

## Cardiac Tumor Presenting as Acute Right Hemiparesis in a 13-Year-Old Boy

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### INTRODUCTION

Cardiac myxoma is the most common cardiac tumor in adult group, but very rare in pediatric patient.<sup>1,2</sup> Among pediatric population, the most common primary cardiac tumor is rhabdomyomas, the second is fibroma, and myxoma is the third most common.<sup>1</sup> The clinical manifestation of cardiac myxoma depends on the tumor size, shape and surface contents.<sup>2</sup> The variable clinical symptoms can range from non-specific symptoms to some neurological deficits, or even cerebral stroke and peripheral embolisms due to thrombi from tumor fragments.<sup>1-4</sup> In literature review, most cardiac myxoma is located in left atrium and the size can range from small to as large as 8 cm.<sup>3-5</sup> Therefore, early diagnosis and management is important to save patient from morbidity, mortality and further complication.<sup>2-4</sup> We reported a 13-year-old boy with sudden onset of right hemiparesis. Further echocardiography revealed a huge mass in the left atrium. Cardiac myxoma was diagnosed after surgical removal of the mass.

### CASE

A 13-year-old boy was referred to our cardiology center due to right hemiparesis for one day. He was

healthy before without symptoms such as short of breath, tachypnea, chest tightness and palpitation, etc. The hemiparesis was of sudden onset, accompanied with numbness and unsteady gait of right limbs. He was brought to a local hospital initially, where brain computed tomography (CT) was done without specific finding. Right hemiparesis persisted, so he was brought to another hospital for further evaluation. Young stroke survey was done completely. Brain magnetic resonance imaging (MRI) showed high signal intensity on T2WI and T2 FLAIR with restricted diffusion over left thalamus (Figure 1), suggestive of acute infarction. Notably, transthoracic echocardiography revealed a huge intracardiac tumor, attached to mitral valve inside left atrium. Under the diagnosis of acute left thalamus infarction and intracardiac tumor, he was then transferred to our hospital on the third day of symptom onset for further survey and management.

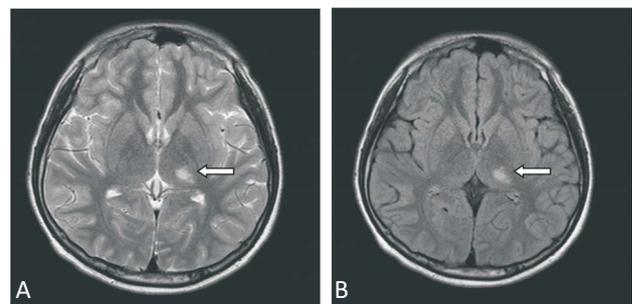
On physical examination, he had weakness of right extremities along with right paresthesia. Deep tendon reflex in lower extremities was two plus and the Babinski sign was negative. Heart auscultation revealed regular heart beats without audible murmur. Transthoracic echocardiography revealed a non-mobile 3.8 × 2.6 cm solitary mass within left atrium (Figure 2A), suspicious cardiac myxoma, with preserved mitral valve contour and left ventricle function (Figure 2B). Cardiac CT

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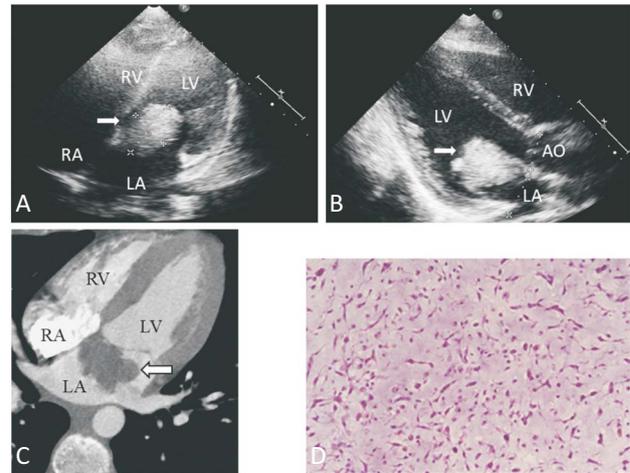
**Figure 1.** (A) T2W. (B) T2W Flair image showed high signal intensity within left thalamus suggesting left thalamus infarction.

also showed cardiac mass compatible with suspicious cardiac myxoma in left atrium (Figure 2C). Further work-up for stroke survey was performed, including vascular, inflammatory and metabolic origin, without specific abnormal finding. After supportive treatment, his condition was stable. Surgical resection of cardiac tumor with atrial septum repair was performed successfully on the 12th day. Pathological report showed cardiac myxoma (Figure 2D). Follow-up echo showed no visible residual tumor. There was no significant neurological sequelae except mild numbness of right limb after operation. He was thus discharged uneventfully and followed up at our outpatient department.

## DISCUSSION

Cardiac myxoma is very rare in children, and accounts for about 2-4% of primary benign tumor.<sup>6</sup> The low incidence of cardiac myxoma and the non-specific clinical presentations can lead to misdiagnosis and neglect until transthoracic echocardiogram is performed. Despite benign nature of cardiac myxomas, the symptoms such as embolism can be serious.<sup>3,4</sup> It is important to recognize that cardiac myxoma can initially be seen as stroke, and echocardiography can easily be done to document the diagnosis.

The clinical symptoms of cardiac myxoma vary from being asymptomatic to sudden death.<sup>2-5</sup> Nonspecific symptoms include dyspnea, palpitation, fatigue, chest pain, and possible fever related to tumor secretion of interleukin-6.<sup>3,5</sup> Potential lethal symptoms can be caused by intra-cardiac obstruction of valve and outflow tract by tumor, arrhythmia, heart failure, and embolic complications by fragmentation of tumor cells and adherent surface thrombus.<sup>2-5</sup> Stroke is the first clinical manifestation in one third of cases<sup>4</sup>, as demonstrated in our case. Most embolic events arising from cardiac myxoma involve the cerebral arteries, but embolization of coronary, renal, and femoral arteries, etc may rarely occur.<sup>7</sup> The middle cerebral artery and cerebellum were the predominant locations for myxoma-related embolic stroke in the previous report.<sup>7</sup> Our case with a single thalamus infarction is uncommon and requires cautious differential diagnosis in identifying possible etiologies other than myxoma. The mechanism of stroke due to myxoma



**Figure 2.** (A) Two-dimensional transthoracic echocardiogram, four-chamber view showed a non-mobile 3.8 × 2.6 cm solitary mass within left atrium. (B) Long-axis view showed a mass with irregular borders and homogeneous echogenicity. (C) Computed tomography of the cardiac showed a filling defect, an occupying lesion suggesting LA tumor. (D) Pathological diagnosis: the cardiac tumor composed of myxoma cells existing as cords and nests in a myxoid background. (hematoxylin and eosin staining, X200). AO, aorta; LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

is still unclear. Cerebral aneurysm due to tumor cell penetration, irregular papillary myxoma with a soft mobile surface and friable structure, tumor size, atypical tumor location, and high platelet count were suggested to be risk factors of embolic stroke.<sup>7,8</sup> Further investigation with large sample size is necessary to elucidate the mechanism of embolic stroke in patients with cardiac myxoma. The prompt definite diagnosis can lead the clinical physician to make right decision for early surgical intervention.<sup>9</sup> Stroke survey, including hematologic, vascular, and metabolic factors is important to exclude other etiologies before operation.<sup>9</sup> Besides, before surgical intervention, cardiac CT or MRI is recommended for definite diagnosis and providing the surgeon adequate information.

The surgical outcome of cardiac myxoma is generally good,<sup>5</sup> as demonstrated in our case. However, late onset atrial fibrillation is not rare in patients after resection of atrial myxoma.<sup>5</sup> There is also a potential recurrence risk of 1-3% in sporadic cases and 20% in familial cases.<sup>10</sup> Regular postoperative examinations, including echocardiography and electrocardiography, are suggested for patients with resected atrial myxoma.

Cardiac myxoma frequently presents with symptoms

of embolism, but with little cardiac symptoms. Pediatric patients with unknown hemiparesis or ischemic stroke should immediately undergo echocardiography to exclude cardiac etiologies.

### LEARNING POINTS

1. Cardiac myxoma is very rare in pediatric patient. The most common original site of cardiac myxoma is from left atrium, and the variable manifestations depend on the tumor size, location, shape or tumor contents.
2. Cardiac myxoma frequently presents with symptoms of embolism, but with little cardiac symptoms. Pediatric patients with unknown hemiparesis or ischemic stroke should immediately undergo echocardiography to exclude cardiac etiologies.
3. The surgical outcome of cardiac myxoma is generally good. However, late onset atrial fibrillation is not rare in patients with resection of atrial myxoma, and there is also a potential recurrence risk. Regular postoperative examinations, including echocardiography and EKG, are suggested for patients with resected atrial myxoma.

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### CONFLICT OF INTEREST

All the authors declare no conflict of interest.

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