Rare Etiology of Acute Embolic Stroke in a 17-Year-Old Girl: Cardiac Papillary Fibroelastoma

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Abstract

A cardiac papillary fibroelastoma is a benign, pedunculated tumor with strand-like projections arising from the endocardium. While often discovered incidentally, these tumors can embolize resulting in major neurologic events. There is a dearth of pediatric literature describing the necessity and optimal timing of surgical intervention for this rare primary cardiac tumor after a cerebrovascular event. We report an interesting case and the subsequent management dilemma in a teenage patient presenting with acute embolic strokes secondary to an aortic valve papillary fibroelastoma. Despite its rarity, a high index of suspicion should be maintained for such tumors in previously healthy children presenting with acute stroke.

Keywords

- ▶ pediatric
- ▶ stroke
- ▶ cardiac tumor

Introduction

The incidence of primary cardiac tumors is 0.002 to 0.01% in multiple large autopsy series.^{1–3} Cardiac papillary fibroelastomas (CPFs) comprise less than 10% of all cardiac tumors and their prevalence is highest in the eighth decade of life.⁴ They are found predominantly on the aortic and mitral valves and are the most common benign tumor of cardiac valves with the propensity to embolize resulting in stroke.^{1–3} Although primary cardiac tumors in children are rare with only sporadic cases reported, here we report the case of a teenager with acute embolic strokes due to a CPF that was subsequently successfully resected.

Case

A 17-year-old previously healthy girl presented to an outside hospital emergency department with right-sided hemiplegia, right-sided facial droop, and dysarthria approximately 6 hours after the onset of symptoms. She suffered a second episode after admission to the outside hospital (~8 hours after the first episode) characterized by mild loss of left-sided strength with worsening dysarthria and visual field

defects. Magnetic resonance imaging of the brain at this time revealed acute infarcts of the left caudate, putamen, and posterior limb of the internal capsule and right insula (▶Fig. 1). Aspirin therapy was initiated and a transthoracic echocardiogram demonstrated a 1 cm × 1 cm, mobile mass on the aortic valve (▶Fig. 2). The patient was then transferred to our institution's pediatric cardiac intensive care unit for further management, approximately 18 hours after the initial symptoms. As the etiology of the mass (thrombus vs. vegetation) was unclear, the patient was systemically anticoagulated with heparin (continued until the time of surgery), started on broad-spectrum antibiotics and a hypercoagulability workup was pursued. Serial echocardiography revealed no improvement in the size of the mass. Her hypercoagulable workup was negative and the neurologic deficits were slowly improving. Given the size and location of the mass and clinical evidence of recurrent embolic events, a consensus was reached by the multidisciplinary team caring for this patient to proceed with surgical resection on day 7 of admission to our institution.

The patient underwent surgical repair involving cardiopulmonary bypass (CPB) with systemic heparinization with 400 units/kg of heparin. A transverse aortotomy was made in

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Fig. 1 Initial magnetic resonance imaging: bilateral areas of acute infarct (white arrows).

the proximal ascending aorta exposing the aortic valve. On examination, both right and left coronary cusps were normal in appearance. There was a tumor (► **Fig. 3**) on the ventricular surface of the noncoronary cusp, which was shaved off without injury to the leaflet. The remainder of the operation and post-operative course was unremarkable and the patient was discharged on daily aspirin. She continued to improve clinically and on last visit (postoperative day 140), her exam revealed mild residual right hand weakness, but no other deficits. The final pathology demonstrated a villiform mass with histologic features consistent with papillary fibroelastoma.

Discussion

CPFs are typically mobile, less than 1.5 cm and have an 80% predilection for the left heart valves.¹⁻³ While often asymptomatic, presenting findings may include transient

Fig. 3 Intraoperative images of aortic valve mass (black arrow).

ischemic attack, stroke, angina, myocardial infarction, and pulmonary embolus. CPF is seen as a small, well-delineated stalk-like mass on transthoracic echocardiography, although transesophageal echocardiography is more sensitive.³ Differential diagnosis of CPF comprises vegetation, thrombus, other primary cardiac tumors, and Lambl's excrescence.

In our literature review, we found three adult studies that recommend surgical intervention for CPF based on patient symptomatology, size of the tumor, location, and mobility. Gowda et al concluded that both symptomatic and asymptomatic patients with CPF and evidence of tumor mobility should undergo surgical resection via shave excision.¹ Boodhwani et al developed an algorithm recommending that symptomatic as well as asymptomatic CPF that are more than 1 cm, mobile, or left sided should be surgically removed.⁵ Sun et al concluded that the decision to excise rested on the presence of a stalk and mobility. This group recommends observation of asymptomatic patients with CPF that are less than 1 cm in size and nonmobile.³

There is no adult or pediatric literature regarding timing of surgical intervention in those symptomatic patients who

Fig. 2 Transthoracic echocardiographic images of aortic valve mass. Panel A: Parasternal long-axis view (white arrow). Panel B: Parasternal short-axis view with real-time three-dimensional echocardiography visualization (black arrow).

have suffered a neurologic event secondary to a CPF. In reviewing the literature describing timing of surgical intervention in adult acute infective endocarditis patients with cerebrovascular complications, we found recommendations ranging from prompt surgery to delaying for up to 4 weeks. Eishi et al found that cerebral injury was exacerbated in 43.8% of patients with cerebral infarct who underwent cardiac surgery within 7 days with this rate falling to 10% at 2 weeks and 2.3% at 4 weeks.⁶ Gillinov et al reported that the mean interval of time from onset of embolic cerebrovascular accident without hemorrhage to surgery was 22.1 ± 3 days and 96% of patients survived surgery and were discharged home with either improved neurologic function or no change in neurologic exam.⁷ Piper et al found that the prognosis of patients undergoing cardiac surgery within 72 hours was more favorable than those treated medically when the risk of hemorrhagic conversion is low. In addition, they concluded that delaying surgery by more than 8 days was associated with a poor prognosis.⁸

In our patient, surgical resection was warranted given the recurrent neurologic events, size, location, and mobility of the lesion. We weighed the risk of hemorrhagic conversion of the acute cerebral infarcts against the benefit of preventing further embolic sequelae in this teenager and pursued surgical intervention. Although we may never formally answer the question of the optimal timing of CPB in the setting of a stroke or an evolving neurological event, each care team must critically weigh various clinical characteristics to best risk stratify each patient and their possible complications/postoperative course. Most importantly, a high index of suspicion for primary cardiac tumor is neces-

sary for optimal diagnosis and presurgical planning in pediatric patients presenting with acute embolic stroke.

Conflict of Interest

None.

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