Recurrent Facial Palsy as an Initial Presentation of Cardiac Myxoma

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Facial palsy in childhood may result from a variety of diseases. However, it rarely presents initially as cardiac myxoma. In this article, we report on a 12-year-old boy who suffered from recurrent facial palsy as the initial clinical manifestation, which resolved spontaneously within several days of hospital admission. The predisposing factor of the symptom was physical exercise. Brain magnetic resonance imaging showed encephalomalacia and multiple cystic changes compatible with an old infarction over the left caudate nucleus, head-body junction and the left putamen. Echocardiography revealed a tumor mass on the left atrium, and surgical intervention was performed. The pathological finding revealed cardiac myxoma, and a diagnosis of intracardiac myxoma associated with brain infarction and recurrent facial palsies was made. During the three year follow-up, no residual cardiac myxoma or recurrent facial palsy were noted. Although cardiac myxoma associated with stroke rarely occurs in children, it should be considered as a differential diagnosis in patients with paroxysmal neurologic disorders.

Key Words: Atrial myxoma • Cardiac myxoma • Facial palsy • Facial paralysis • Stroke

INTRODUCTION

Acquired facial palsy is a common problem encountered in pediatric outpatient and emergency departments. Most cases are related to viral infection which resolve spontaneously and the prognosis is usually favorable.[1]

Nevertheless, facial palsy can be the initial presentation of several underlying diseases which may lead to serious morbidity and mortality. Its etiology includes central nervous system infection, head trauma, brain tumor, leukemia, coagulation disorders, vasculitis, or congenital malformations.[1] Other unusual causes include hypertensive encephalopathy, Kawasaki disease, Guillain-Barré syndrome, Lyme disease, and neurofibromatosis.[1] Facial palsy had been reported as a symptom of cardiac myxoma, and patients in most of those cases suffered from brain infarction with subsequent neurological sequelae.[2,3]

We herein report a 12-year-old boy with cardiac myxoma presenting as recurrent facial palsies which resolved spontaneously. This is a rare case of cardiac myxoma-associated brain infarction with such subtle neurological symptoms.

CASE REPORT

The 12-year-old boy was born after an uncomplicated pregnancy and delivered at term as the first child of healthy, nonconsanguineous parents. The family history was unremarkable for cardiac or central nervous system diseases.

He suffered his first episode of sudden onset of right hand weakness, asymmetric facial expression and

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dyspnea during exercise. The boy was brought to a clinic and brain computed tomography showed no evidence of brain tumor or intracranial hemorrhage. His symptoms resolved ten days later and he was discharged with a diagnosis of hyperventilation syndrome.

Three months later, the second attack presented with left upper limb weakness, slurred speech, dyspnea and mouth deviation to the right side during exercise, he again was brought to our emergency department. Upon admission, his vital signs were heart rate 82/minute, respiratory rate 19/minute, and blood pressure 120/70 mmHg. Physical examination of the patient revealed a clear breathing sound with smooth breathing pattern, no heart murmur, and no remarkable skin lesions. Routine neurological examination disclosed clear consciousness, unequal pupil sizes (left/right 3 mm/3.5 mm) with light reflexes, normal external ocular motion, bilateral forehead wrinkles with absence of left nasolabial fold, and mouth angle deviation to the right side. Increased deep tendon reflexes over bilateral lower limbs without ankle clonus and barbinski sign were also found, and decreased muscle power (2+~3+) was found over the left upper limb.

Laboratory data revealed a white blood cell count 12300/mm^3 (normal 4500-13,000), and a C reactive protein level of 1.9 mg/dl (normal < 0.3). Biochemistry parameters showed sodium 140 mEq/L (normal 137~153), potassium 3.9 mEq/L (normal 3.5~5.3), calcium 10 mg/dL (normal 8.4~10.2), and blood glucose 105 mg/dl (normal 70~110). Immunologic survey, including C3, C4, anti-double strand DNA, and anti-nuclear antibody, showed negative results.

There was no obvious abnormality on the patient's chest film. Brain magnetic resonance imaging (MRI) was arranged due to left limb weakness and left central facial palsy, which revealed cystic encephalomalacia over the left caudate nucleus, head-body junction and left putamen (Figure 1). Magnetic resonance angiography was unremarkable. His electrocardiogram showed normal sinus rhythm and the QT interval was within normal limits. However, cardiac sonography revealed a large mass over the mitral valve, measuring 31 × 29 mm in size (Figure 2A). Multiple detector computed tomography (CT) of the cardiovascular system showed a polypoid mass measuring 41.2 mm over the left atrium with a peduncle of 12 mm based on atrial septum (Figure 2B).

The patient's symptoms spontaneously subsided one day after admission. Excision of the intracardiac tumor was performed and a myxomatous mass measuring 3 cm in the left atrium with stalk attached to the atrial septum was noted grossly. Further pathologic analysis showed a loose myxoid stroma with round and polygonal cells

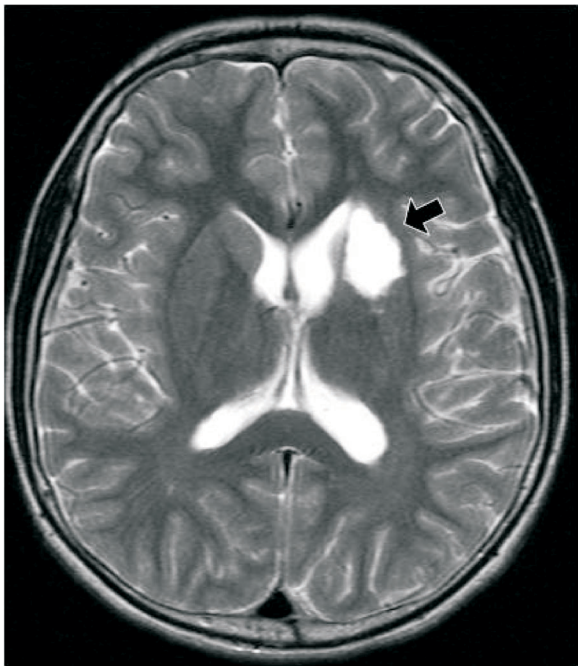

Figure 1. *Brain magnetic resonance imaging of the case. Axial view (TE/TR 97 ms/4500 ms) showed cystic encephalomalacia (arrow) over the left caudate nucleus, head-body junction and left putamen.*

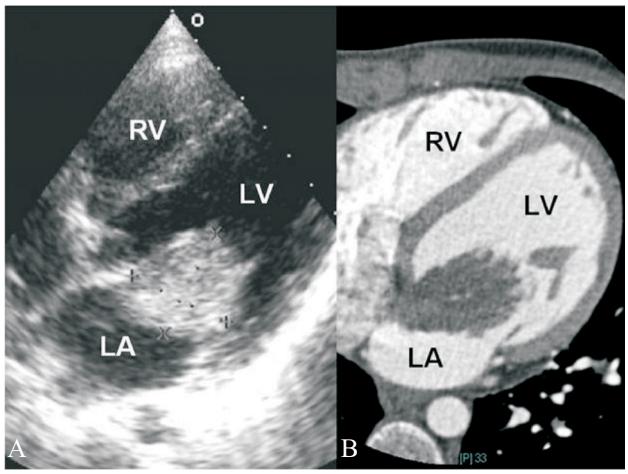

Figure 2. *Echocardiography of the case. (A) Parasternal long axis view showed a mass measuring 3.09 cm × 2.91 cm over left atrium. (B) Cardiac multiple detector computed tomography showed a 41.2 mm polypoid tumor with a pedicle of 12 mm based on atrial septum. LA, left atrium; LV, left ventricle; RV, right ventricle.*

forming solid cords and nests. During a period of three year follow-up, no additional facial palsies or other neurologic symptoms were noted, and subsequent echocardiography showed no residual tumor.

DISCUSSION

Cardiac diseases have been shown to be etiological factors associated with stroke events.[2,4,5] They include arrhythmias, infective endocarditis, rheumatic fever, cardiomyopathy, mitral valve prolapse, prosthetic valves, aneurysm, and valvular calcification.[2,4] When a stroke is the result of heart disease, the initial physical examination may not reveal any clue about the underlying heart disorder. It has been reported in the literature that 64% of patients with cardiac myxoma had an abnormal heart murmur, and 15% might have discerned a tumor polyp on cardiac auscultation.[6] In our case, no heart murmur was found. In general, underlying cardiac abnormalities may be discovered when serious neurological events are encountered. However, neuroimaging studies might also show shortcomings in the detection of early brain ischemia. CT and MRI sensitivity for identifying early stage brain lesions had been reported to be 26% and 83%, respectively.[7] The brain MRI performed in our case did not reveal any new lesions in the patient's second attack.

Cardiac neoplasm has been reported as a cause of brain infarction in some cases. The most common cardiac tumor in childhood is rhabdomyoma. Cardiac myxoma is less common in children than in adults,[8] and account for 10~15% of pediatric cardiac tumors.[4] Approximately 75% of cardiac myxomas were found over the left atrium.[8] Some cases may be the familial type, which is inherited in an autosomal dominant manner.[4,8-10] Furthermore, the familial form of myxoma may be associated with other organ involvement, such as skin lesions, neoplasm, or endocrinopathy.[9] Generally, the location and size of myxoma determine the clinical manifestations. The fragments of the atrial tumor are carried with the blood flow to all parts of the body, and may produce symptoms if they become lodged and subsequently form a stenotic lesion.[4] Dyspnea on exertion, pulmonary edema, orthopnea and/or embolic events are found in the left atrial myxoma, while the right cardiac tumor tends to cause syncope, right heart failure, pulmonary embolism, pulmonary hypertension, and/or atrial fibrillation.[10]

Most cardiac myxoma associated with stroke reportedly resulted in severe and permanent neurologic sequelae, although this differs from those symptoms seen in our case.[3,4] Our patient suffered from transient limb weakness, facial paralysis, and dyspnea, and these symptoms recovered spontaneously without severe neurological sequelae. A possible explanation for this phenomenon was the increased cardiac load during exercise, which may have caused fragmentation of the cardiac myxoma or the thrombus around the tumor, thus allowing for greater blood flow.[4] Early detection of cardiac myxoma is important as early surgical intervention may reduce the risk of further complications.[10] Delayed surgery increases the risk of recurrent cerebral emboli, and cardiac myxoma has been reported as a source of bacterial and fungal infections leading to endocarditis and septic emboli.[4,10]

In conclusion, cardiac myxoma resulting in brain infarction may present as a neurological symptom, such as transient facial palsy, and is easily misdiagnosed.[3] In children with paroxysmal neurological symptoms, an underlying cardiac problem should be included in the differential diagnoses.[4]

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