The Journal of Emergency Medicine, Vol. 57, No. 5, pp. 710–712, 2019

0736-4679/$ - see front matter

https://doi.org/10.1016/j.jemermed.2019.06.006

# Ultrasound in Emergency Medicine

## ATRIAL MYXOMA PRESENTING AS ACUTE BILATERAL LIMB ISCHEMIA

Roshan Mathew, MD, Naman Agrawal, MD, Praveen Aggarwal, MD, and Nayer Jamshed, MD

Department of Emergency Medicine, All India Institute of Medical Sciences, New Delhi, India
*Corresponding Address:* Roshan Mathew, MD, Department of Emergency Medicine, All India Institute of Medical Sciences, Ansari Nagar, New Delhi 110029, India

☐ **Abstract—Background: Cardiac myxoma is the most common primary benign tumor of the heart and it has diverse clinical presentations. It is known to embolize into systemic circulation. However, presentation with complete occlusion of the aorta is uncommon. Case Report: We report an 18-year-old female who presented to the emergency department with features of acute bilateral limb ischemia. Arterial Doppler ultrasonography showed infrarenal aortic occlusion. A bedside cardiac ultrasound was done in the emergency department which clinched the diagnosis of atrial myxoma. Complete surgical excision of the tumor and subsequent histopathologic examination confirmed the diagnosis of atrial myxoma. Why Should an Emergency Physician be Aware of This?: This report puts emphasis on the fact that atrial myxoma, though rare, may be considered as a source of embolism in patients presenting with acute limb ischemia. The importance of bedside ultrasonography for early diagnosis in such presentations is also highlighted.** © 2019 Elsevier Inc. All rights reserved.

☐ **Keywords—acute limb ischemia; myxoma; POCUS**

## INTRODUCTION

Atrial myxomas are benign cardiac tumors that can have life-threatening clinical presentations because of mechanical cardiac obstruction, dysrhythmias, or peripheral embolization (1). There can be tumor fragmentation or complete tumor detachment, which can embolize, causing syncope, dyspnea, neurologic symptoms, or ischemic limb pain (2,3). Myxomas originate from entrapped embryonic foregut and have multipotent mesenchymal cells capable of both neural and epithelial differentiation (1,4). Operative mortality is <5% with early diagnosis and immediate surgery, resulting in good recovery of the patient (5). We report a patient who presented with acute bilateral lower limb ischemia and was diagnosed with atrial myxoma with complete aortic occlusion in the emergency department. The aortic occlusion was caused by embolization of the myxomatous tissue.

### *Case Presentation*

An 18-year-old female came to the emergency department with complaints of pain and weakness in both the lower limbs for the past 3 days. The patient had been previously healthy with no pertinent medical, social, or family history. On presentation, she was conscious, with a respiratory rate of 18 breaths/min and oxygen saturation of 98% on room air. She had a radial pulse rate of 86 beats/min and blood pressure of 120/68 mm Hg in the right arm. The results of her cardiopulmonary examination were normal, with no murmurs or audible extra heart sounds. On local examination of lower limbs, there was bluish discoloration of all the toes of her left lower limb. There was no apparent swelling or skin ulceration. There was no line of demarcation for the dusky/bluish



Received: 22 April 2019; Final submission received: 27 May 2019;
Accepted: 8 June 2019

discoloration. Both lower limbs were cold to the touch. Femoral, popliteal, posterior tibial, and dorsalis pedis pulses were absent bilaterally. Touch and pain sensations were decreased in both the lower limbs. Muscle power was 3 of 5 in the extensors and flexors of both the lower limbs around all joints. An arterial Doppler ultrasound of her abdomen and lower limbs revealed a filling defect in infrarenal aorta (possible thrombus) with dampened flow in bilateral lower limb arteries. An immediate bedside cardiac ultrasound was done to find any cardiac source of embolization. It revealed a large (20 mm × 57 mm) lobulated left atrial mass attached to interatrial septum (Figures 1 and 2). The mass was heteroechoic in appearance and was prolapsing into left ventricle during diastole (Video 1). In addition, there was mild mitral regurgitation. Her hemoglobin was 12.4 g/dL, her total leukocyte count was 14,300 $\mu$L, and her platelet count was 1.62 × $10^5$ $\mu$L. The 12-lead electrocardiogram showed a normal sinus rhythm and chest radiography was also normal.

The patient was started on unfractionated heparin and was immediately taken up for surgery. Bilateral aortofemoral embolectomy was performed followed by excision of the left atrial mass. The left atrial mass (Figure 3) was found attached to the interatrial septum. The embolized tumor was at the aortofemoral bifurcation with distal thrombus in left lower limb (Figure 4) and no distal thrombus on the right side. Since the left lower limb had already developed gangrene because of the late presentation of the patient to the emergency department, an above-knee amputation of the left lower limb was performed. Three days later, her right leg developed tense swelling for which right lower limb fasciotomy was performed. She was discharged and enrolled in the institutional rehabilitation center for further care. The excised tumor was sent for histologic examination and it revealed papillary frond like structure with a myxomatous matrix and dispersed myxoma cells showing calretinin positivity on immunostaining.

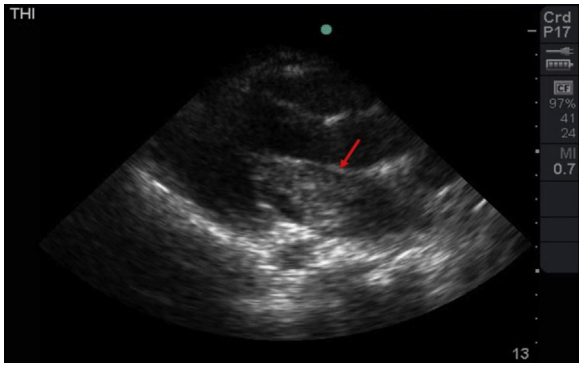


**Figure 1. Parasternal long axis view showing a large mass in the left atria.**

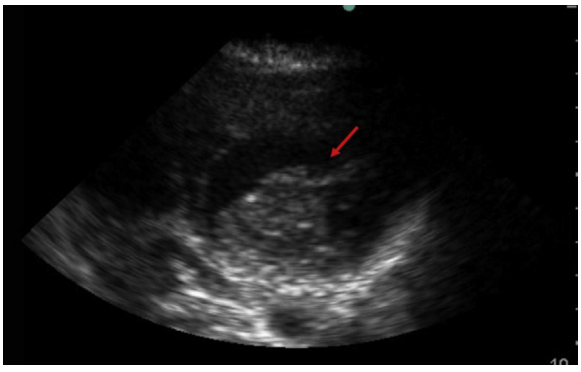

**Figure 2. Parasternal short axis view showing a large mass in the left atria.**

## DISCUSSION

Acute limb ischemia is a sudden decrease in limb perfusion that threatens limb viability and requires urgent intervention (6). Eighty percent of emboli to peripheral arteries have a cardiac source (rheumatic valve disease, atrial fibrillation, mural thrombus, infective endocarditis, prosthetic valve thrombus, or tumors), of which cardiac tumors account for only 1.5% (7). Definitive surgery is usually needed to salvage the limb in such cases. Use of point of care ultrasound has traditionally been described for life-threatening cardiac conditions like cardiac tamponade, pulmonary embolism, and aortic dissection (8). It is also a valuable tool for emergency physicians in the setting of acute limb ischemia and empowers providers to look for a possible cardiac source, thus facilitating decision making and early intervention.

Embolic phenomena in cardiac myxoma occurs in about 30–40% of cases (1). However, the incidence reported from Indian studies is around 9% (9). Emboli usually show a predilection for cerebral circulation but can affect other organs, such as the liver, spleen, retina,

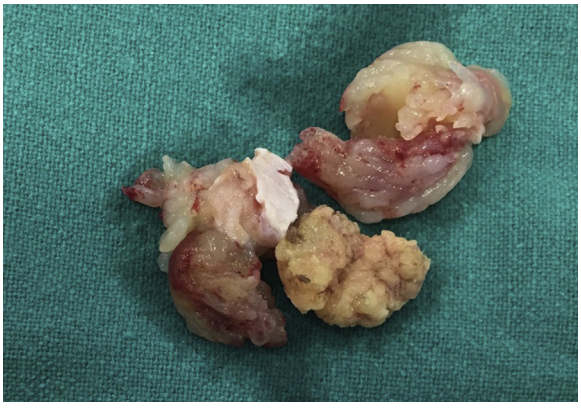

**Figure 3. Myxoma mass excised from the left atria showing an irregular surface.**

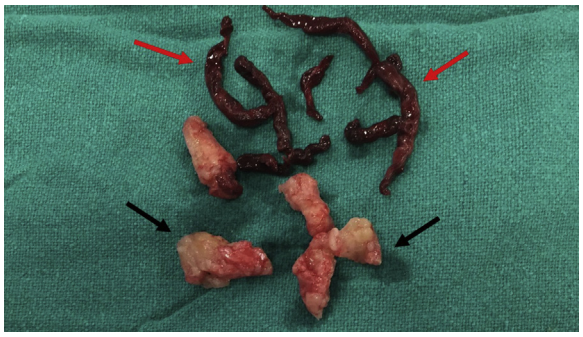

**Figure 4. Aortic embolus (black arrow) with femoral thrombus (red arrow).**

coronary artery, and peripheral arteries. Large saddle emboli in the abdominal aorta without preceding symptoms of cardiac myxoma have been reported in a few patients (10,11). There have been reports of cases where total detachment of cardiac myxoma caused saddle embolization of the abdominal aorta (12,13).

On a macroscopic level, myxomas are usually pedunculated and gelatinous in consistency. Their surface may be smooth, villous, or friable. The incidence of systemic embolism is higher in tumors with an irregular and friable surface than in those with a smooth surface. It is also frequent for prolapsing and polypoid tumors to embolize (14,15). Larger myxomas (>5 cm) usually present with cardiac obstructive symptoms and smaller ones with systemic embolism (1,14). In our patient, the left atrial myxoma was polypoid in shape with an irregular surface, and it prolapsed across the mitral valve orifice.

Early surgical resection of the tumor is the mainstay of treatment to prevent further cardiovascular complications. Recurrence is uncommon except in familial cases. However, follow-up with echocardiography is recommended.

## WHY SHOULD AN EMERGENCY PHYSICIAN BE AWARE OF THIS?

Providers should keep in mind atrial myxoma as a differential in young patients with acute limb ischemia presenting to the emergency department. Prompt use of beside ultrasound can aid in diagnosis and reduce the time to intervention, which is critical for good patient outcomes in such cases.

## SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.jemermed.2019.06.006.

## REFERENCES


1. Pinede L, Duhaut P, Loire R. Clinical presentation of left atrial cardiac myxoma. A series of 112 consecutive cases. Medicine (Baltimore) 2001;80:159–72.
2. Markel ML, Waller BF, Armstrong WF. Cardiac myxoma. A review. Medicine (Baltimore) 1987;66:114–25.
3. St John Sutton MG, Mercier LA, Giuliani ER, Lie JT. Atrial myxomas: a review of clinical experience in 40 patients. Mayo Clin Proc 1980;55:371–6.
4. Amano J, Kono T, Wada Y, Zhang T, Koide N, Fujimori M, et al. Cardiac myxoma: its origin and tumor characteristics. Ann Thorac Cardiovasc Surg 2003;9:215–21.
5. Rahmanian PB, Castillo JG, Sanz J, Adams DH, Filsoufi F. Cardiac myxoma: preoperative diagnosis using a multimodal imaging approach and surgical outcome in a large contemporary series. Interact Cardiovasc Thorac Surg 2007;6:479–83.
6. Creager MA, Kaufman JA, Conte MS. Clinical practice. Acute limb ischemia. N Engl J Med 2012;366:2198–206.
7. Lyaker MR, Tulman DB, Dimitrova GT, Pin RH, Papadimos TJ. Arterial embolism. Int J Crit Illn Inj Sci 2013;3:77–87.
8. Seif D, Perera P, Mailhot T, Riley D, Mandavia D. Bedside ultrasound in resuscitation and the rapid ultrasound in shock protocol. Crit Care Res Pract 2012;2012:1–14.
9. Aggarwal SK, Barik R, Sarma TCSR, Iyer VR, Sai V, Mishra J, et al. Clinical presentation and investigation findings in cardiac myxomas: new insights from the developing world. Am Heart J 2007;154:1102–7.
10. Horn KD, Becich MJ, Rhee RY, Pham SM. Left atrial myxoma with embolization presenting as an acute infrarenal aortic occlusion. J Vasc Surg 1997;26:341–5.
11. Huang CY, Chang YY, Hsieh MY, Hsu CP. Atrial myxoma presenting as total occlusion of the abdominal aorta and its major four branches. J Chin Med Assoc 2012;75:349–52.
12. Neff CM, McCowan CL. Complete aortic occlusion caused by cardiac myxoma emboli. Am J Emerg Med 2008;26:110. e1–e2.
13. Coley C, Lee KR, Steiner M, Thompson CS. Complete embolization of a left atrial myxoma resulting in acute lower extremity ischemia. Tex Heart Inst J 2005;32:238–40.
14. Goswami KC, Shrivastava S, Bahl VK, Saxena A, Manchanda SC, Wasir HS. Cardiac myxomas: clinical and echocardiographic profile. Int J Cardiol 1998;63:251–9.
15. Ha JW, Kang WC, Chung N, Chang BC, Rim SJ, Kwon JW, et al. Echocardiographic and morphologic characteristics of left atrial myxoma and their relation to systemic embolism. Am J Cardiol 1999;83:1579–82. A8.