Papillary fibroelastoma associated with congenital heart disease: a coincidental association or a potential new syndrome?

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Introduction

Cardiac papillary fibroelastomas (PFs) are rare cardiac tumors (1). They may be asymptomatic or may present with embolic phenomena (2). To the best of our knowledge, only a few cases of PF associated with congenital heart disease (CHD) have been previously reported (3-6).

Here, we present three cases of PF. The first two cases of PF are associated with ostium secundum-type atrial septal defect (ASD), and the third case is associated with patent ductus arteriosus (PDA). No cases with a history of infective endocarditis or thrombosis were recorded. The coexistence of these lesions is extremely rare.

Case Report

Case 1

A 44-year-old woman with a complaint of shortness of breath was admitted to our institution; electrocardiography was performed that revealed right atrial enlargement and incomplete right bundle-branch block. A transthoracic and two-dimensional (2D) transesophageal echocardiography (TEE) demonstrated a mass on the aortic valve and an ostium secundum-type ASD with an enlargement of the right atrium and ventricle. The size of the defect was 12×18 mm on 2D-TEE, which was surrounded with thin, floppy interatrial septum. On performing cardiac catheterization, the calculated Qp/Qs and Rp/Rs were 2.1 and 0.03, respectively. Subsequently, she underwent surgery when ASD was closed with an autologous pericardial patch and the tumor was completely excised (Fig. 1). The histology of the mass was consistent with PF. A follow-up echocardiography after 2 years of the procedure showed no residual shunt, aortic regurgitation, and mass.

Case 2

Thirteen years ago, a 52-year-old woman with a complaint of angina was admitted to our institution. Chest X-ray showed moderate cardiomegaly, particularly on the right side. Electrocardiography demonstrated atrial fibrillation, right atrial enlargement, and incomplete right bundle-branch block. TEE detected a mass on the aortic valve and a secundum-type ASD. On performing 2D-TEE, the size of the defect was 17 mm with deficient aortic rim. Right heart catheterization revealed a Qp:Qs ratio of 1.88. A coronary angiogram revealed an anomalous single coronary artery originating from the right coronary sinus without stenosis (Fig. 2). The patient had no previous history of neoplasia. The tumor was completely removed, ASD was surgically closed, and histological examination confirmed the diagnosis of cardiac PF. Postoperative TEE demonstrated normal aortic valve function.

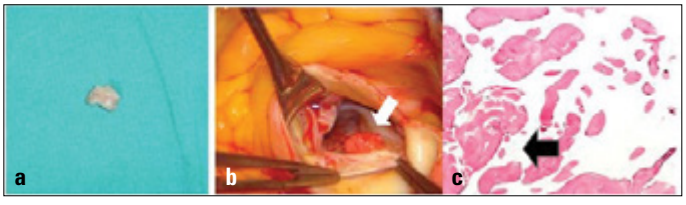

Figure 1. a-c. (a) Gross images of a cardiac papillary fibroelastoma. A 10×15-mm mass has a characteristic flower-like appearance. (b) Papillary fibroelastoma of the aortic valve. Photograph of the lesion obtained at the operating room. (c) Microscopic appearance of the papillary fibroelastoma (H&E stain)

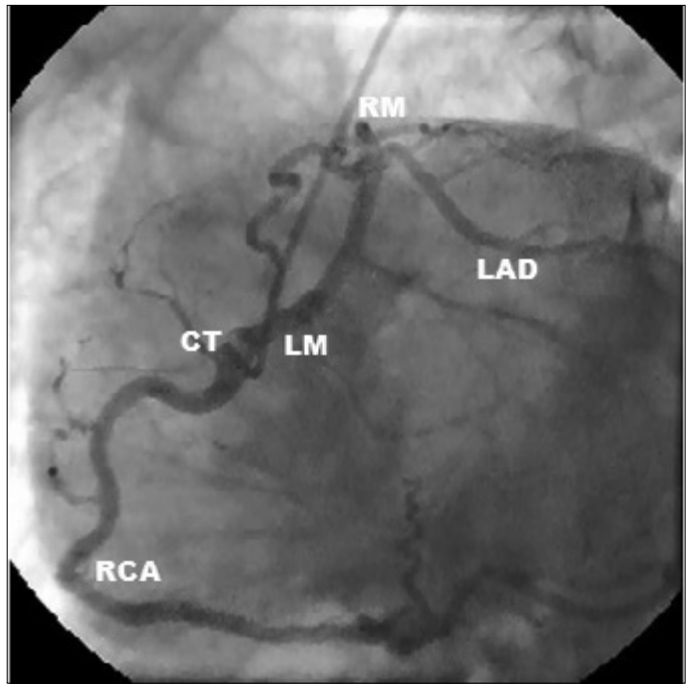

Figure 2. Angiogram obtained from a 52-year-old woman in the left anterior oblique cranial projection. In this view, the entire coronary system is visualized from a single ostium that is located at the right sinus. Common trunk (CT), left main trunk (LM), septal branch (SB), left anterior descending (LAD), and ramus (RM) branches

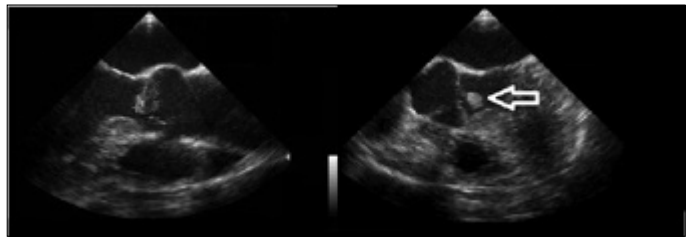

Figure 3. Transesophageal echocardiography reveals a 1-cm^2 mass on the aortic valve, and continuous murmur was heard on the left sternal border

Case 3

A 42-year-old woman with a complaint of dyspnea on effort was admitted to our hospital in 2006. On conducting physical examination, blood pressure was 152/84 mm Hg, and continuous murmur was heard on the left sternal border. TEE revealed PDA and a mass with a size of 11 mm on the aortic valve (Fig. 3, Video 1). Abdominal ultrasonography showed an atrophic left kidney. PDA was ligated and aortic mass was excised. After 4 years, a continuous murmur was again heard on the left sternal border. In the thoracic X-ray, a prominent pulmonary artery trunk and increased pulmonary vasculature were observed. Transthoracic and TEE again revealed

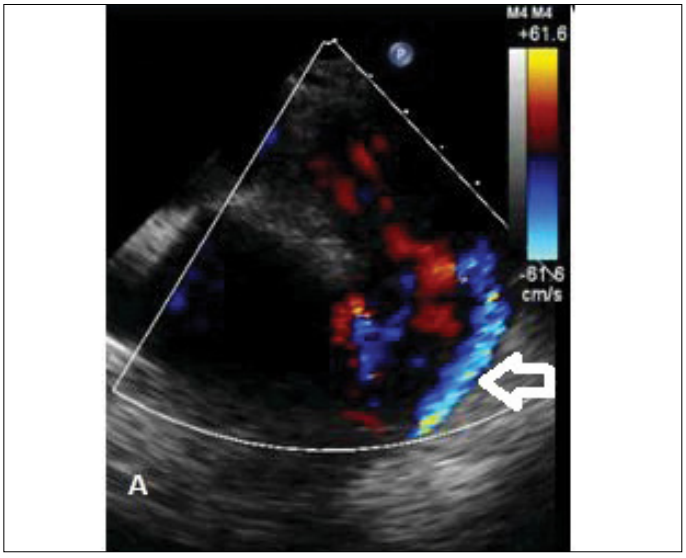

Figure 4. Color Doppler TEE image of a PDA

Table 1. Cases of papillary fibroelastoma associated with congenital heart diseases

Author, year	Age, sex	CHDs	PF location and size, mm
Morishita, 2013	76, M	PDA	AoV, 5
Betigeri, 2011	33, M	AV canal defect (ASD + Cleft mitral)	IVS crest, 20×30
Abad, 2008	60, M	PLSVC, ASD	RA (IAS), 15×20
Watanabe, 1996	64, F	ASD	TV, 11
Current Study			
Patient 1	44, F	ASD	AoV, 9
Patient 2	52, F	ASD, coronary anomaly	AoV, 6
Patient 3	42, F	PDA	AoV, 11

AoV - aortic valve; ASD - atrial septal defect; AV - atrioventricular; CHDs - congenital heart diseases; F - female; IAS - interatrial septum; IVS - interventricular septum; M - male; PDA - patent ductus arteriosus; PF - papillary fibroelastoma; PLSVC - persistent left superior vena cava; RA - right atrium; TV - tricuspid valve.

PDA, which was considered to be caused by suture loosening, and an absence of mass on the aortic valve (Fig. 4). An 8×10-mm Cardiofix device (Starway Medical Technology Inc., Beijing, China) was successfully implanted for PDA. The follow-up course was uneventful. Moderate-to-severe hypertension developed, and nephrectomy was performed a year ago.

Discussion

PFs are uncommon, with an incidence of 7%–8% in all primary cardiac tumors. A majority of PFs occur on the left side of the heart and generally involve the heart valves (1, 2, 7). An association of PF with ASD or other CHDs is rare. To date, four cases of PF associated with CHDs have been reported in the literature (Table 1) (3-6).

In this report, we present a potential new syndrome, which may explain some types of PFs associated with CHDs. To our knowledge, there has been no previous report with direct suggestion of the PF as a more prevalent link of CHDs. Further research on PF associated with CHD syndromes is required with a focus on epidemiology, physiopathological mechanisms, clinical/radiological features, and treatment strategies.

Conclusion

On the basis of the obvious similarities between our cases and those of the other published reports, we propose that a combination of PF and CHDs may represent a recognizable, albeit a rare spectrum of anomalies. We report these cases in the hope that the presence of CHDs will alert the cardiologist to detect a possible PF or vice versa.

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Video 1. Transesophageal echocardiography showed a mass on the aortic valve short axis

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