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Calcific Pulmonary Valve Stenosis in Patient with Ostium Secundum Defect

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Abstract

Dystrophic calcification of the pulmonary valve in a Ostium secundum defect patient requiring valve replacement is a rare occurrence.

In this paper, we report one such patient, a case of Ostium Secundum atrial septal defect with calcific pulmonary valve stenosis in middle age. Infective endocarditis was the most probable etiology for the calcification of the pulmonary valve. He underwent a pulmonary valve replacement with a bioprosthetic valve, pericardial patch closure of the atrial septal defect and tricuspid valve annuloplasty. The literature review shows four such reported cases.

Keywords: Calcific pulmonary valve; Pulmonary valve replacement; Infective endocarditis; Atrial septal defect

Introduction

Calcification of the pulmonary valve (PV) is a rare entity, in contrast to aortic calcific stenosis. Calcific pulmonic stenosis occur secondary to rheumatic, congenital, carcinoid, infective endocarditis etiologies. To date, 16 cases have been reported in the literature [1]. Despite similar predisposing factors such as raised ventricular ejection pressure against a dysplastic valve leading to calcification of the valve, the occurrence of calcium deposition on the pulmonary valve is extremely rare.

Occurrence of isolated pulmonic valve endocarditis is also rare, incidence less than 1.5% [2]. The literature includes 45 cases described between 1960 and 2005 [3].

Here, we report one such patient, a case of Ostium Secundum atrial septal defect with calcific pulmonary valve stenosis with healed vegetation's requiring pulmonary valve replacement.

Presentation

A 38-year-old gentleman was admitted in our hospital for complaints of dyspnea, intermittent chest discomfort of 2 years duration and worsening of symptoms from one month prior to admission.

There was no previous exposure to infectious disease, dialysis or intravenous drug abuse. Patient had undergone inguinal hernia repair 5 years prior, with no record of central venous catheter usage during the perioperative period.

Cardiovascular examination revealed signs of right ventricular enlargement, a systolic murmur grade 3/6 loudest over left upper sternal border. On abdominal examination liver was palpable about 5 cm below right costal margin, tender, smooth surface, firm in consistency.

Blood investigation revealed leukocytosis (17,900 cells / cubic mm), raised bilirubin (1.5 milligram/ deciliter) and normal renal function. Blood cultures drawn at the time of admission were negative for any pathogens.

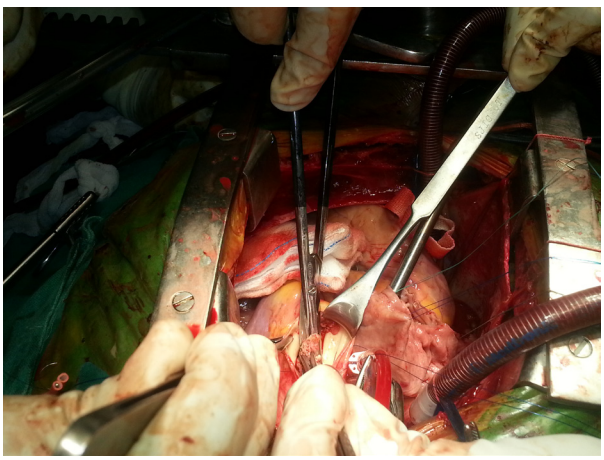
Chest X- ray showed cardiomegaly with cardiothoracic ratio of 0.8, right atrial and ventricular enlargement, enlarged pulmonary artery segment and pulmonary plethora.

Transthoracic Echocardiography showed dilated right heart chambers (right ventricular dimension in diastole = 3.30mm) and severe right ventricular dysfunction, thickened pulmonary valve leaflets with dense echoes on leaflets suggestive of healed vegetation in a background of dense calcification and reduced opening. *Continuous* wave Doppler measured a peak systolic gradient of 90 and a mean gradient of 45 across the pulmonary valve. Color Doppler showed grade I pulmonary regurgitation, grade III tricuspid regurgitation, a left to right shunt across a ostium secundum atrial septal defect (ASD) with normal pulmonary venous drainage.

The ASD was not amenable to device closure as the size was more than 40 mm in its largest dimension and had deficient aortic and Caval rims. The pulmonary valve was not tackled by balloon dilatation as it was a thickened and a calcific valve with grade III regurgitation in a setting of severe RV dysfunction. Besides need for surgical pulmonary valve intervention would preclude a device closure of the ASD.

Intra-operatively, right atrium, right ventricle & pulmonary trunk were dilated. Annulus was densely calcified, with calcification extending to the pulmonary valve leaflets, commissures and extending into the infundibulum. There were healed vegetations on the valve leaflets.

Pulmonary valve leaflets were excised and calcific deposits over the annulus debrided. A bioprosthetic valve, a 25 mm HANCOCK II T510 Cinch (Medtronic, Santa ana, CA, USA) aortic valve implanted in an intra annular position with nonpledgeted interrupted polyester sutures (Centibond, Murdabad, Thane, India) (Figure 1).

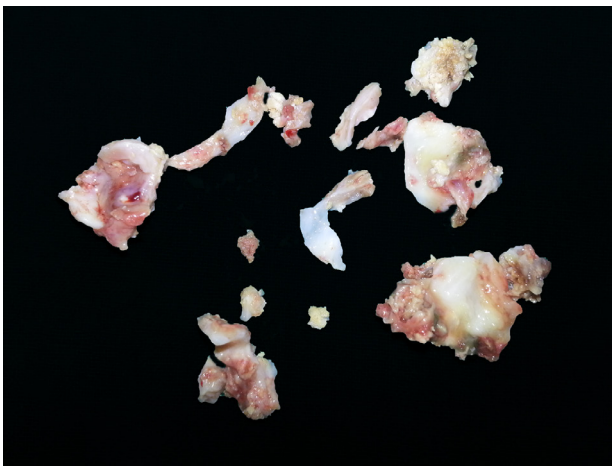


a. Excision of the calcified pulmonary valve

Atrial septal defect was closed with pericardial patch & tricuspid annuloplasty was done with a Teflon collar. Pulmonary artereotomy required augmentation with a freshly harvested autologous glutaraldehyde fixed pericardial patch.

Postoperative echocardiography showed normally functioning bioprosthetic valve prosthesis, trivial pulmonary regurgitation, mean pulmonary valve gradient of 15, normal left ventricular function.

Histopathological examination of the resected valve specimen revealed a sclerotic calcified valve with vegetation, and dense infiltration of polymorphs. Blood and valve cultures were negative for any pathogens (Figure 2).



Specimen showing dense calcified pulmonary valve leaflets

Postoperative course was uneventful; patient was discharged on 8th post-operative day. Patient is on regular follow up (1 yr.), has resumed his farming activities and remains asymptomatic.

Discussion

Infective endocarditis of a calcific pulmonary valve is a rare entity. Rarity of infective endocarditis of pulmonary valve may be due to the low-pressure gradients within the right heart, the lower oxygen content of venous blood and the differences in the covering and vascularization of the right heart endothelium. The prerequisites for calcium deposition are a severe degree of valvular pulmonary stenosis and prolonged elevation of right ventricular pressure. In our patient, the longstanding functional pulmonary stenosis and the left to right shunt might have created an ideal setting for infective endocarditis of the pulmonary valve to occur and subsequent dystrophic calcification .

The diagnosis of pulmonary valve probable infective endocarditis in our patient was made based on modified Duke criteria [6].

Reports of isolated pulmonary stenosis with pulmonary valve replacement are also rare. Because of the heavy calcific deposits at pulmonary valve leaflets, pulmonary valvotomy was not feasible. Hence pulmonary valve was replaced with bioprosthetic valve.

Conclusion

Calcific pulmonary valve stenosis of infective etiology is a rare entity. Here, we report a rare case of calcified pulmonary valve with healed vegetation in a middle aged gentleman with ostium secundum ASD requiring pulmonary valve replacement .With literature review, to our knowledge this is the fourth case of Ostium Secundum ASD with calcific pulmonary valve [5].

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