Stroke and peripheral embolisms in a pediatric patient with giant atrial myxoma
Case report and review of current literature
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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.1–3 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.

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 left lower extremity was level 4, while that of the left side was level 5. 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 × 60 × 40 mm) in the left atrium, which arose from the area of the fossa ovalis, was noted.
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.
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 yrs | M     | LA               | 15*14          | Heart murmur (grade 5, holosystolic) | None              | None                  | None              | Dead    | [6]       |
| 5 yrs | M     | LA               | None           | None             | None               | None                  | None              |         |           |
| 4 yrs | M     | LA               | 20*15          | None             | None               | None                  | None              |         |           |
| 6 mos | F     | LV               | None           | None             | Heart murmur       | None                  | None              |         |           |
| 12 yrs | M   | LV               | None           | None             | None               | None                  | None              |         |           |
| 12 yrs | F   | LV               | 35*25          | None             | None               | Fever as high as 39.5°C for 3 weeks | None              |         |           |
| 11 yrs | F   | LA               | 44*35*28       | Left-sided hemiparesis | None              | Rash on distal limbs, splinter hemorrhages | None              |         |           |
| 13 yrs | F   | LA               | 50*40*30       | Headache and dizziness for 18 months, loss of consciousness for the preceding 2 h | None | None | None | Normal | [13] |
| 16 yrs | F   | LA               | 25*25          | None             | None               | Chest pain, shortness of breath | None              |         |           |
| 2 yrs | M     | RV               | 30*15*15       | Heart murmur     | None               | None                  | None              | Normal  | [14] |
| 12 yrs | F   | RV               | 35*20          | None             | None               | None                  | None              | Normal  | [14] |
| 11 yrs | M   | LA               | 40*20          | Sudden dizziness and right-sided paresthesia of the lips and fingers | None | Multiple blanching lesions in feet | None | Normal | [14] |
| 8 yrs | M     | LA               | 55*24*40       | None             | Left-sided hemiparesis and dysarthria | weight loss, anemia | None | Normal | [15] |
| 14 yrs | F   | LA               | 40 of the longest diameter | None | None | None | None| Normal| [14] |
| 8 yrs | F     | LA               | 30*30          | None             | None               | None                  | None              | Normal  | [14] |
| 12 yrs | M   | LV               | 25*50          | Heart murmur     | None               | None                  | None              | Normal  | [14] |
| 11 yrs | M   | LA               | 50 of the longest diameter | None | None | None | None| Normal| [14] |
| 16 yrs | M   | LA               | 70*60*40       | None             | None               | None                  | None              | Normal  | Case 1 |

Table 1
Clinical summary of 19 pediatric cases of cardiac myxoma with different clinical manifestations.

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. 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.

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

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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.
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.

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Conceptualization: Xinmin Zhou.
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