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.[1-5] 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 transesophageal echocardiography.[3,6] Cardiac 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 saline floated and opened like a sea-anemone, a typical finding for fibroelastoma. The anatomopathological analysis confirmed the nature of the lesion (Figure 3).

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Key words: Cardiac papillary fibroelastoma; Bicuspid aortic valve; Children.

Contributions: SD, conception of the research work, acquisition and collection of the data, analysis and critical interpretation of the data; drafting the manuscript, analyzing literature and critically revising the manuscript; final approval of the manuscript; agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. MCS wrote the manuscript; BD and MCS diagnosed the lesion at echocardiography; MB confirmed the anatomopathological nature of the lesion. All of the authors contributed to the revision of the manuscript.

Conflict of interest: the authors declare no potential conflict of interest.

Funding: none.

Received for publication: 7 February 2019.
Revision received: 6 July 2019.
Accepted for publication: 2 August 2019.

Clinics and Practice 2019; 9:1135
doi:10.4081/cp.2019.1135

Discussion

CPFE are small solitary avascular tumours arising most frequently from the valvular endocardium,[1,4,6-8] involving particularly the aortic valve, as it was the case in our patient.[7-10] CPFE are generally acquired lesions secondary to endothelial damage.[2,3,7] In our patient, CPFE developed most probably in response to non-laminar flow pattern related to the bicuspid aortic valve. Beside acquired forms, congenital forms of CPFE have been described in neonates and infants, suggesting a hamartous origin of the tumour.[3,7,11]

Macroscopically, when immersed in water, CPFE resemble typically a sea anemone.[1,3,7,9] 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 lesions[1-3,7] 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, Lambl'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.[2,3,7]

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

Since young age is not protective[3,11,14] 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-

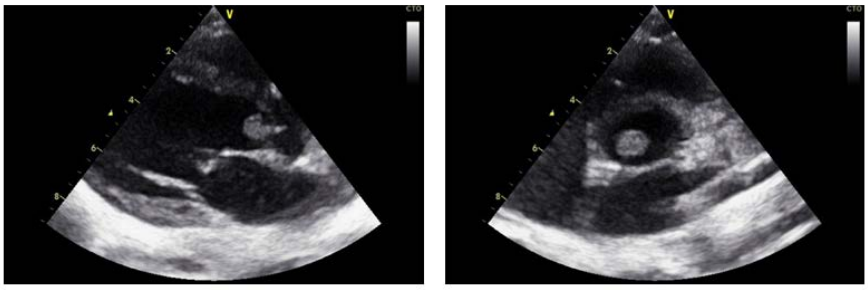
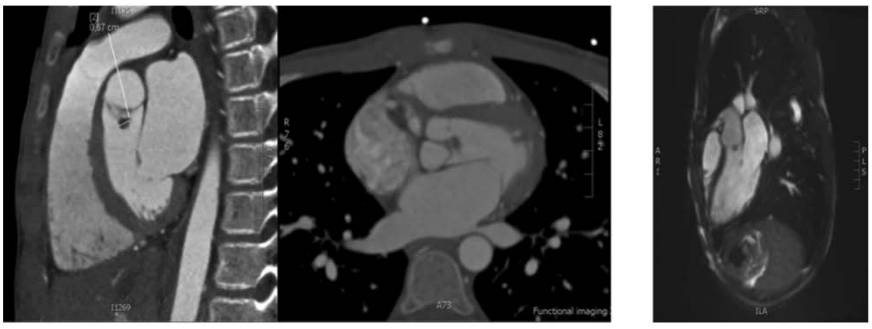
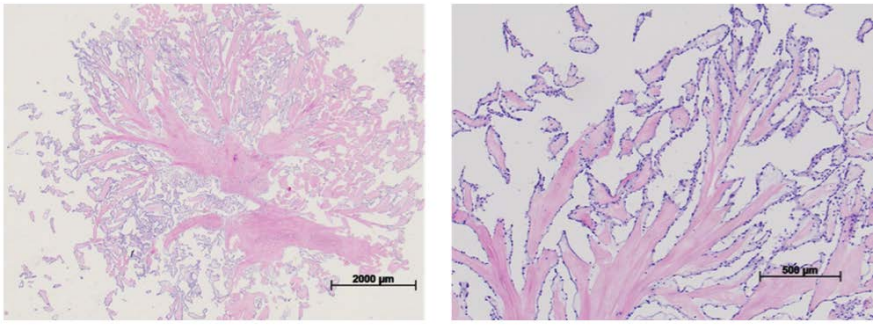


Figure 1. Transthoracic echocardiography. Parasternal short axis (right) and long axis view (left). A circular mobile mass, about 8 mm in diameter, attached to the ventricular side of the aortic valve, projecting at the free edge of the two leaflets. No stalk could be identified.



A **B**

Figure 2. A) ECG-gated cardiac computed tomography, sagittal (left) and transverse (right) view: confirmation of a bicuspid aortic valve. Just below the raphe connecting the non-coronary and right coronary cusp, there is a nodular lesion of 6×7 mm evoking a fibroelastoma. No calcifications. Normal cardiac measurements with a right dominant coronary artery and the absence of coronary lesions. The ejection fraction is calculated to be 66%. B) Cardiac magnetic resonance imaging, sagittal view: presence of a nodular lesion, approximately 6 mm in diameter, appended under the raphe connecting the non-coronary and right coronary cusp of the bicuspid aortic valve. No other morphological anomalies were highlighted.



A **B**

Figure 3. Microscopic analysis. Hematoxylin and eosin staining; space bars represent 2000 µm (A) and 500 µm (B). On the cross-section, a papillary lesion built up of thin, branching avascular papillary axes consisting of collagen and elastic fibers is seen. The papillae are bordered by a single-layered endothelium without nuclear atypia.

gery via mini sternotomy has been shown effective and safe for the excision of CPFE.[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.[17,18] In elective cases, percutaneous resection of CPFE in adults has also proven feasible and safe.[19] However, in children, conventional cardiac surgery remains at time the gold standard.[11]

Fortunately, recurrence after surgery has not been reported but careful follow-up is necessary.[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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