Dubai Medical Journal

Educational Corner – Case Report

Dubai Med J
DOI: 10.1159/000523927

Received: June 25, 2021
Accepted: March 1, 2022
Published online: April 4, 2022

A Case Report of Atrial Myxoma in the Time of Corona Presenting with Systemic Embolization

Maher Younes[a] Alma Al Mansour[b] Mohammad Jawdat Al Halaki[b]
Lubna Lutfi[b]

[a]Department of Emergency, Rashid Trauma Center, Dubai Health Authority, Dubai, United Arab Emirates;
[b]Medical Internship Program, Medical Education Department, Dubai Health Authority, Dubai, United Arab Emirates

Keywords
Atrial myxoma · Acute limb ischemia · Multiple infarcts · COVID-19

Abstract
Myxomas are very rare intracardiac benign tumors. They can arise in any of the cardiac chambers, although 75% occur in the left atrium. Atrial myxomas (AMs) show a female predominance with a sex ratio of 3:1. There is a diversity in the clinical presentation; it may resemble many cardiovascular or systemic diseases. Furthermore, it is possible to present as an acute upper or lower limb ischemia or organ infarction due to systemic emboli. Hereby, we report a previously healthy 38-year-old male, who presented to the emergency department with acute pain of both lower limbs, unilateral upper limb, and acute abdomen. A bedside transthoracic echocardiogram was done which showed a mass that was confirmed to be an AM. The patient underwent surgical embolectomy and tumor resection. As this tumor is rare, the duration between the onset of symptoms to finally reaching the correct diagnosis is usually prolonged, and meanwhile, the patient can experience irreversible neurological damage or even death. Hence, we would like to highlight the role of echocardiogram in detecting AMs. In addition, excluding the primary cause of the thrombi, the patient's ischemic pain has a spectrum of differential diagnosis ranging from peripheral vascular disease to COVID-19. We would also like to emphasize the importance of a high index of suspicion by emergency physicians when such cases present.

Background

Atrial myxomas (AMs) are the most common primary benign tumors of the heart that account for 50% of all cardiac tumors and 0.25% of all heart disease [1, 2]. They have a spectrum of clinical presentations; nearly 50% of patients present with symptoms that result from either central or peripheral embolism or intracardiac obstruction, in addition to constitutional symptoms. However, 10% of patients may be completely asymptomatic [3]. Myxomas can arise in any of the cardiac chambers, although 75% occur in the left atrium [4]. Echocardiography is the gold standard for diagnosis of myxomas and surgical resection is the definitive treatment. A rare but tragic complication of AMs is complete myxoma embolization to the peripheral vessels.

Karger@karger.com
www.karger.com/dmj

Correspondence to:
Alma Al Mansour, alma_almansour@outlook.com

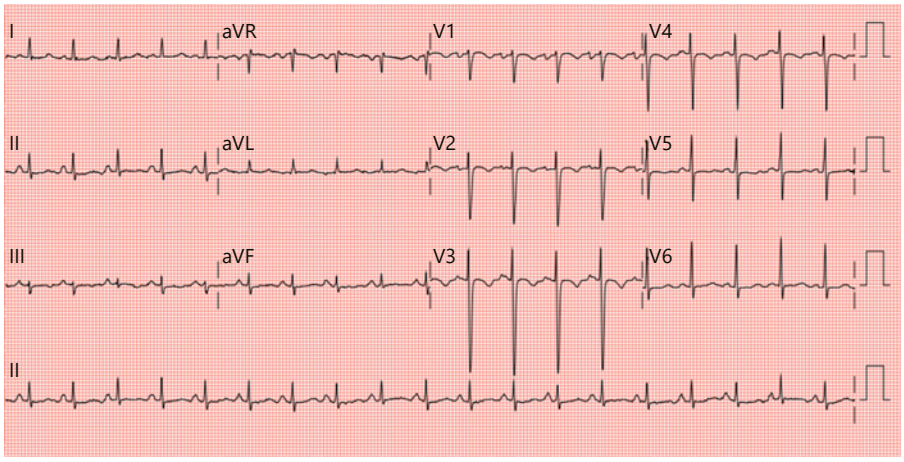

Fig. 1. ECG showing sinus tachycardia, extensive T-wave changes, and left ventricular hypertrophy. ECG, electrocardiography.

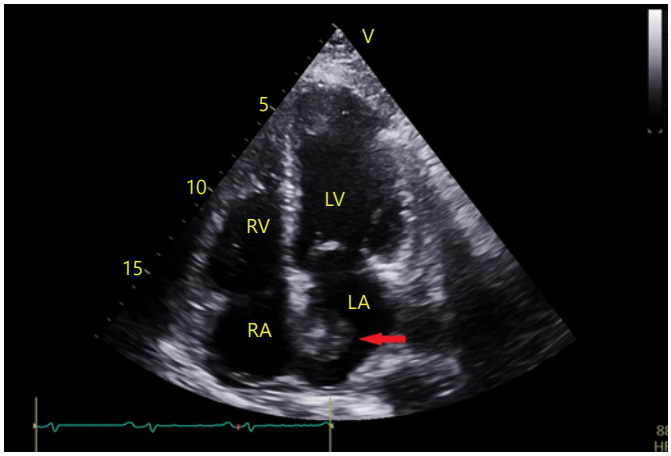

Fig. 2. Transthoracic echocardiogram showing left AM: large lobulated fragile mass seen attached to fossa ovalis, measuring 2.2 × 2.4 cm.

Table 1. Blood investigations were carried out to assess inflammatory markers, cardiac enzymes

Laboratory	Value	Reference range
WBC	14.1×10^3/μL	$3.6–11.0 \times 10^3$/μL
Neutrophils	10.6×10^3/μL	$2.0–7.0 \times 10^3$/μL
Hematocrit, %	38.9	40.0–50.0
Random blood glucose	230 mg/dL	65–140 mg/dL
HbA1c, %	6.1	<5.7
CRP	26.1 mg/L	<5.0 mg/L
Procalcitonin	0.09 ng/dL	<0.05 ng/dL
ESR	53 mm/1 h	2–28 mm/1 h
D-dimer	2.47 μg/mL	0.5 μg/mL
Troponin	92 ng/L	<14 ng/L
Natriuretic pro-BNP	2,550 pg/mL	<125 pg/mL

CRP, C-reactive protein; ESR, estimated sedimentation rate.

Case Report

A 38-year-old previously healthy male presented to the emergency department (ED) complaining of a sudden onset of bilateral lower limb pain, as well as abdominal pain associated with lightheadedness. The lower limb pain started abruptly in both legs 30 min prior to his arrival to the ED; it was nonradiating and was sharp in nature. It progressively worsened and was not relieved or exacerbated by any factors or movements, and upon presentation, the patient reported a pain score of 8/10. This is the first episode of this nature; the patient denied any history of claudication in the past.

Systemically, he did not complain of palpitations, loss of consciousness, cough, fever, or vomiting. However, he reported intermittent chest pain along with difficulty in breathing on mild exertion for the past month, as well as loss of appetite. The patient has no positive family history of heart disease. He does not smoke or drink alcohol and has no known allergies.

Upon evaluating the patient, he was tachycardic with a heart rate of 118 beats per minute, a slightly raised blood pressure of 150/101 mm Hg, but the other vitals were stable. On examination, he was alert and oriented to time, place, and person; however, he was agitated.

Cardiovascular examination revealed pallor and cold extremities with delayed capillary refill. Peripherally, he had absent bilateral pulses in the femoral, popliteal, posterior tibial, and dorsalis pedis arteries in the lower limbs as well as absent brachial and radial pulses in the right upper limb. Upon inspection of the chest, it appeared normal with no visible pulsations or previous surgical scars. Chest palpation was negative for heaves and thrills, and apex beat was palpated on the 5th intercostal space. Auscultation revealed clear lungs, regular heart rhythm, and normal heart sounds without a murmur. There also was no jugular venous distention.

Lower limb examination revealed bilateral weakness with a muscle power of 1/5, accompanied by loss of fine touch; however, deep reflexes were intact. No redness or edema was noted. Upper limb neurological examination was normal. Other system examinations were negative.

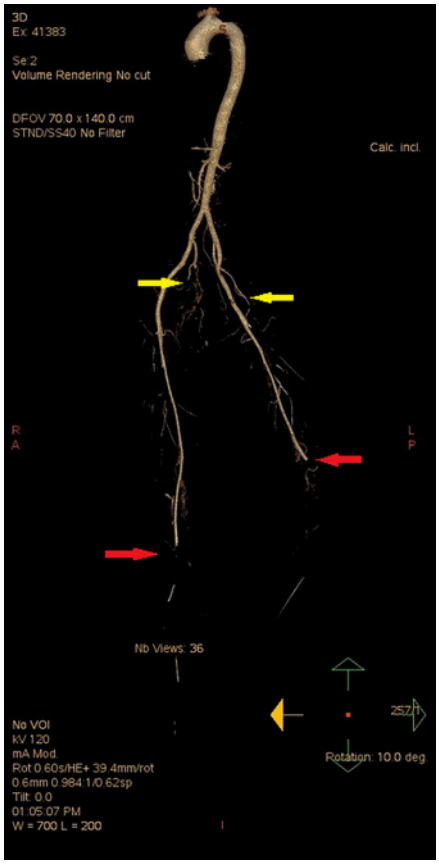

Fig. 3. 3D CT angiogram of the lower limb vessels showing total occlusion of bilateral deep femoral and popliteal arteries, bilateral posterior tibial arteries are opacified, bilateral dorsalis pedis are not seen. CT, computed tomography.

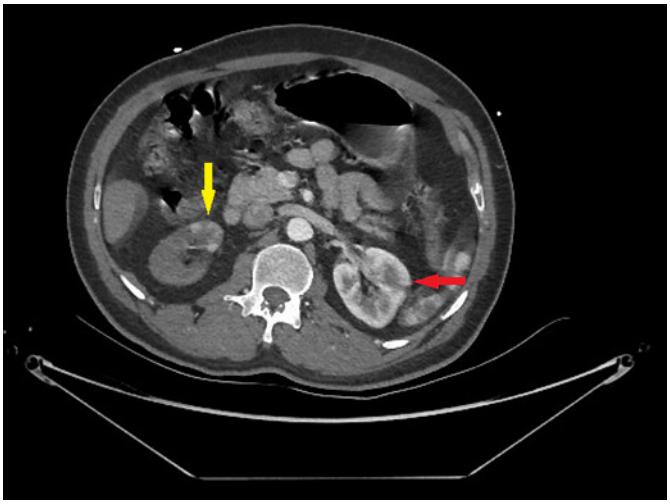

Fig. 4. Axial section abdominal CT angiogram showing wedge-shaped perfusion defect noted on the upper pole of the left kidney suggesting infarction due to segmental branch occlusion as well as infarction of the entire right kidney due to total occlusion of the right renal artery. CT, computed tomography.

Table 2. Arterial blood gas

Ref range and units		Value
PH	7.35–7.45	7.346
PCO_2	35–45 mm Hg	38.4 mm Hg
PO_2	83–108 mm Hg	27.2 mm Hg
HCO_3	21–28 mmol/L	19.7 mmol/L
Lactic acid	0.5–1.6 mmol/L	3.7 mmol/L

Initial bedside investigations in the ED included a 12-lead electrocardiography, a handheld doppler, and transthoracic echocardiogram. The electrocardiography showed sinus tachycardia with extensive T-wave changes and left ventricular hypertrophy as shown in Figure 1. Lower limb doppler revealed absent pulses bilaterally of popliteal, posterior tibial, and dorsalis pedis arteries with faint pulses in the femoral arteries (+1). In the upper limb, the right radial pulse was absent while the left was faint (+1), the rest of the pulses were intact. The bedside transthoracic echocardiogram revealed global hypokinesia, moderate to severe mitral regurgitation, and an ejection fraction of 35–40%. It also revealed the presence of an echogenic, mobile mass in the left atrium, measuring 2 × 3 cm compatible with a thrombus or a myxoma as shown in Figure 2.

Blood investigations were carried out to assess inflammatory markers and cardiac enzymes, revealing elevation in both parameters, shown in Table 1. Arterial blood gas was carried out revealing metabolic acidosis, shown in Table 2. The COVID-19 PCR test was negative. Furthermore, coagulation profile and creatinine were normal.

To further characterize the extent of the thrombosis, computed tomography angiography of the abdomen and lower limbs was carried out. It revealed total occlusion of bilateral deep femoral and popliteal arteries as shown in Figure 3 and total occlusion of right renal artery and segmental occlusion of the left renal artery with resultant renal infarction as shown in Figure 4. A splenic infarction was also noted due to segmental occlusion of splenic artery shown in Figure 5 and no dissection of the abdominal aorta.

A multidisciplinary team approach was undertaken; emergency medicine, cardiology, and vascular surgery physicians were involved in the management plan. The patient was initially started on furosemide 40 mg BID, spironolactone 25 mg OD, bisoprolol 2.5 mg OD, enoxaparin 60 mg BID.

He was admitted to the hospital and within an hour underwent an emergent bilateral femoropopliteal artery embolectomy to revascularize the lower limbs. Furthermore, 2 days post-admission, the patient developed compartment syndrome in his left leg post-procedure, and therefore, a left lower limb fasciotomy was done. On the same day, he was operated on for AM excision. The surgery included right atriotomy and septostomy on the fossa ovalis as well as mass excision from the left atrium which extended to the atrial septum. Grossly, the mass was 3 × 4 cm in size. Histologic examination confirmed a benign AM.

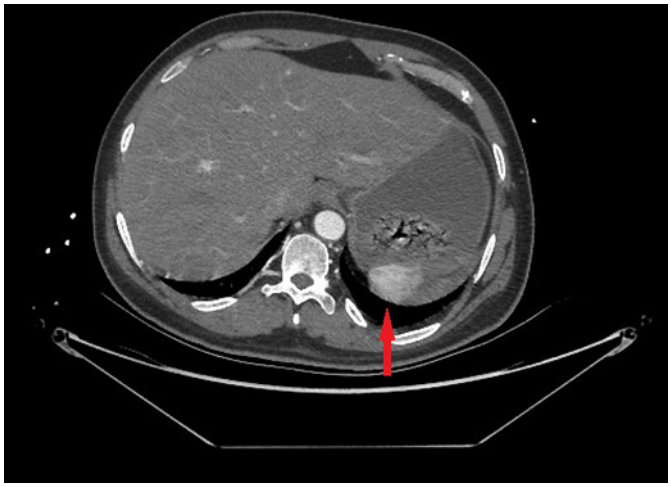

Fig. 5. Axial section abdominal CT angiogram showing a wedge-shaped perfusion defect of the upper pole of spleen likely due to segmental artery occlusion. CT, computed tomography.

The patient presented to the clinic for a follow-up visit 3 months post-surgery, he was doing well, free of disability or new complaints and reported no further embolic events. Repeated transesophageal echocardiography failed to indicate any residual intracardiac tumor.

Discussion

Cardiac tumors are generally rare with an incidence rate of <0.33%. They are classified as primary tumors, secondary tumors, or metastatic tumors. Primary cardiac tumors comprise 5% of all cardiac tumors and 75% of those are benign [5]. AMs make up most benign cardiac tumors with a female sex predominance of 70%. Approximately 90% of AMs are solitary tumors that arise from the left atrium and 75% of these tumors originate from the inferior aspect of the interatrial septum. Myxomas can be polypoid, round, or oval and can range in size from 1 to 15 cm in diameter. Sporadic myxomas are usually solitary and tend to present at older ages in contrast with familial myxomas which constitute 10% or less of all cases [6]. Familial cardiac myxomas can be part of a syndrome like Carney syndrome. Patients with familial myxoma syndromes are more likely to have them in atypical locations, have multiple tumors, have higher rates of recurrence after appropriate surgical interventions [7], and have associated dermatological and endocrine abnormalities [8].

Though the patient can remain asymptomatic, the clinical presentation of AMs is often unspecific. Constitutional signs include fever, weight loss, Raynaud's phenomenon, arthralgia, and myalgia. Signs of heart failure and cardiac obstruction can result from the mass obstructing the blood flow. Furthermore, prolapse of the mass can result in valve destruction and therefore valve disease [3, 6].

In regard to AM complications, it includes a range of sequelae from congestive heart failure, cardiac arrhythmias, rupture, and myocardial infarction to sudden death. In addition, because of the friable nature of the tumor, systemic embolization is a common complication that happens in 30–40% of cases, making it the most common cause of tumor embolism of all [9]. Rarely, myxomas can become infected and thus increases the embolic risk. Streptococcus is the most frequently incriminated germ.

Moreover, bedside echocardiography is the imaging modality of choice for diagnosis. Transthoracic echocardiography is approximately 95% sensitive for the detection of cardiac myxomas, and transesophageal echocardiography approaches 100% sensitivity [10]. The importance of bedside ultrasonography for early diagnosis in such presentations is also highlighted [11]. The most important clue to diagnosing AMs is their location in the left atrium and origin from the midportion of the atrial septum [8]. This cannot be elicited properly using other imaging modalities such as computed tomography or MRI [12]. Surgical removal of the myxoma is the treatment of choice and is usually curative; however, recurrence is possible and is most frequently associated with familial syndrome but can also occur due to incomplete tumor resection.

The novelty of this case lies in the fact that AMs showed a higher prevalence in females, but in our case, it was a male patient; previous cases reported in the Arabian Gulf region reported female patients [13, 14]. Our case also depicts an unusual presentation of acute lower extremity ischemia, splenic and renal infarction due to complete embolization that showered from a left AM, while the patient had absolutely no symptoms prior to the embolic events. Henceforth, this report puts emphasis on the fact that AM, though rare, should be considered as a source of embolism in patients presenting with acute limb ischemia. A case series study published in the year 2017 found that 4 out of 46 cases of left AM experienced multisystem embolization [15]. Common causes of embolism must also be excluded, including COVID-19 viral infection which is known to cause a hypercoagulable state [16]. Finally, high index of suspicion is essential, especially that this entity is rare, and early diagnosis is crucial in such cases as it has significantly reduced the morbidity in our patient and helped overcome further complications.

Conclusion

AM has not been reported in a male patient before in the region, he was also completely asymptomatic prior to his acute presentation. This report puts emphasis on the fact that AM, though rare, should be considered as a source of embolism in patients presenting with acute limb ischemia and organ infarction. As during the COVID-19 pandemic, the viral infection is known to cause a hypercoagulable state; a high index of suspicion should be there to exclude other possible causes of embolization. Early diagnosis using a bedside echocardiogram and quick surgical intervention is essential to reduce morbidity and prevent complications.

Statement of Ethics

This case report complying with the guidelines for human studies was conducted ethically in accordance with the World Medical Association Declaration of Helsinki. Ethical approval was not required for this case report in accordance with the Dubai Scientific Research Ethics Committee policies. Written informed consent for publication of the case report and any accompanied images was obtained from the patient.

Conflict of Interest Statement

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding Sources

The authors have not declared a specific grant for this research from any funding agency in the public, commercial, or not-for-profit sectors.

Author Contributions

All authors were involved in the acquisition and interpretation of the data, the article's conception and design, and the final approval of the version to be published.

Data Availability Statement

All data generated or analyzed during this case report are included in this article. They are available from the corresponding author (A.A.M.) upon reasonable request.

References

1 Fayard JM, Maurice P. Myxome cardiaque. Conférence de Cardiologie. 1980;25:31–3.
2 Goldberg HP, Glenn F, Dotter CT, Steinberg I. Myxoma of the left atrium; diagnosis made during life with operative and post-mortem findings. Circulation. 1952;6(5):762–7.
3 Boutayeb A, Mahfoudi L, Moughil S. Atrial myxoma: from diagnosis to management. Clin Surg. 2017;2:1498.
4 Yuan SM, Yan SL, Wu N. Unusual aspects of cardiac myxoma. Anatol J Cardiol. 2017;17(3):241–7.
5 Ren DY, Fuller ND, Gilbert SAB, Zhang Y. Cardiac tumors: clinical perspective and therapeutic considerations. Curr Drug Targets. 2017;18(15):1805–9.
6 Pernick N. Cardiac myxoma. PathologyOutlines.com, Inc; 2015 [cited 2021 Apr 25]. Available from: https://www.pathologyoutlines.com/topic/hearttumormyxoma.html.
7 Zipes DP, Libby P, Bonow RO, Mann DL, Tomaselli GF, Braunwald E. Braunwald's heart disease a textbook of cardiovascular medicine. 11th ed. Philadelphia (PA): Elsevier; 2019.
8 Armstrong WF, Ryan T, Feigenbaum H. Feigenbaum's echocardiography. Philadelphia: Wolters Kluwer; 2019.
9 Sharma GK. Atrial myxoma. Background, pathophysiology, etiology. Medscape. 2020 [cited 2021 Apr 25].
10 Percell RL, Henning RJ, Siddique PM. Atrial myxoma: case report and a review of the literature. Heart Dis. 2003;5(3):224–30.
11 Ragland MM, Tak T. The role of echocardiography in diagnosing space-occupying lesions of the heart. Clin Med Res. 2006;4(1):22–32.
12 Otto CM. Textbook of clinical echocardiography. Philadelphia: Saunders Elsevier; 2018.
13 Karkout KA, Amrani S, Jamil G. Atrial myxoma – the first case report in Emirati patients. Hamdan Med J. 2016;10(2):129–35.
14 Al-Zamkan BK, Hashem AM, Alaaeldin SA, Abdel Aziz M. An exceptionally giant left atrial myxoma: a case report and literature review. Eur Heart J Case Rep. 2020;4(6):1–7.
15 Zhang RD, Zeng ZH, Zheng JY, Li TD, Zhao YQ, Liu YH, et al. Left atrial myxoma complicated with multi-system embolization. J Cardiothorac Surg. 2017;12(12):76.
16 Ranucci M, Ballotta A, Di Dedda U, Bayshnikova E, Poli MD, Resta M, et al. The procoagulant pattern of patients with COVID-19 acute respiratory distress syndrome. J Thromb Haemost. 2020;18(7):1747–51.