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. A transthoracic and two-dimensional (2D) transesophageal echocardiography (TEE) detected a mass on the aortic valve and an ostium 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.

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. Transthoracic and TEE again revealed
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, physiological 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.

References

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

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Bonsai-induced Kounis Syndrome in a young male patient

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Introduction

The use of cannabis and its synthetic derivative, bonsai, has recently increased, and it has become an important health problem (1). Kounis Syndrome is a rare and potentially life-threatening syndrome that occurs when an allergic reaction to an irritant is superimposed on a coronary atherosclerotic plaque, leading to an acute coronary syndrome. The syndrome is characterized by the sudden onset of chest pain, ECG changes, and a typical temporal pattern, usually starting with the ingestion of the irritant and progressing over a few days or weeks.

Bonsai is a synthetic cannabinoid that is often used as a recreational drug. It is similar in structure to tetrahydrocannabinol (THC), the main psychoactive component of marijuana, but it has a more potent effect. Bonsai-induced Kounis Syndrome is a rare and potentially life-threatening syndrome that occurs when an allergic reaction to bonsai is superimposed on a coronary atherosclerotic plaque, leading to an acute coronary syndrome.

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

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

PDA - patent ductus arteriosus; ASD - atrial septal defect; IVS - interventricular septum; AoV - aortic valve; F - female; IAS - interatrial septum; M – male; PLSVC - persistent left superior vena cava; TV - tricuspid valve.