Cardiac papillary fibroelastoma of a bicuspid aortic valve in an adolescent: A case report

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Abstract

Cardiac papillary fibroelastomas (CPFE) are exceptional primary benign cardiac tumours affecting the heart valves. We report here the case of a 15-year-old boy in whom echocardiography performed for non-specific chest pain during follow-up for bicuspid aortic valve showed as accidental finding the presence of a round mobile mass without stalk attached on the inferior side of the aortic valve. The mass did not cause any outflow tract obstruction or aortic insufficiency. Electrocardiogram-gated cardiac computed tomography and magnetic resonance imaging allowed to suspect CPFE. Although the patient was asymptomatic, open cardiac surgery with elective surgical resection of the tumour was performed to avoid systematic emboli. Histology confirmed the diagnosis of CPFE. This is an exceptional case of acquired CPFE in a young patient with bicuspid aortic valve. Due to the risk of systemic embolization, aortic or coronary ostium obstruction, elective excision of such lesions is recommended.

Introduction

Cardiac papillary fibroelastomas (CPFE) are exceptional primary cardiac tumours with a reported incidence lower than 0.03% in adults.¹¹² They arise from the cardiac endothelium and principally affect the heart valves.¹ Endothelial damage, iatrogenic factors, or organizing thrombi are probably involved in its pathogenesis. CPFE are benign tumours with high potential for acute life-threatening complications such as sudden cardiac death, stroke, and myocardial infarction in until then asymptomatic patients.² Due to the paucity of symptoms, CPFE are often accidentally discovered at autopsy, during heart surgery or at cardiac imaging. In general, the lesion is well depicted at transthoracic and/or transoesophageal echocardiography.³,⁶ Cardiovascular magnetic resonance imaging and/or Electrocardiogram-gated cardiac computed tomography (ECG-gated TDM) may be helpful to precise the diagnosis of CPFE.⁶,⁷ Owing to the high risk of embolization in the pediatric population, prophylactic tumour excision is considered to be the definitive and safe treatment of choice.¹,³

We report the case of a 15-year-old boy with CPFE on a bicuspid aortic valve, accidentally discovered by echocardiography at follow-up.

Case Report

A 15-year-old boy was referred to our department for further evaluation of a polypoid formation accidentally discovered during a routine transthoracic echocardiography (TTE) conducted for chest pain. A former TTE performed 4 years earlier in the context of palpitations had showed a thin bicuspid aortic valve without any stenosis or insufficiency.

Physical examination was normal but cardiac auscultation revealed a protosystolic click at the level of the second and third intercostal space of the left sternal border, followed by a 1-2/6 systolic murmur with a punctum maximum in the second right intercostal space and with irradiation to the carotid arteries.

ECG was normal. TTE confirmed the bicuspid aortic valve. A round mobile mass, approximately 8 mm in diameter, was attached to the ventricular side of the aortic valve, projecting at the free edge of the two leaflets. No stalk could be identified (Figure 1). The mass did not cause any outflow tract obstruction or aortic insufficiency. Systolic flow velocity was slightly increased. Cardiac function was normal. An exercise stress test on bicycle was negative. An ECG-gated cardiac TDM confirmed the presence of a bicuspid aortic valve. Just below the raphe connecting the non-coronary and right coronary cusp, a nodular lesion of 6×7 mm was noted evoking a CPFE. There was no calcification (Figure 2A). Cardiac structure and function were otherwise normal. Cardiac magnetic resonance imaging (MRI) performed during preoperative assessment confirmed the diagnosis of CPFE (Figure 2B).

Decision was made to perform prophylactic excision of the CPFE under cardio-pulmonary bypass. The mass immersed in water, CPFE resemble typically a sea anemone.⁸ On histologic examination, CPFE present with a central core of dense hyalinised connective tissue. Surrounding this central core are multiple papillary fronds made of a single layer of hyperplas-
tic endocardial cells, elastic fibers and loose connective tissue. Most CPFE are asymptomatic and usually, they are incidentally diagnosed at autopsy, heart surgery or cardiac imaging with a mean age at diagnosis of 60 years.1-4,6,7,12

Clinical presentation is determined by tumour localisation, size and tendency for embolization that is higher for left-sided than for right-sided lesions1-3,7 and manifests by systemic ischemia. The embolic debris originate either from the tumour itself, or from thrombi that form at the tumour surface.1,13 Besides embolic complications, mechanic interferences impairing valve function or obstructing a coronary ostium and conduction system disturbances have also been described.6,7,14

In children, TTE is the most reliable diagnostic tool that may be implemented by transesophageal echocardiography in cases of sub-optimal window. The differential diagnosis of CPFE includes other heart tumours, Lambli’s excrescences, intracardiac thrombi, vegetations and valvular calcifications.1,6,7

In adults, CPFE are described as solitary, round or oval, or irregular-shaped tumours, generally small, with a mean size of 10 mm, half of them have stalks and are often mobile.3,8 In some particular cases, magnetic MRI is useful to assist in the diagnosis of CPFE. MRI allows a high soft-tissue characterization, multi-planar imaging, and high temporal resolution. Along with echocardiography, MRI is the modality of choice for the diagnosis of CPFE in children because it doesn’t require the use of ionizing radiation in contrast to CT imaging. Furthermore, cine-MRI allows an assessment of myocardial and valvular function comparable with echocardiography. However, cardiac MRI is dependent on patient cooperation. The evaluation of small mobile masses, such as CPFE or valvular vegetations, may also be limited due to poor spatial resolution. In addition, MRI does not provide detailed assessment of the coronary arteries, in contrast to angio-computed tomography.6,7,15

In the presented case, ECG-gated cardiac CT was performed to exclude mass calcification and to better evaluate the potential for coronary ostium obstruction by the tumour.

Routine preoperative coronary angiography is not recommended due to the friable nature of the lesion and to the potential risk of embolization.2,12

Surgical excision is curative and is indicated in symptomatic CPFE.1,3,7

Since young age is not protective11,14 and because CPFE size does not correlate with the development of serious embolic complications,2,3 surgery is always indicated in asymptomatic children. Surgical removal of CPFE is simple, safe, and highly efficacious. In most cases, the tumour is pedunculated and can be easily removed with low perioperative morbidity and mortality.2,3,7 Should valvular damage occur during resection of the tumour, standard techniques of valve repair are effective.13

In adult patients, minimal invasive sur-
surgery via mini sternotomy has been shown effective and safe for the excision of CPFE.\textsuperscript{16} The technique can be performed under robotic-assisted endoscopy as recently reported in adults presenting CPFE located in the mitral chordae or in the right ventricle, respectively.\textsuperscript{17,18} In elective cases, percutaneous resection of CPFE in adults has also proven feasible and safe.\textsuperscript{19} However, in children, conventional cardiac surgery remains at time the gold standard.\textsuperscript{11}

Fortunately, recurrence after surgery has not been reported but careful follow-up is necessary.\textsuperscript{6-8,13}

Conclusions

Among benign cardiac tumours, CPFE are extremely rare in children. Although most CPFE do not cause symptoms, there is sufficient evidence to state that these tumours can cause life-threatening complications. Therefore, the diagnosis of CPFE in infants and children is of vital importance due to the high risk for embolization. Accordingly, prophylactic tumour excision with valve repair or replacement when necessary is considered to be the treatment of choice, even in asymptomatic patients.

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