



Main Pulmonary Artery Aneurysm and Fish Mouth Pulmonary Valve: A Case Report

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Abstract

Introduction: Bicuspid pulmonary valve as a rare cardiac anomaly results in hemodynamic alterations leading to aneurysmal dilation of the pulmonary artery (PA), which has been reported in association with other heart defects. Pulmonic stenosis is usually presented as a part of a complicated disease (e.g. tetralogy of Fallot) or in association with other abnormalities such as corrected transposition of great arteries, although it can be developed as a distinct entity.

Case Presentation: The case described in this study was a 36-year-old man with a history of syncope and dyspnea. The complementary studies revealed isolated bicuspid PS and aneurysmally dilated main PA.

Conclusion: Both main PA aneurysm and fish mouth bicuspid pulmonary valve are rare cardiac anomalies that are sometimes presented in association with other heart diseases. Pulmonic stenosis can occur as an isolated anomaly but more often is a part of complex defects. The present report emphasizes the fact that an isolated bicuspid anomaly of the pulmonary valve may be compatible with normal cardiac anatomy and remain uneventful even on its own accord, although our case presented signs and symptoms of severe pulmonic stenosis at the age of 36.

Keywords: Bicuspid pulmonary valve, Main pulmonary artery, Aneurysm

Introduction

Both aortic and pulmonic valves are originated from the junction of conus arteriosus and the bulbus cordis during cardiovascular system development (1). Both of these semilunar valves are normally tricuspid guarding the 2 orifices which are formed by sequential events out of the existing 4 primitive cushions (ventral, dorsal, right and left). The left and the right cushions each are separated into 2 distinct cusps by a transversely oriented 'distal bulbar septum', then join with the ventral and dorsal cushions cusps (2-5). Therefore both these semilunar valves consist of 3 leaflets with 3-dimensional attachment being the mirror image of the other.

The segment located in the anterior part of the bulbar septum consists of the pulmonary opening whereas the posterior part of septum develops into the aortic orifice (6).

The development of anomalous pulmonary valves as a consequence of abnormal function and the migration disorder of neural crest cells has been previously explained (7).

Bicuspid pulmonary valve as a rare cardiac anomaly results in hemodynamic alterations leading to aneurysmal dilation of the pulmonary artery (PA), which has been reported in association with other heart defects. A study of

3861 donor hearts, dissected at the European Homograft Bank revealed only 4 cases (0.1%) of bicuspid pulmonary valves. PA aneurysms are even rarer, with only 8 cases documented in 109 571 autopsies. Aneurysmal dilation of main PA could be as a result of the stenotic pulmonary valve as well as the abnormal migration of neural crest cells. Normal embryologic development of semilunar valves and great vessels, the outflow tract division (into the PA and aorta) and completion of aortic arch are necessarily depended on migrating neural crest cells (8).

The congenital etiologies for abnormal pulmonary valves include the absence or addition of one or more cusps including acommisural, unicommissural, bicuspid or dysplastic (9).

Pulmonic stenosis is usually presented as a part of a complicated disease (e.g. tetralogy of Fallot) or in association with other abnormalities such as corrected transposition of great arteries, although it can be developed as a distinct entity. Two-dimensional echo imaging of the bicuspid pulmonic valve shows thickened leaflets with systolic bowing. On Doppler interrogation, the antegrade velocity is increased with the corresponding maximum and mean pressure gradient via the Bernoulli equation (10).

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Case Presentation

The present report describes a pulmonary valve with only 2 cusps in a 36-year-old man who was admitted to hospital with syncope, dyspnea and fatigue. In the first evaluation, the patient was tachypneic with arterial desaturation (O₂ sat = 85%). The inspection of the patient showed that there were a remarkable kyphoscoliosis and nail clubbing.

The patient was evaluated with CXR and echocardiography and there was obvious main PA dilation in CXR (Figure 1) and chest CT-scan (Figure 2).

Two-dimensional echocardiography revealed aneurysmally dilated main PA (about 51 mm) (Figure 3). A bileaflet



Figure 1. CXR Showing Chest Deformity and Main PA Aneurysmal Dilation.

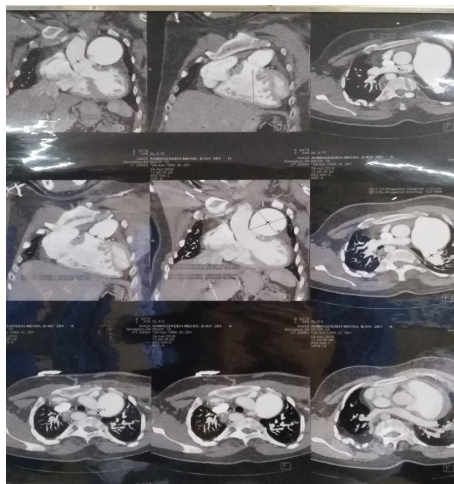


Figure 2. Spiral Chest CT-Scan Showing Main PA Dilation.

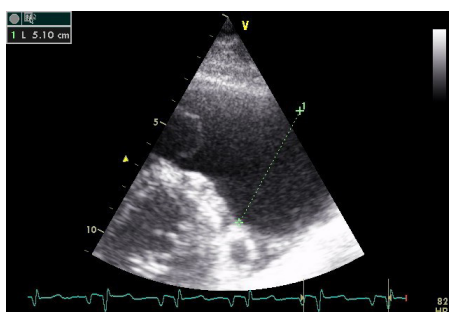


Figure 3. Echo (Modified Parasternal Short-Axis View) Indicating Bowing PV Leaflets and Dilated Main PA.

pulmonic valve has been discovered via modified parasternal short-axis view (Figure 4, Online video file 1) with severe PS but because of the associated chest deformity, Doppler alignment was not possible so peak velocity was underestimated (Online video file 2, Figure 5).

Discussion

Bicuspid pulmonary valve as a rare distinct cardiac anomaly has been previously reported, however, it has been usually explained in relation with other expected cardiac anomalies (11).

Bicuspid pulmonary valve is a rare cardiac anomaly, expressed in association with aneurysmally dilated PA which is usually developed in response to hemodynamic alterations caused by the bicuspid pulmonary valve (poststenotic dilation). However, abnormality in migrating neural crest cells which is essential for normal development of semilunar valves can be another etiological factor (8).

Given our patient's severe degree of pulmonary valve stenosis, it is possible that his PA aneurysm is secondary to hemodynamic alterations attributable to his bicuspid pulmonary valve stenosis.

Conclusion

The congenital etiologies for abnormal pulmonary valves include absence or addition of one or more cusps including a commissural, unicommissural, bicuspid or dysplastic. Pulmonic stenosis can occur as an isolated anomaly but more often is a part of complex defects (for

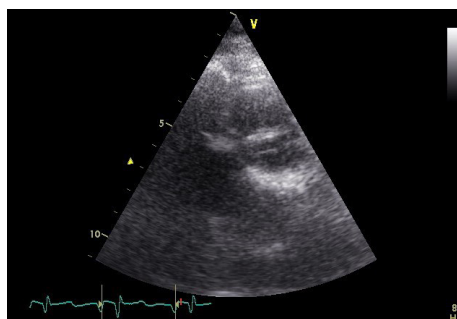


Figure 4. 2D Echo (Modified Parasternal Short-Axis View) Indicating Bicuspid Pulmonary Valve.

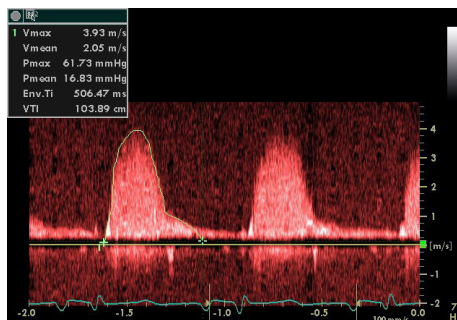


Figure 5. Echo Doppler Indicating Severe Pulmonary Valve Stenosis.

example, tetralogy of Fallot or corrected transposition).

Both main PA aneurysm and fish mouth bicuspid pulmonary valve are rare cardiac anomalies that are sometimes presented in association with other heart anomalies. Pulmonic stenosis can occur as an isolated anomaly but more often is a part of complex defects. The present report emphasizes the fact that an isolated bicuspid anomaly of the pulmonary valve may be compatible with normal cardiac anatomy and connection. Though very unique and rare in its incidence, a bifoliate pulmonary valve can exist without other expected cardiac anomalies and remain uneventful even on its own accord (12-14), although our case presented signs and symptoms of severe pulmonic stenosis at the age of 36.

Conflict of Interests

None to be declared.

Ethical Issues

Written informed consent was obtained from the patient for the publication of this case report and any connected images and videos.

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None.

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Supplementary Materials

The online version of this article contains video files 1 and 2.

Video 1. Modified parasternal short axis view indicating bicuspid pulmonary valve.

Video 2. Parasternal short axis view illustrating dome shaped opening bicuspid pulmonary valve.

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