

A Rare Case of Atrial Myxoma Presenting with Splenic Infarction

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Abstract

Background: Cardiac myxomas are the most frequent benign primary cardiac tumors. Clinical symptoms vary from constitutional to clinical characteristics caused by intracardiac blockage, such as mitral stenosis, coronary embolization, or systemic embolization. The only effective therapy for this disabling and fatal condition is surgical resection.

Case Presentation: A 17 years old male patient, with no chronic medical illness, presented to emergency department by his family due to repeated non-projectile vomiting of everything. He eats for 2 months duration associated with loss of appetite, generalized fatigue, low grade fever and weight loss of about 40kg in months. Patient underwent computed tomography (CT) scan which revealed splenic infarction. Patient underwent echocardiography showed large left atrial myxoma.

Discussion: The majority of cardiac myxomas are spontaneous and develop as a solitary lesion in middle-aged population from the left atrium. The preferred diagnostic method is echocardiography. Long-term survival following surgical excision is excellent, and recurrence is uncommon.

Keywords: Atrial Myxoma; Splenic Infarction; Case Report; Echocardiography

Introduction

Cardiac tumors are quite uncommon in medicine. Primary cardiac tumors are rare, with an estimated prevalence of less than 0.03 percent, 75 percent of which are benign and half of which are myxomas. Cardiac myxoma (CM) is still a rare clinical entity, with an incidence of 0.5 - 0.7 cases per million people and a frequency of 5 per 10,000 people [1]. They are most common during the third and sixth decades of life, with a female preponderance. In 75% of cases, the injured site is in the left atrium, whereas in 15% of cases, it is in the right atrium. The embolic phenomenon occurs in 40 - 50 percent of cases [2].

Atrial myxomas are thought to be caused by an entrapped embryonic foregut. They typically arise with emboli due to their beginnings in multipotent mesenchymal cells capable of both neural and epithelial development. They reveal scattered cells inside a mucopolysaccharides stroma under the microscope [3]. They are pedunculated and gelatinous on the surface, with a smooth, villous, or friable surface [4].

Clinical signs might vary greatly. Most tumors emit fragments or thrombi into the bloodstream, causing embolization and systemic and pulmonary symptoms. The most serious result is of a neurological origin, with stroke being the most common. Other symptoms include heart failure, arrhythmia, and pericardial effusion [4].

There is currently no effective medicinal treatment that can stop the tumor’s development; hence, surgical removal of the tumor mass is the best therapeutic option. We provide a case of a 17-year-old guy who complained of vomiting, loss of appetite, and weight loss. He was thoroughly checked to determine the source of his vomiting and weight loss, and a rare diagnosis of atrial myxoma was obtained.

Case Presentation

A 17 years old male patient, with no chronic medical illness, presented to emergency department by his family due to repeated non-projectile vomiting of everything he eats for 2 months duration associated with loss of appetite, generalized fatigue, low grade fever and weight loss of about 40 kg in months. Patient also had chronic dry cough for 2 months. Patients also had an emotional stress due to general high school exam. Patient had no history of trauma. Systematic review during history taking revealed only vomiting and dry cough. Otherwise, no palpitation, no orthopnea, no urinary symptoms, no skin manifestations and no musculoskeletal symptoms were noticed. Patient had one previous hospitalization due to abdominal pain and appendicitis was suspected, but was ruled out by investigations and imaging.

Upon physical examination, patient was conscious, oriented, but looked ill, pale, not jaundiced, and not dyspneic. There is temporal muscle wasting and his BMI was 21.6 kg/m². There was finger clubbing. His vital signs were: blood pressure was 122/78 mmHg; temperature 37.8; pulse was regular rhythm, heart rate 122 beats/min and SaO₂ was 98% on room air. Nothing was remarkable in the examination. Laboratory investigations upon admission are presented in table 1.

Complete Blood Count		
HGB		9.1
MCV		65
WBC		7
PLT		477
Clinical Chemistry		
Cr		1.0
Urea		21
RBS		116
ALT		23
AST		20
Albumin		3.7
Total protein		6.4
Bilirubin total, direct		0.8, 0.1
Uric acid		4.7
LDH		553
Na		139
K		3.8
Ca		1.24
Mg		1.68
Phosphorus		5
ESR		115
INR		1
Reticulocyte count		1.8
Blood Film		
Microcytic RBCs with hypochromia, few poikilocytes, Neut 64%, eosinophil 3%, lymph 22%, increased no. of platelets		
Arterial Blood Gases		
pH		7.47
PCO ₂		40
HCO ₃		29

Table 1: Laboratory investigations upon admission.

Patient underwent several abdominal ultrasound imaging with a conclusion of normal study. Patient's ECG is presented in figure 1.

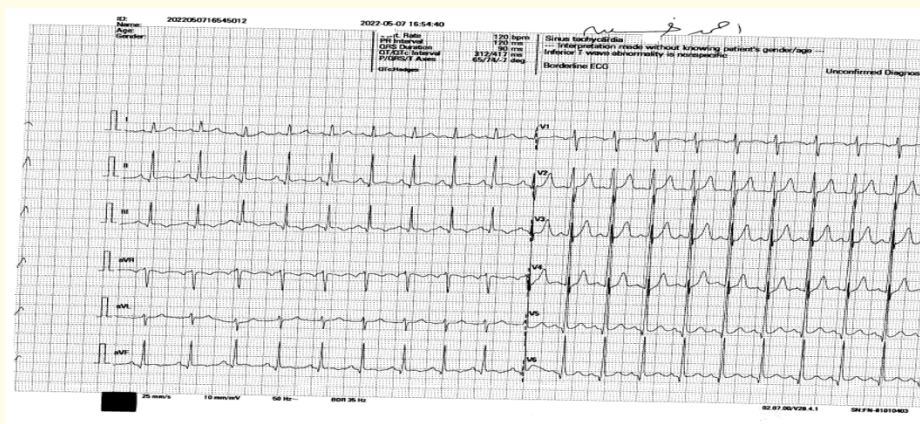


Figure 1: Electrocardiogram shows normal sinus rhythm.

Patient underwent computed tomography (CT) scan which revealed splenic infarction as presented in figure 2.

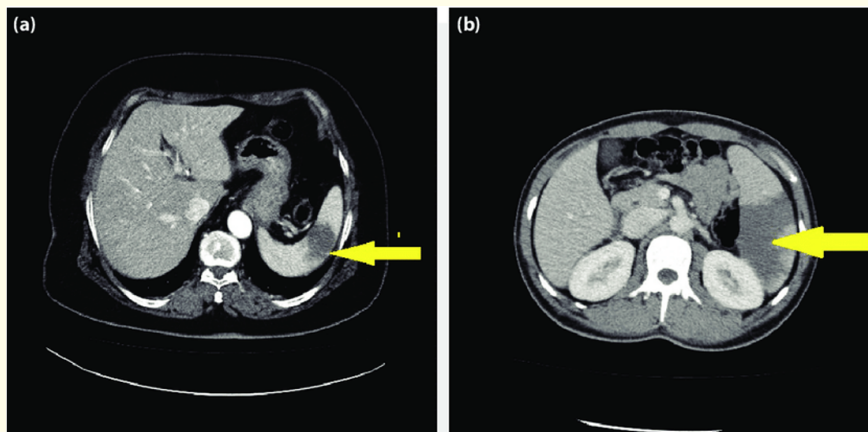


Figure 2: CT scan showing huge splenic infarction (Yellow arrow).

After that, patient underwent echocardiography with the following results:

- Normal internal dimensions
- Ejection fraction: 55%
- Large mass in the left atrium protruding in the left ventricle causing severe mitral stenosis (Figure 3).

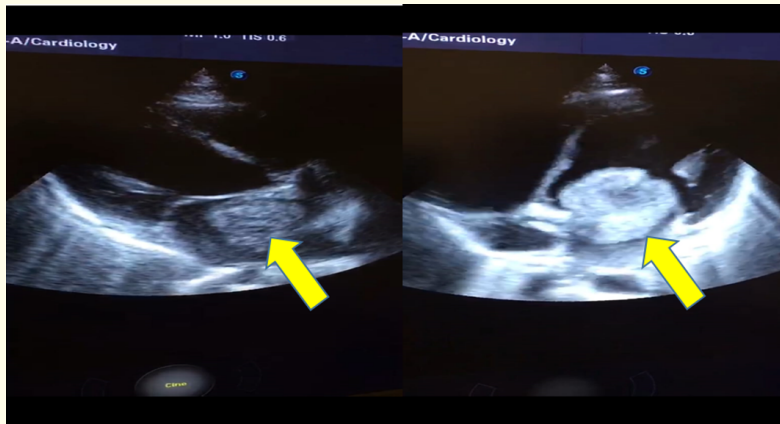


Figure 3: Echocardiography showing left atrial myxoma (Yellow arrow).

Finally, the patient underwent left atrial mass excision and mitral valve band annuloplasty (Cosgrove-Edwards band 30 mm).

Discussion

Cardiac myxomas (CMs) are the most prevalent kind of primary cardiac tumor. The total incidence ranges from 0.0017 to 0.02 percent, with the majority of cases presenting as benign tumors in the left atrium (60 - 75 percent) [1]. The majority of instances are sporadic, with no known cause, and women aged 30 - 60 years are twice as likely to be afflicted. Carney syndrome is an autosomal dominant inheritance cluster that occurs in only around 3 - 10% of all cases [2]. CMs appear as a slimy, friable lesion with a smooth or villous surface, with the latter having a higher risk of embolic consequences [2,3]. They can be sessile, emerging from a wide base, or they can emerge from a stalk or pedicle with an average diameter of 5 - 6 cm and a maximum height of 15 cm [2,5]. Most tumors are pedunculated, positioned in the atria, and discovered in the fossa ovalis area [1,2], as seen in figure 2A. Patients under the age of 40 with non-left atrial location or numerous CMs should be investigated for "syndrome myxoma," which is linked with blue nevi, cutaneous lentiginosis, peripheral tumors, and endocrine neoplasms [1,4,5].

Clinical symptoms might be induced by the tumor directly through blockage or indirectly through embolization. In 52 - 67 percent of patients, symptoms are caused by intracardiac blockage [5]. They are caused by either a blockage of the mitral orifice or a wrecking-ball effect, which causes valve damage and mitral regurgitation [4]. Pulmonary edema, dyspnea, orthopnea, malaise, syncope, and palpitations can also occur [1]. The second most frequent consequence is embolization (30-40%), which causes neurologic sequelae (12 - 30%) or systemic infarction (4.1-23%) of the extremities, splanchnic, or coronary circulations [2,5-7]. It is crucial to highlight that embolic events are not affected by tumor size and can occur in tiny tumors as well [3]. Our patient was one of the rare cases presenting with splanchnic infarction as seen in figure 2.

In asymptomatic individuals, the diagnosis of CMs is frequently an accidental discovery on imaging for another reason [4]. On imaging, a tentative diagnosis is determined upon the exclusion of thrombus or vegetation and the presence of a mobile mass connected by stalk or a stalk remained after the mass has been systemically embolized [1]. To assist better visualize the intracardiac mass for preoperative planning, echocardiography (Transesophageal > Transthoracic) is the diagnostic test of preference, followed by cardiac MRI and CT scans [2,3]. If an echo does not reveal a stalk or pedunculated mass, MRI can be utilized to diagnose CMs and differentiate them from thrombus

or pseudotumor [3]. CMs on MRI are heterogeneous in cine imaging due to intermittent calcifications or hemosiderin-related artifacts, hyperintense on T2 weighted images, and isointense on T1 weighted images [2,3]. If an MRI is contraindicated or inconclusive, a CT can be used to detect a left atrial mass with a limited connection to the atrial septum and heterogeneous low attenuation [2,3].

On CT, CMs may be distinguished from thrombi based on their size, origin, form, movement, and prolapse. CMs are bigger, start in the fossa ovalis (vs. an appendage in thrombi), are villous shaped (vs. polypoid in thrombi), are very mobile, and can prolapse through the mitral valve, whereas thrombi never do [8].

To reduce the danger of systemic embolization, cerebral infarction, and sudden death, surgical excision should be performed as soon as possible [6]. The overall prognosis is favorable, with recurrence rates ranging from 1 to 5% due to inadequate resection, intraoperative tumor seeding, or tumor multifocality [2]. The 5-year survival rates for benign tumors are 83 percent, 30 percent for malignant neoplasms, and 26 percent for metastases [5].

Conclusion

Cardiac myxomas should always be considered as a possible cause of embolism in otherwise healthy patients with systemic thromboembolism. Early detection and surgical excision are critical for preventing additional embolization.

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