Clinical Case Report

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Stroke and peripheral embolisms in a pediatric patient with giant atrial myxoma

Case report and review of current literature

Yin Wu, MD, Xian-Ming Fu, MD, Xiao-Bo Liao, MD, Xinmin Zhou, MD*

Abstract

Rationale: Cerebral stroke with peripheral embolism due to left atrial myxoma is very rare in children. Misdiagnosis may occur because of nonspecific symptoms in the heart.

Patient concerns: We present a case of a 16-year-old boy who presented with ischemic stroke and embolisms in the lower extremity, caused by a giant left atrial myxoma.

Diagnoses: Left atrial myxoma.

Interventions: A giant gelatinous mass was completely excised, and the histopathological findings confirmed the diagnosis of atrial myxoma.

Outcomes: The temperature of the right lower extremity recovered gradually, and pulse of the right dorsalis pedis artery became palpable 10 days after the surgery. The strength of the bilateral lower extremity was level 5 at discharge.

Lessons: Our case, along with the review of the literature, highlights the fact that myxomas often initially present with multiple embolisms but with few cardiac symptoms. Transthoracic echocardiography should be performed immediately to make a definitive diagnosis.

Abbreviations: CT = computed tomography, CTA = computed tomography angiography, LA = left atrium, LV = left ventricle, MRI = magnetic resonance imaging, RV = right ventricle, TTE = transthoracic echocardiography.

Keywords: embolization, myxoma, pediatrics, stroke

1. Introduction

Cardiac myxoma is rare in children, and early diagnosis is challenging because of nonspecific symptoms. The initial presentations of the tumor range from neurological deficits to cutaneous eruptions, resulting from thrombi or embolization of tumor fragments.[1] We here describe a case of a child who initially presented with cerebral stroke and peripheral embolisms in the lower extremity. We also review the clinical features of 18 other cases of pediatric cardiac myxoma reported in the literature. Our description of these pediatric cases can alert others to suspect this rare disease in children who present with atypical symptoms, thereby reducing the mortality rate.

Editor: N/A.

The authors declare no conflicts of interest.

Department of Cardiovascular Surgery, The Second Xiangya Hospital, Central South University, Changsha, Hunan, People's Republic of China.

** Correspondence: Xinmin Zhou, Department of Cardiovascular Surgery, the Second Xiangya Hospital, Central South University, Changsha, Hunan 410011, People's Republic of China (e-mail: zhou6xinmin@163.com).*

Medicine (2018) 97:30(e11653)

Received: 28 March 2018 / Accepted: 28 June 2018

http://dx.doi.org/10.1097/MD.0000000000011653

2. Case report

A 16-year-old boy was transferred to our emergency department with complaints of left-sided hemiplegia for 2 months, along with pain and swelling in the right lower extremity for 1 day. The symptoms of left hemiplegia started 2 months before admission. Brain magnetic resonance imaging (MRI) in a local hospital revealed cerebral infarcts scattered throughout the right basal ganglia and lateral ventricle (Fig. 1). The patient was treated for dehydration and nerve nutrition, and he underwent rehabilitation training. The symptoms of left hemiplegia improved gradually. One day before admission to our hospital, pain symptoms and swelling in the right lower extremity developed. Transthoracic echocardiography (TTE) revealed a giant space-occupying mass (64×33 mm in diameter) in the left atrium with a high possibility of atrial myxoma (Fig. 2A).

The patient was then transferred to our hospital for surgical treatment. On admission the patient was conscious, and his vital signs were stable. Physical examination revealed a regular rhythm with no murmurs. The patient had no pulse in the right popliteal artery and his right foot was pale, cold, and insensate. The pulse of the left lower extremity was normal. The muscle strength of the right lower extremity was level 5, while that of the left side was level 4. Computed tomography angiography (CTA) confirmed the giant mass in the left atrium (Fig. 2B and C) and obstructions of the bilateral lower limb arteries (Fig. 3A). Surgical resection of the left atrial myxoma was performed through a median sternotomy under cardiopulmonary bypass. A giant gelatinous mass ($70 \times 60 \times 40$ mm) in the left atrium, which arose from the area of the fossa ovalis, was noted.

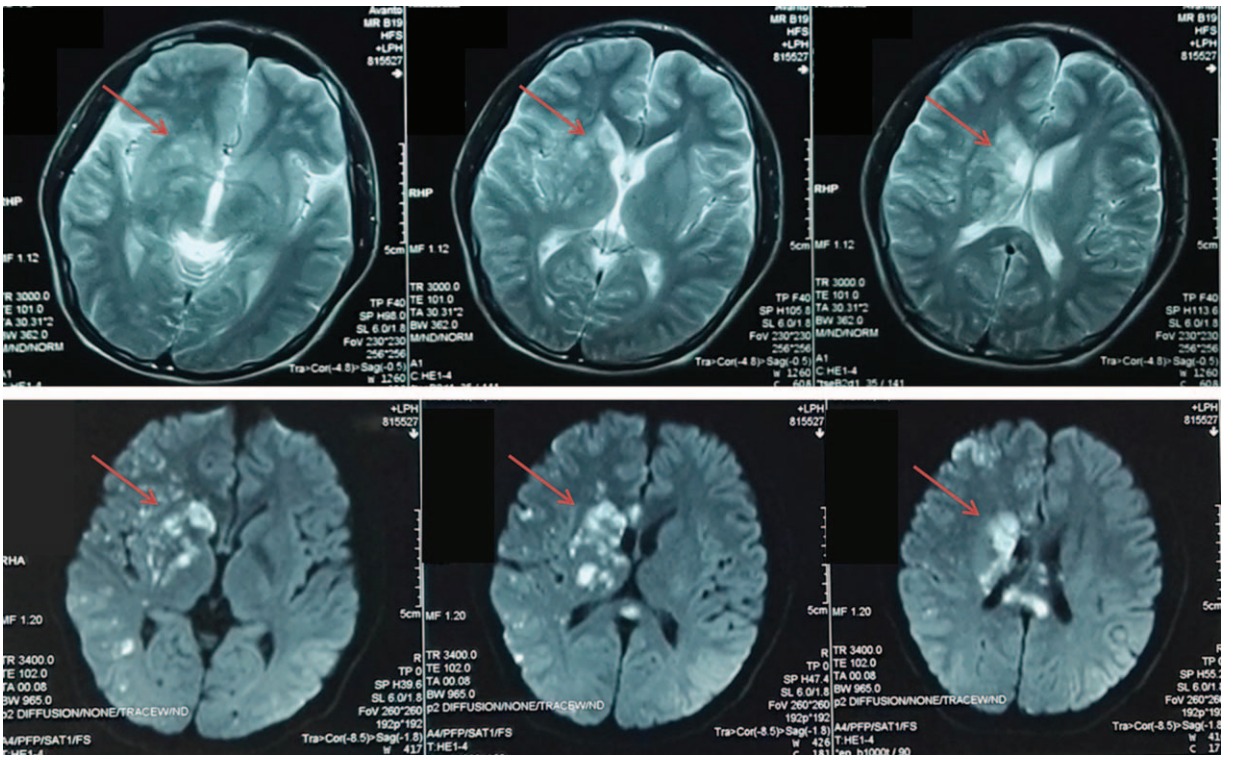

Figure 1. Brain MRI revealed cerebral infarcts scattered throughout the right basal ganglia and lateral ventricle lacunar. MRI = magnetic resonance imaging.

Complete excision of the tumor was performed, and subsequent histopathological findings confirmed the diagnosis of atrial myxoma. The conventional medical therapy such as dopamine and diuretics were performed once the surgery completed. The temperature of the right lower extremity recovered gradually and the pulse of the right dorsalis pedis artery became palpable 10 days after the surgery. The strength of the bilateral lower extremity was level 5 at discharge. CTA confirmed the revascularization of the bilateral lower limb arteries (Fig. 3B).

This study was performed according to the guidelines of the Medical Ethics Committee of the Second Xiangya Hospital of Central South University, in compliance with Helsinki Declaration of 1964 and later versions. The patient has provided informed consent for publication of the case.

3. Discussion

Within the pediatric population, primary cardiac myxoma is extremely rare. Most cardiac myxomas occur in the left atrium and arise in the area of the fossa ovalis. The majority of left atrial myxomas is histologically benign but may result in systemic embolization, atrioventricular valve obstruction, and constitutional symptoms. Of these, the most serious is cerebrovascular embolism, which may result in cerebral stroke. Previous reports have demonstrated that cerebral stroke is the first clinical manifestation in one third of cases.[2–4] In addition, embolization of coronary arteries, kidneys, intestines, and extremities can also occur. Cardiac myxomas are typically diagnosed by echocardiography. Computed tomography (CT) and magnetic resonance imaging (MRI) can help determine the location and the extent of the cardiac myxoma.

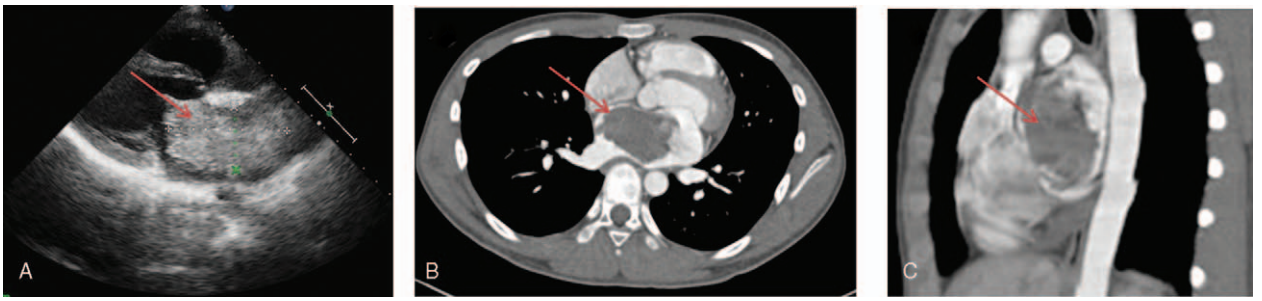

Figure 2. A giant space-occupying mass in the left atrium revealed by TTE (A) and CTA (B and C). CTA = computed tomography angiography, TTE = transthoracic echocardiography.

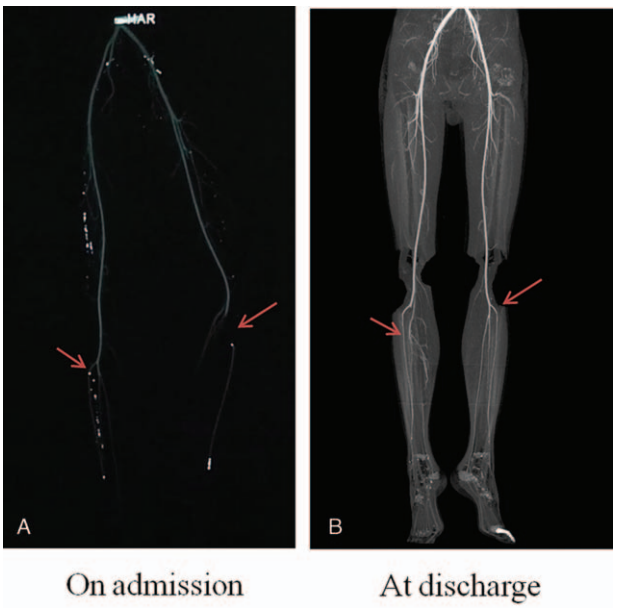

Figure 3. Obstructions of bilateral lower limb artery revealed by CTA (A). Revascularization of bilateral lower limb artery revealed by CTA (B). CTA = computed tomography angiography.

A few pediatric cases of cardiac myxoma with different clinical manifestations have been reported in the published literature, as summarized in Table 1. Based on their location, size, mobility, and embolism position, the clinical features of myxomas are quite variable. Early diagnosis might be challenging because of nonspecific symptoms, ranging from cardiovascular to neurological symptoms. Most often, less obvious cardiac symptoms are discovered only after an embolism.[5] Therefore, cardiac myxomas are often misdiagnosed in the pediatric population. In the 18 previously reported pediatric cases plus our own, 10 children presented with cerebral ischemic stroke prior to diagnosis. However, only 5 of the cases had a cardiac murmur at the time of presentation with cerebrovascular disease. The patient in our case had no previous medical conditions and neither cardiac nor constitutional symptoms before embolism occurred. Left atrial myxoma was not diagnosed by TTE until the peripheral embolism developed 2 months after his stroke. These findings were similar to the reported cases in the literature.

In the 18 previously reported cases, only 2 patients had erythematous rashes on the extremities, which were described as "red spots." This uncommon feature is important because it provides a clue that should prompt the urgent acquisition of an echocardiogram. Additionally, noncardiac signs and symptoms of cardiac myxomas such as fever and weight loss might result from the release of cytokines.

Table 1

Clinical summary of 19 pediatric cases of cardiac myxoma with different clinical manifestations.

Age/ Gender	Location of Tumor	Tumor size, mm	Cardiac symptoms	Neurological symptoms	Constitutional symptoms	Imaging features	Outcome	Reference
2 mos/M	LA	15*14	Heart murmur (grade 5, holosystolic)	None	None	None	Dead	[6]
5 ys/M	LA	None	None	Drowsy, sluggish, occasionally irritable, abnormal gait and seizure-like activity	None	Brain MRI: embolic strokes. Echocardiography: a large atrial mass	Normal	[7]
6 mos/F	LV	None	Heart murmur	None	Vomiting, hematochezia, anemia, acidosis, and anuria	CT: total embolic occlusion of the suprarenal abdominal aorta and an abnormal LV shadow defect. Echocardiography: LV tumor. Cardiac MRI: a solid LV tumor.	Normal	[8]
12 ys/M	LV	None	None	None	None	TTE: mobile-appearing mass attached to the posteromedial papillary muscle of the mitral valve	Normal	[9]
12 ys/F	LV	35*25	None	None	Fevers as high as 39.5 °C for 3 weeks	Echocardiography: a LV mass attached to the anterior mitral leaflet	Normal	[10]
11 ys/M	LA	44*35*28	None	Left-sided hemiparesis	Rash on distal limbs, splinter hemorrhages on nail beds	CTA: a complete occlusion of the proximal right M1 branch of the middle cerebral artery. TTE: a left atrial, pedunculated mass	Normal	[11]
13 ys/F	LA	50*40*30	None	Headache and dizziness for 18 months, loss of consciousness for the preceding 2 h	None	Cranial CT: a patch of low density in the area of the left basal nuclei and cerebral ganglion. Echocardiography: an enlarged left atrium with a LA mass	Normal	[12]
16 ys/F	LA	25*25	None	None	Chest pain, shortness of breath	Echocardiography: a LA mass	Normal	[13]
2 ys/F	RV	30*15*15	Heart murmur	None	None	Echocardiography: a mass occupying the main pulmonary trunk	Normal	[14]
12 ys/F	LA	35*26	None	Sudden dizziness and right-sided paresthesia of the lips and fingers	None	Brain MRI: demyelinating lesions or vasculitis. Echocardiography: a LA mass	Normal	[15]
11 ys/M	LA	40*30	None	Syncope, left-sided hemiparesis, facial droop and dysarthria.	Multiple blanching lesions in feet	Cranial CT: hypodense changes in the right hemisphere and complete occlusion of the middle cerebral artery	Normal	[16]
8 ys	LA	55*24	None	None	weight loss, anemia	None	Normal	[17]
5 ys /M	LA	40*40	None	Left-sided hemiparesis and dysarthria	None	Echocardiography: a LA mass	Normal	[4]
14 ys /F	LA	40 of the longest diameter	None	Syncope, transient left hemiparesis with expressive aphasia	fever and rash on the distal extremities	Cranial CT: negative. Cranial MRI: scattered restricted diffusion, acute arterial infarction. Echocardiography: a LA mass	Normal	[18]
8 ys /F	LA	30*30	None	Pain and weakness of the left leg, one episode of loss of consciousness	Skin rashes on the distal extremities	MRI: altered intensity in multiple areas of kidney cortex. Echocardiography: a LA mass.	Normal	[19]
12 ys /M	LV	25*50	Heart murmur	None	None	TTE: a LV mass. MRI: a heterogeneous, T2-enhancing LV mass.	Normal	[20]
11 ys /M	LA	50 of the longest diameter	None	Ischemic stroke	None	TTE: a LA mass	Normal	[21]
9 ys /M	LA	60*50*45	Heart murmur	None	Leg pain, paresthesia, and pallor	TTE and chest CT: a polypoid LA mass, mild mitral stenosis and regurgitation	Normal	[22]
16 ys /M	LA	70*60*40	None	Left-sided hemiplegia	pain and swelling in the right lower extremity	Cranial MRI: cerebral infarcts scattered throughout the right basal ganglia and lateral ventricle lacunar. TTE: a giant LA mass. CTA: a giant LA mass and obstructions of bilateral lower limb artery	Normal	Case 1

CT = computed tomography, LA = left atrium, LV = left ventricle, MRI = magnetic resonance imaging, TTE = transthoracic echocardiography.

Urgent surgical resection of myxoma is usually recommended to prevent further embolic complications. In our case, the atrial myxoma was surgically removed and the embolisms in the extremity resolved without any therapy. In most cases in the literature, early removal of the tumor has led to early rehabilitation. Sudden death from myxomas has only been reported in the case of Kure et al,[6] due either to tumor embolization with subsequent infarction or mechanical disturbances leading to conductive and hemodynamic aberrations. Early suspicion of this potentially curable tumor in children who present with neurological symptoms or peripheral embolisms can help reduce the mortality and disability rate.

4. Conclusion

Cardiac myxoma should be considered in any child presenting with cerebral stroke or peripheral embolism. TTE should be performed to reach a definitive diagnosis, and surgical resection is recommended to prevent further embolic complications.

Author contributions

Conceptualization: Xinmin Zhou.
Data curation: Yin Wu, Xiao-Bo Liao.
Formal analysis: Yin Wu.
Investigation: Yin Wu, Xian-Ming Fu.
Methodology: Xian-Ming Fu.
Project administration: Xinmin Zhou.
Resources: Xiao-Bo Liao.
Supervision: Xinmin Zhou.
Validation: Xiao-Bo Liao.
Writing – original draft: Yin Wu.
Writing – review & editing: Xian-Ming Fu, Xinmin Zhou.

References

[1] Schroeder L, Zyblewski S, Forbus G, et al. Left ventricular myxoma. J Pediatr 2016;168:249–52.
[2] Al-Mateen M, Hood M, Trippel D, et al. Cerebral embolism from atrial myxoma in pediatric patients. Pediatrics 2003;112:e162–7.
[3] Fuchs J, Leszczyszyn D, Mathew D. Cardiac myxoma causing acute ischemic stroke in a pediatric patient and a review of literature. Pediatr Neurol 2014;50:525–9.
[4] Juaneda I, Peirone A, Contreras A, et al. A rare cause of pediatric stroke. World J Pediatr Congenit Heart Surg 2017;8:220–3.
[5] Zhang R, Zeng Z, Zheng J, et al. Left atrial myxoma complicated with multi-system embolization. J Cardiothorac Surg 2017;12:76.
[6] Kure K, Lingamfelter D, Taboada E. Large multifocal cardiac myxoma causing the sudden unexpected death of a 2-month-old infant-a rapidly growing, acquired lesion versus a congenital process? Am J Forensic Med Pathol 2011;32:166–8.
[7] Fleurat MR, Zaia BE. Altered mental status in a child with an unwitnessed fall: a case report. Pediatr Emerg Care 2016;32:392–4.
[8] Kawabata T, Kasahara S, Ohtsuki S, et al. Left ventricular myxoma occluding the suprarenal abdominal aorta in an infant. Ann Thorac Surg 2015;100:309–11.
[9] Schroeder L, Zyblewski S, Forbus G, et al. Left ventricular myxoma. J Pediatr 2016;168:242–9.
[10] Yao F, Xu ZY, Liu YL, et al. Infective mitral valve myxoma with coronary artery embolization: surgical intervention followed by prolonged survival. J Thorac Cardiovasc Surg 2009;137:749–51.
[11] Thom CD, Sparks SE. Acute pediatric stroke-what's the hurry? A case for emergency physician-performed echocardiography. Am J Emerg Med 2014;32:1440–3.
[12] Xu J, Gao Y, Li Y, et al. Left atrial myoxma presenting as headache in the pediatric patient. J Emerg Med 2015;48:161–4.
[13] Kaneko T, Walkes JC, Tsai FW. Left atrial myxoma in a child with unique presentation: chest pain. Pediatr Cardiol 2011;32:230–1.
[14] Kumagai K, Sai S, Endo M, et al. Right ventricular myxoma obstructing the pulmonary artery during early childhood. Gen Thorac Cardiovasc Surg 2008;56:351–3.
[15] Cardoso R, Chaves M, Prado S, et al. Adolescent with neurological manifestations of cardiac cause treated surgically. Rev Port Cardiol 2009;28:97–103.
[16] Vega RA, Chan JL, Anene-Maidoh TI, et al. Mechanical thrombectomy for pediatric stroke arising from an atrial myxoma: case report. J Neurosurg Pediatr 2015;15:301–5.
[17] Sernich S, Chauhan A, Singh D, et al. Left atrial myxoma in a child: a challenging diagnosis of a rare lesion. World J Pediatr Congenit Heart Surg 2013;4:220–2.
[18] Takenouchi T, Sasaki A, Takahashi T. Multiple cerebral aneurysms after myxomatous stroke. Arch Dis Child 2014;99:849.
[19] Sawhney S, Agarwal M, Roy S, et al. A patient with rashes and limb weakness. Indian Pediatr 2009;46:867–73.
[20] Chlebowski M, O'Brien J, Hertzenberg C, et al. Asymptomatic Left Ventricular Myxoma in a 12-Year-Old Male. Tex Heart Inst J 2016;43:267–9.
[21] Ihsen Z, Hela M, Khadija M, et al. Cerebral embolism complicating left atrial myxoma: a case report. Pan Afr Med J 2016;24:140.
[22] Lee EJ, Moon S, Lee JY, et al. Concomitant percutaneous aortic embolectomy and excision of left atrial myxoma in a child. Pediatr Int 2015;57:172–4.