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Report

Cardiac Papillary Fibroelastoma in a 20-month-old Child: A Case Report and Literature Review

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Cardiac papillary fibroelastoma (PFE), a rare primary benign tumor in children, is a potential cause of stroke, systemic embolism, myocardial infarction, and sudden death. Herein, we report a 20-month-old girl with PFE showing acute onset of hemiplegia. A comprehensive review of the literature is also presented.

Key Words: cardiac papillary fibroelastoma, stroke, hemiplegia, children

Introduction

Cardiac papillary fibroelastoma (PFE) is an uncommon primary tumor of the heart. It can be asymptomatic but may also cause severe complications via cerebral or coronary embolization. PFE is rare in children. We report a 20-month-old girl with PFE whose initial manifestion was a stroke.

Case Report

A 20-month-old girl was admitted to Shanghai Children's Medical Center with a four-day history of right extremity weakness. Four days before admission, she had fallen from a chair 50 cm in height and was found to have right hemiplegia. There had been no fever, vomiting, seizures, or changes in consciousness. Her birth and development had been normal. There was no family history of strokes or any other serious neurological events.

Physical examination findings were as follows: Vital signs were stable. The pupils were equal, round, and responsive to light. The right nasolabial fold was flattened relative to that on the left. The muscle strength of the right lower extremity was decreased (grade 3/5). Hyperactive reflexes on the right with a right-sided Babinski sign and ankle clonus were elicited. In addition, a grade 2/6 systolic ejection heart murmur over the precordium was detected by careful auscultation.

The complete blood count including platelets was normal, as were blood biochemistry parameters and cerebrospinal fluid analysis results.

Computed tomography of the brain showed large areas with low-density lesions in the right temporal, occipital, and left parietal lobes. Cranial magnetic resonance imaging (MRI) revealed abnormal signals in the frontal, temporal, occipital and parietal lobes, as well as in the basal ganglia bilaterally, but these lesions were not symmetrical (Fig. 1a). Cranial magnetic resonance angiography (MRA) showed occlusion in the right internal carotid artery (Fig. 1b and Fig. 1c). An abnormal signal at the left ventricle was detected on cardiac MRA (Fig. 1d), and vegetations involving the left ventricle and mitral valve were further verified with echocardiography (Fig. 2). Based on these findings, the patient was diagnosed as having had a stroke accompanied by occlusion of the right internal carotid artery. The embolus was thought to have originated from the cardiac vegetations. Mitral valve replacement and excrescence resection were performed soon after the diagnosis had been confirmed. PFE was confirmed histopathologically after surgical resection (Fig. 3).

Discussion

Stroke is a neurological injury and the reported incidence of pediatric stroke ranges from 1.2 to 13 cases per 100,000 children under 18 years of age[1]. Cardiac diseases are the most common causes of

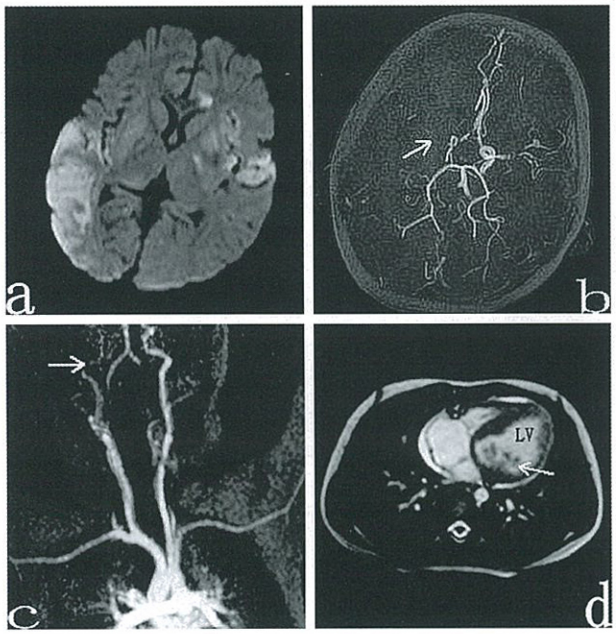

Fig. 1 a: Abnormal signals in the temporal, occipital, and parietal lobes, and in the basal ganglia (MRI). b: Occlusion in the right middle cerebral artery (MRA). c: Occlusion in the right internal carotid artery (MRA). d: Abnormal signal in the left ventricle (Heart MRA).

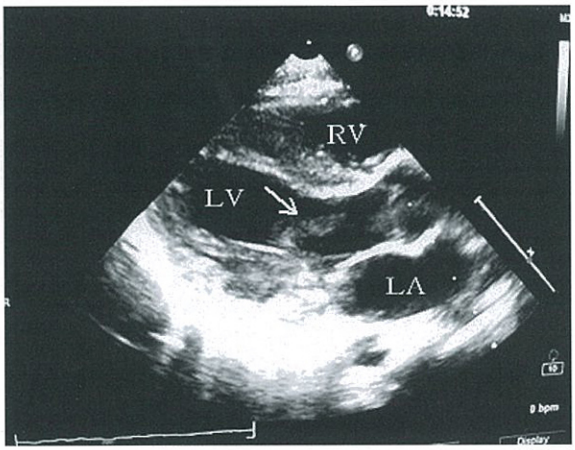

Fig. 2 Large vegetations in the left ventricle (Echocardiography)

ischemic infarction in children[2], accounting for up to one third of all acute ischemic strokes.

Primary cardiac tumors, originating from the myocardium or pericardium, are uncommon in children, with an incidence of 0.0017 to 0.28% at autopsy[3]. The incidence has, however, risen to 0.019% since the advent of identification by echocardiography. PFE, first recognized in 1961 by Pomerance[4], is among the benign, slow-growing, rare tumors of the heart. Though it is the third most common primary benign tumor of the heart after myxoma and lipoma, it accounts for only 7% of total cases. The etiology of PFE development is still not clear, but possible explanations have been suggested, such as previous mechanical damage to the endothelium, hamartomatous origins, or an organizing embolus. There is a growing body of evidence showing that iatrogenic PFE may be relatively common among patients who have undergone previous cardiac surgery[5].

Morphologically, PFE resembles a "sea anemone" with multiple papillary fronds attached to the endocardium by a short stalk. Microscopically, the papillae are avascular structures, containing a core of dense collagen fibers admixed with varying amounts of reticulin and elastin fibers. The most common clinical manifestations of PFE are stroke and myocardial infarction[6]. The initial clinical presentation can be neurological. Hoashi et al[7] reported a 5-year-old boy who presented with acute onset of chorea and dysarthria. Karapanagiotidis et al[8] described an 81-year-old woman whose PFE manifested with recurrent episodes of dizziness. PFE represents a potential cause of systemic embolism, stroke, myocardial infarction and sudden death. Early diagnosis is very important, as surgical excision of these tumors can prevent cerebrovascular and cardiovascular complications. Only one case, through 2012, was reported to have developed a recurrence after initial surgical intervention[9].

Because our case had a history of a fall, head trauma could easily have confused the diagnosis. The patient manifested with right hemiplegia, but radiological tests showed lesions on both sides of her brain. This also suggested that the source of the lesions was extracranial disease. This case highlights the importance of a thorough clinical assessment (history, general examination, and neurological examination), which remains the cornerstone of obtaining a correct diagnosis.

According to the American Heart Association guidelines for the early management of adults with ischemic stroke[10], one way to avoid delays or misdi-

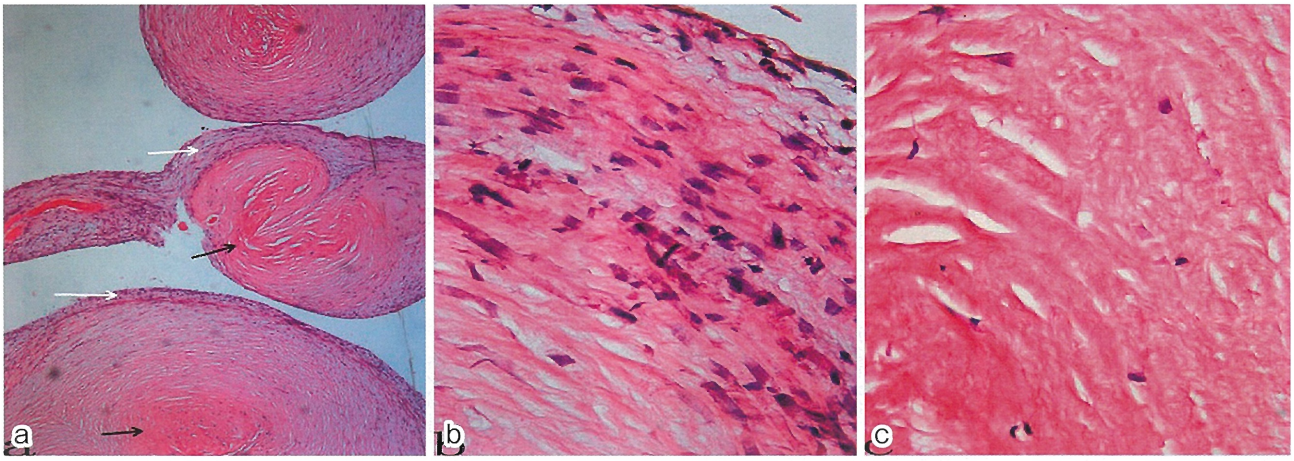

Fig. 3 a: Photo of a light micrographic specimen shows papillary branches containing endothelial tissue surrounding a hyalinized stromal core (H & E-stained, ×10). b: Endothelial tissue corresponding to the area in Fig. 3a (×40) is indicated by the white arrows. c: A vascular hyalinized stromal core in the area in Fig. 3a is indicated by the black arrows (×40).

agnoses in children would be identifying risk factors and causes of stroke in this age group. Since cardiac anomalies are significant risk factors, ECG and chest radiography are necessary, and echocardiography may also be very useful.

Conclusion

When cerebral infarction is diagnosed, physicians should be aware that extracranial diseases are potential causes.

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心乳頭筋線維弾性腫の20ヵ月女児例―症例報告と文献展望

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心乳頭筋線維弾性腫（Cardiac papillary fibroelastoma（以下PFE））は稀な小児期の原発性良性腫瘍である．本症は，小児における脳卒中，塞栓症，心筋梗塞，および突然死を惹起する可能性がある．我々は，PFEにより，急性の片麻痺を呈した20ヵ月女児を経験したので，文献展望とともに報告した．