Platypnea-Orthodeoxia Syndrome Associated with Spontaneously Ruptured Chordae Tendineae of Tricuspid Valve

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INTRODUCTION

Platypnea-orthodeoxia syndrome (POS) was first described in 1949,[1] and a patient with a patent foramen ovale (PFO) presenting POS was first reported in 1981.[2] Cardiac POS has been associated with abnormal intracardiac communications (i.e., PFO, atrial septal defect, and fenestrated atrial septal aneurysm), which represent the most common etiology of POS. Even a patient with previously asymptomatic PFO may develop right-sided heart pressure overload, inducing a right-to-left shunt. A persistent eustachian valve or Chiari network can increase streaming blood flow from the inferior vena cava to the atrial septum, regardless of right heart pressure. Additionally, the upright position deforms the right atrium and atrial septum, which can be exacerbated by aortic elongation or aneurysm, leading to the development of abnormal shunting. These anatomic and hemodynamic characteristics are associated with the mechanism of cardiac POS. In the present case, spontaneously ruptured chordae tendineae of the tricuspid valve anterior leaflet with concomitant high right atrial pressure induced a large amount of right-to-left shunt via the PFO. To date, only a few reports are available of spontaneous tricuspid valve chordal rupture, and the present case is the first to demonstrate POS induced by spontaneous tricuspid valve chordal rupture.

CASE PRESENTATION

A 50-year-old woman, with no medical history, presented to the emergency department with acute dyspnea and cyanosis. The patient's blood pressure was 120/70 mm Hg, exhibiting no shock. Oxygen saturation level was 70% on room air, and cardiac auscultation revealed a high-pitched holosystolic murmur. Chest radiography revealed the absence of cardiomegaly, congestion, or pulmonary arterial dilatation. Following the administration of 5 L/min oxygen, the oxygen saturation level was 90% in the supine position and 77% in the sitting position, with markedly worsened cyanosis. The presence of orthostatic hypoxia with systolic heart murmur suggested cardiac-related POS accompanied by intracardiac shunting. Transthoracic echocardiography was used for verification. The latter showed tricuspid valve regurgitation with dilated right atrium and right-to-left shunt through the atrium septum (Figures 1 and 2, Video 1). Next, transesophageal echocardiography (TEE) demonstrated the following findings: dilated right atrium with atrial septal aneurysm, ruptured chordae tendineae of the tricuspid valve anterior leaflet with severe tricuspid regurgitation, and right-to-left shunt flow via the PFO (Figures 3–5, Videos 2-4). Microbubble contrast promptly appeared in the left atrium within three cardiac cycles following injection of intravenous agitated saline (Video 5). Additionally, TEE confirmed the distinctly increasing shunt flow occurring in the sitting position.

Furthermore, right heart catheterization revealed high right atrial pressure. On the contrary, pulmonary capillary wedge pressure, pulmonary artery pressure, and right ventricular pressure were normal. These physiologic data were consistent with primary tricuspid valve disorder; the diagnosis of tricuspid valve chordal rupture could be confirmed considering the findings on TEE.

The patient underwent urgent surgical intervention, which included tricuspid valve repair and PFO closure. Surgery was successful, demonstrating ruptured chordae tendineae of the tricuspid valve (Figure 6, *arrow*) and the PFO with a diameter of approximately 15 mm (Figure 6, *arrowhead*). Following surgery, the patient's arterial oxygen saturation was recovered regardless of her posture, and her symptoms disappeared.

DISCUSSION

Although POS is relatively rare, it is a clinically significant condition accompanied by posture-dependent hypoxia. A recent study has classified POS into three groups: intracardiac abnormalities, extracardiac abnormalities, and others.[3] Intracardiac POS is accompanied by intracardiac shunting such as PFO, atrial septal defect, and fenestrated atrial septal aneurysm. These conditions cause a right-to-left shunt increase while in the sitting or standing position. Upright posture is believed to cause the right atrium and atrial septum to stretch and deform, resulting in increased venous flow from the inferior vena cava to atrial septum. Of note, an earlier study has reported that an ascending aortic elongation or aneurysm can promote the shunting flow.[4,5] Alternatively, extracardiac POS comprises pulmonary arteriovenous shunts and pulmonary parenchymal diseases (e.g., pulmonary ventilation-perfusion mismatch).

The primary etiology of POS is cardiac-related POS, a condition observed in patients with abnormal intracardiac shunting.[3] In a retrospective analysis, rates of cardiac and extracardiac POS were 87.0% and 13.0%, respectively.[3] The evaluation of oxygen saturation levels in both the supine and sitting positions is an elementary and essential examination for POS diagnosis. Additionally, a history of worsening cyanosis and dyspnea in the upright position is critical. Furthermore, cardiac auscultation has diagnostic significance. Specifically,

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VIDEO HIGHLIGHTS

Video 1: Transthoracic echocardiography, modified short-axis view, showing right-to-left shunt through atrial septum with dilated right atrium and severe tricuspid regurgitation.
Video 2: TEE showing tricuspid valve anterior leaflet flail.
Video 3: TEE showing atrial septal aneurysm and the PFO.
Video 4: TEE showing massive right-to-left shunt flow via the PFO.
Video 5: TEE showing the prompt occupation with micro-bubble contrasts through the PFO to left atrium and ascending aorta.

View the video content online at www.cvcasejournal.com.

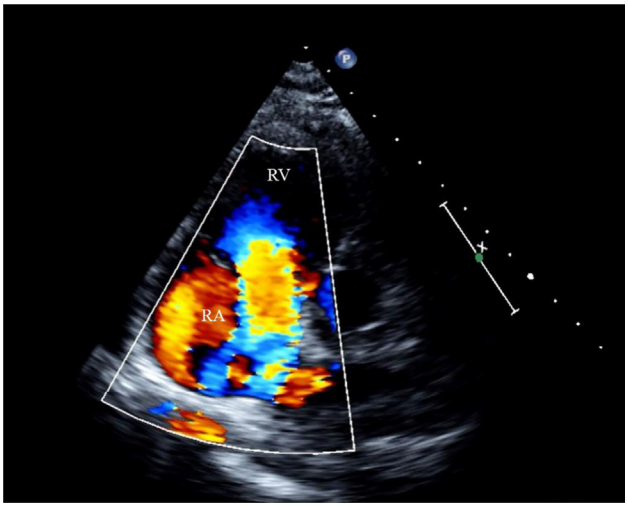

Figure 1 Transthoracic echocardiography, modified parasternal short-axis view, showing dilated right atrium (RA) and severe tricuspid regurgitation. *RV*, Right ventricle.

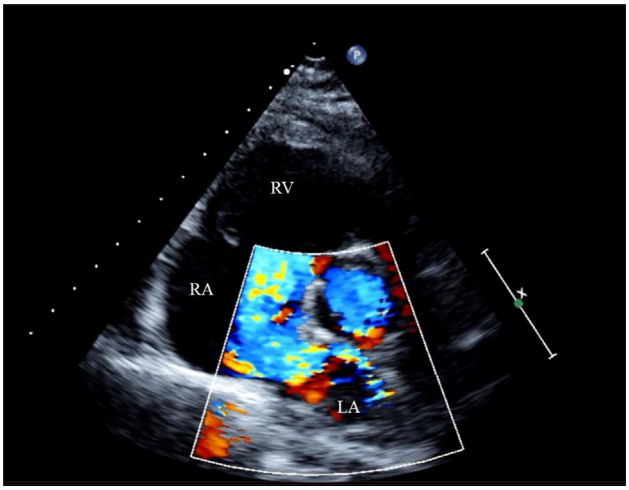

Figure 2 Transthoracic echocardiography, modified parasternal short-axis view, showing right-to-left shunt flow through atrial septum. *LA*, Left atrium; *RA*, right atrium; *RV*, right ventricle.

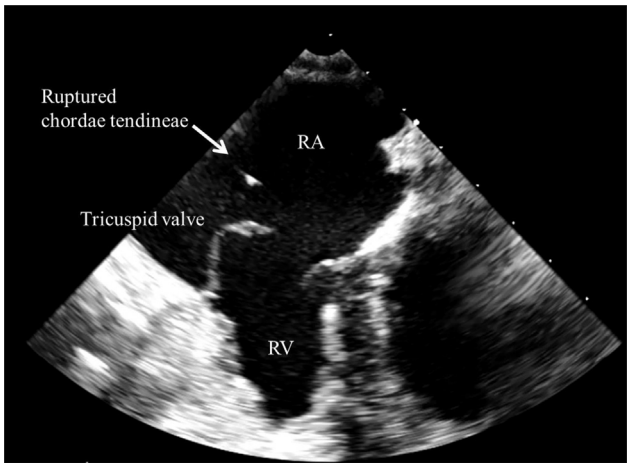

Figure 3 TEE showing severe tricuspid prolapse following ruptured chordae tendineae of anterior leaflet. *RA*, Right atrium; *RV*, right ventricle.

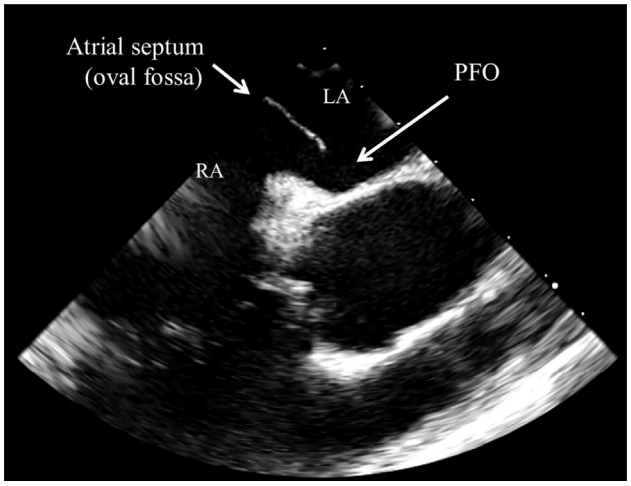

Figure 4 TEE showing atrial septal aneurysm and the PFO. *LA*, Left atrium; *RA*, right atrium.

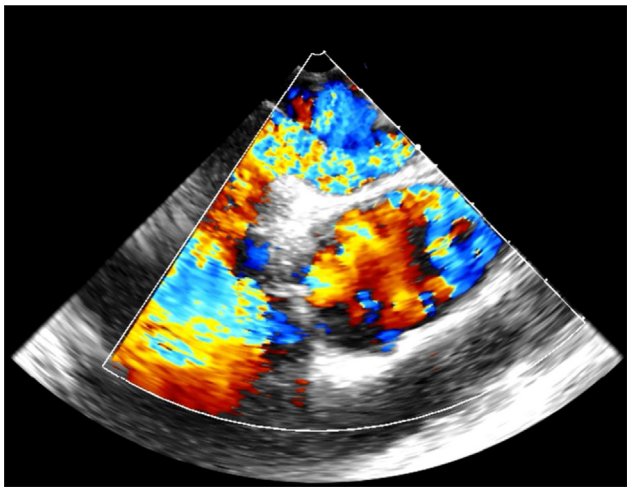

Figure 5 TEE showing massive right-to-left shunt flow via the PFO.

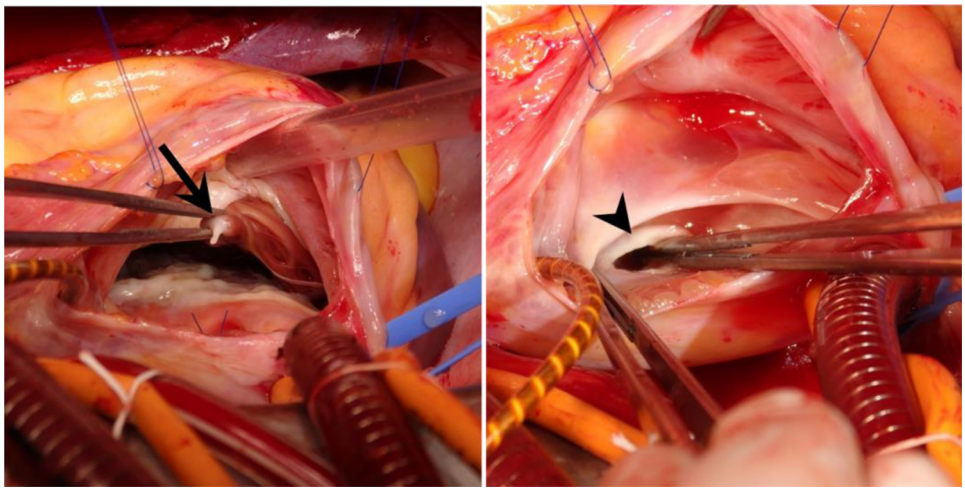

Figure 6 Surgical findings showing the ruptured chordae tendineae of tricuspid valve anterior leaflet (*arrow*) and the PFO (*arrowhead*).

orthostatic hypoxia with systolic heart murmur suggests cardiac POS accompanied by intracardiac shunting. Finally, a combination of both anatomic and hemodynamic findings detected with precision by TEE confirmed the diagnosis of POS.

In our case, a patient with potential asymptomatic PFO exhibited sudden onset of dyspnea and cyanosis. Spontaneous rupture of the chordae tendineae of the tricuspid valve caused tricuspid regurgitation and right atrial pressure overload, resulting in right-to-left shunting. Such observations accounted for an extremely unusual case. The patient had no apparent triggers, and dyspnea started suddenly, during an ordinary meeting at work.

Generally, rupture of the chordae tendineae of the tricuspid valve is primarily induced by blunt chest trauma, and previous reports have described the occurrence of tricuspid chordal rupture due to trauma.[6-9] Although other etiologies have been reported (e.g., infective endocarditis, myocardial infarction, congenital disorders,[10,11] iatrogenic complication of endomyocardial biopsy[12,13]), only a few studies have described spontaneous tricuspid chordal rupture.[14,15] Irrespective of the cause, POS induced by rupture of the chordae tendineae of the tricuspid valve should be rapidly diagnosed to promptly proceed with the adequate surgery needed to resolve the hemodynamic compromise.

CONCLUSION

We report a case of cardiac-related POS induced by an uncommon spontaneous rupture of the chordae tendineae of the tricuspid valve. The condition should be immediately diagnosed by measuring oxygen saturation level in both the supine and upright positions and accurately evaluating anatomic and hemodynamic abnormalities using TEE. Appropriate treatment of tricuspid valve repair and PFO closure were successfully performed in the present case.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2019.10.006.

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