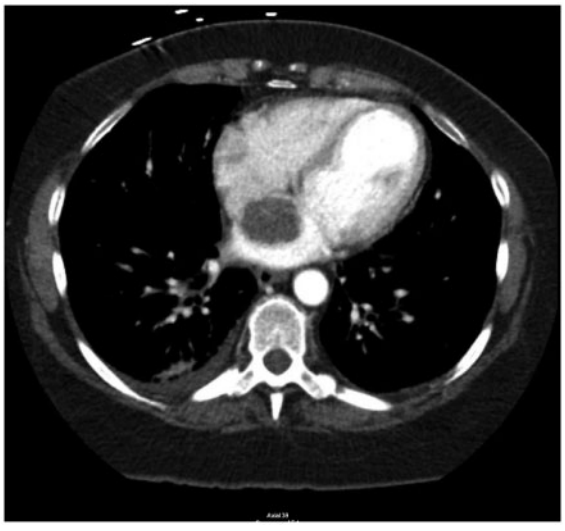

Figure 4 Computed tomography angiography of chest revealing a 3.4-cm solid ovoid filling defect in the left atrium.

Transthoracic echocardiography showed an ejection fraction of 50–55% with apical and septal wall motion abnormalities and a large echogenic mass attached to the interatrial septum. The patient was immediately started on heparin infusion. A day after, transoesophageal echocardiography (TOE) suggested the mass to be consistent with myxoma (*Figure 5*) and there was resolution of the previously seen wall motion abnormalities. Coronary angiography revealed normal coronaries and the patient underwent successful resection of left atrial myxoma with autologous pericardial patch repair of the atrial septal defect. Pathology report subsequently confirmed the diagnosis of myxoma. She recovered well from surgery and follow-up transthoracic echocardiography 1 year later did not show recurrence of myxoma.

Discussion

Primary tumours of the heart are rare with approximate prevalence of 0.02 percent in autopsy series.[1] Seventy-five percent of cardiac tumours are benign, with myxoma accounting for 50% and rhabdomyoma comprising 20% of lesions.[2] The majority of cardiac myxomas are sporadic and mostly occur as an isolated lesion in middle-aged women.[3]

Approximately 75% of cardiac myxomas arise from the left atrium, 20% of them from the right atrium, and 5% in both atria and the ventricle.[4] Cardiac myxomas produce symptoms by intracardiac

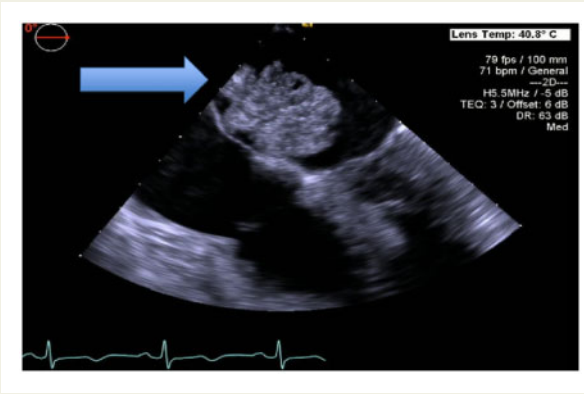

Figure 5 Transoesophageal echocardiography showing left atrial mass consistent with myxoma.

obstruction, such as mitral stenosis, coronary embolization, systemic embolization, or systemic constitutional manifestations. Systemic emboli occur in 30–40% of left atrial myxoma patients. In the majority of cases, the cerebral arteries, including the retinal arteries, are affected.

Embolization to other arterial systems including upper and lower extremities, kidney, spleen, and liver can also occur. Complete obstruction of the abdominal aorta and renal arteries by a large tumour embolus originating from the left ventricle and aortic saddle embolism have even been reported.[5] Therefore, cardiac myxomas should always be in differential diagnosis as a source of embolism in otherwise healthy patients presenting with systemic thromboembolism. The tumour size, location, and macroscopic appearance, along with mean platelet volume and platelet count, are closely associated with embolic events in patients with cardiac myxoma.[6]

The incidence of coronary artery embolization from atrial myxoma resulting in MI is exceedingly rare (0.06%).[7] One possible explanation for this low occurrence is that the coronary apertures form a right-angled junction within the aortic root, which allows a measure of protection of the coronaries by the aortic valve cusps.[7] A study by Braun *et al.*,[8] 40 cases of myxoma-related MI were reviewed from 1970 to 2002, and it was noted that the right coronary artery is most commonly involved and up to one-third of documented coronary angiogram was normal. In another study of 17 cases of MI due to atrial myxoma, 10 out of 17 (59%), patients had a normal coronary angiogram.[9] Our patient's elevated troponin and wall motion abnormalities on transthoracic echocardiogram and then the resolution of abnormal wall motion on TOE within 24 h is consistent with non-ST-elevation MI due to embolization of coronary artery and subsequent spontaneous recanalization prior to coronary angiography. The reason behind having a normal coronary angiogram in patients with atrial myxoma and acute MI is still not clearly known. A suggested hypothesis is the high rate of spontaneous recanalization after the myxomatous embolization from myxoma.[9–12]

Myocardial infarction should be considered as an embolic complication in patients with atrial myxoma who present with chest pain, ischaemic EKG changes, or elevated troponin. Echocardiography is the method of choice for diagnosis of cardiac myxoma. Transthoracic echocardiography is ~93% sensitive for the detection of cardiac myxoma, and TOE increases the sensitivity of myxoma detection up to 97%, which will be indicated especially in cases of small myxomas, right atrial myxomas, or when a poor transthoracic window exists.[13,14] While echocardiography is the traditional and easily available tool to use as a first diagnostic modality, it can be challenging to differentiate between atrial myxoma and thrombi. Cardiac computed tomography is a helpful imaging modality for differentiation between left atrial myxomas and thrombi by assessing the size, origin, shape, mobility, and prolapse of the mass.[15] After the diagnosis of atrial myxoma has been established, surgery should be performed promptly, because of the possibility of embolic complications or sudden death. Surgical resection of the myxoma is usually safe with low morbidity and mortality. Pericardial or Dacron patch can be used to close the surgical defect caused by excision of the tumour. During the surgery, to reduce the risk of tumour fragmentation and embolization, vigorous palpation or manipulation should be performed only after cardioplegia.[6,16] The long-term survival after myxoma resection is excellent and recurrence is rare. The overall risk of recurrence is about 12% and 22% for familial and complex myxoma, respectively, whereas it is only 1–3% for sporadic tumours. Regular follow-up examinations with echocardiography are indicated in all cases.[6,17,18]

Conclusion

Cardiac myxomas should always be in differential diagnosis as a source of embolism in otherwise healthy patients presenting with systemic thromboembolism. Early diagnosis and prompt surgical resection is essential to prevent further embolization.

Lead author biography

Ahmad Nawid Latifi completed MBBS course in 2013 from All India Institute of Medical Sciences, New Delhi, India. Currently, he is working as an internal medicine resident physician at Saint Mary's Hospital, an affiliated hospital of Yale University, CT, USA. His work has been published in journals such as *Metabolic Syndrome and Related Disorders* and *Indian Journal of Cancer*.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

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