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EC CARDIOLOGY
Case Report

Atypical Presentation of Infective Endocarditis in Congenital Heart Diseased Patient

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Abstract

Echocardiographic criteria of typical infective endocarditis include presence of vegetations at atrial side of atrioventricular valves (mitral and tricuspid valves) and ventricular side of semilunar valves (aortic and pulmonary valves). Right side infective endocarditis which affect tricuspid valve (TV) and pulmonary valves are very rare and most commonly affect the drug abusers and who have congenital heart disease. In addition, if connective tissue disorder affected the left side heart valves of the same patient with right sided infective endocarditis, it will cause more confusion and dilemma in proper diagnosis of the case. In this case report we present a patient with Marfanoid habitus and atypical tricuspid valve (TV) vegetation at ventricular side, flail mitral valve with severe mitral regurgitation and severe aortic valve regurgitation.

Keywords: *Infective Endocarditis; Congenital Heart Disease; Tricuspid Valve*

Abbreviations

TV: Tricuspid Valve; MVP: Mitral Valve Prolapse; MV: Mitral Valve; MR: Mitral Regurgitation; ARDS: Adult Respiratory Distress Syndrome; TVE: Tricuspid Valve Endocarditis; IE: Infective Endocarditis; AR: Aortic Valve Regurgitation; TTE: Trans Thoracic Echo; TEE: Trans Esophageal Echo; RCC: Right Coronary Cusp

Introduction and Case Report

A tall statured 17Y old previously healthy boy was transferred to our hospital from another facility on ventilator with diagnosis of Septic shock, ARDS and presumed infective endocarditis. bed side echo there, revealed a mass attached to TV and MVP with severe MR.

His family denied history of any high-risk behavior for sexually transmitted diseases, alcohol abuse, intravenous drug abuse, recent dental procedures, persisting skin infections, congenital heart disease or rheumatic fever.

At our institution, the physical exam was remarkable for pan-systolic murmur over the apex, bilateral basal to mid-zone lung crackles. His physique was remarkable for disproportionate ration of the lower extremities to torso and very large upper extremities span, he has a high arched palate and the thumb wrist test was positive all suggested marfanoid habitus. His initial laboratory data showed significant leukocytosis and elevated inflammatory markers and microscopic hematuria. Blood, respiratory and urine cultures were negative. Chest imaging showed pulmonary edema like picture/ARDS. TTE and TEE revealed large sessile cauliflower like mass attached to ventricular

side of TV annulus and septal leaflet with highly mobile sphere like mass attached to its tip without hemodynamic compromise, Flail anterior MV leaflets with ruptured chord at A2, A3 scallops and severe MR, Aortic valve showed retraction of RCC with triangular gape causing severe AR.

The consensus was for surgery to remove the mass and send for C/S and histopathology, repair/replacement of valvular lesions.

Vegetations were removed and sent for culture and histopathology. All culture specimens were negative, Aortic valve specimen revealed No evidence of IE, only myxoid changes (C/W connective tissue diseases). Both aortic and mitral valves were replaced by mechanical valves. Patient was stable post operatively and went home after one week, Returned back to his school, Unfortunately, 4 months later he was expired with sudden cardiac death.

Discussion

We did a literature review about native TV Endocarditis with atypical vegetation at ventricular side and if there any relation to connective tissue disorder like Marfan syndrome, We found that, Ninety-two percent of patients with infective endocarditis, the involvement is in the left side and 8% is in the right side [1]. Isolated tricuspid (TV) endocarditis accounts for 5% - 10% of cases of infective endocarditis (IE)and is uncommon in an immunocompetent adult in absence of risk factors or congenital heart disease [2,3]. TVIE is thought to be rare because the right-sided congenital heart diseases are rare and the tricuspid and pulmonary valves are not strained because of low pressure, and oxygen saturation is low [2]. Persistent fever associated with pulmonary events, anemia, and microscopic hematuria is known as 'tricuspid syndrome', and should alert for TVE, Early Echo is recommended in such patients [4]. Sometimes atypical presentation of vegetations at ventricular side of TV may occur in some Patient with VSD and L-> R shunt which Encroach on the Papillary Muscle and Right Ventricular Cavity.

Echocardiography is the mainstay of assessment of Marfan's syndrome which may include aortic valve with Annuloaortic ectasia, especially with dilatation of aortic root, is found in 60% to 80% of adult cases which can cause severe AR or may progress to aortic root dissection. Aortic root dilatation is the most specific finding in Marfan's syndrome. The pattern of dilatation, a so called "onion shape," is specific. Several issues should be considered while performing a cardiac echo in Marfan's syndrome patient. The Aortic root diameter should be adjusted according to body surface area (BSA) Roman., *et al.* [5] developed a nomogram of the AR diameter at the sinus of Valsalva in relation to BSA according to age; it is currently the most widely cited reference because it covers the entire spectrum of ages. Although the Aortic root in Marfan syndrome is typically described as an onion shape, several variations have been observed. Dilatation of the aortic annulus is associated with the severity of aortic regurgitation

Also, Mitral valve may suffer from MVP which is less benign than the common type of MVP identified in the general population. MVP is often not the only pathology in Marfan syndrome. The prevalence of chordae tendineae rupture in prolapsed valves is higher than that in the normal population probably because of the underlying tissue pathology or the stressed hemodynamics on the prolapse leaflets. Furthermore, because an eccentric turbulent flow is commonly observed from a prolapsed mitral leaflet, risk of infection is higher than that in the normal structure [6]. One retrospective study of mitral valve surgery in Marfan syndrome patients revealed that regurgitation was caused by leaflet prolapse in five of nine patients and by chordal rupture because of endocarditis in two patients; annulus dilatation was observed in all patients [7]. Flail leaflet is an independent predictor of progression of MR and MV-related clinical events.

Back to our case, we found no single case report in the literature with combination of those rare findings of atypical Tricuspid valve vegetation, Marfan syndrome with MVP and ruptured chord and retracted RCC of Aortic valve.

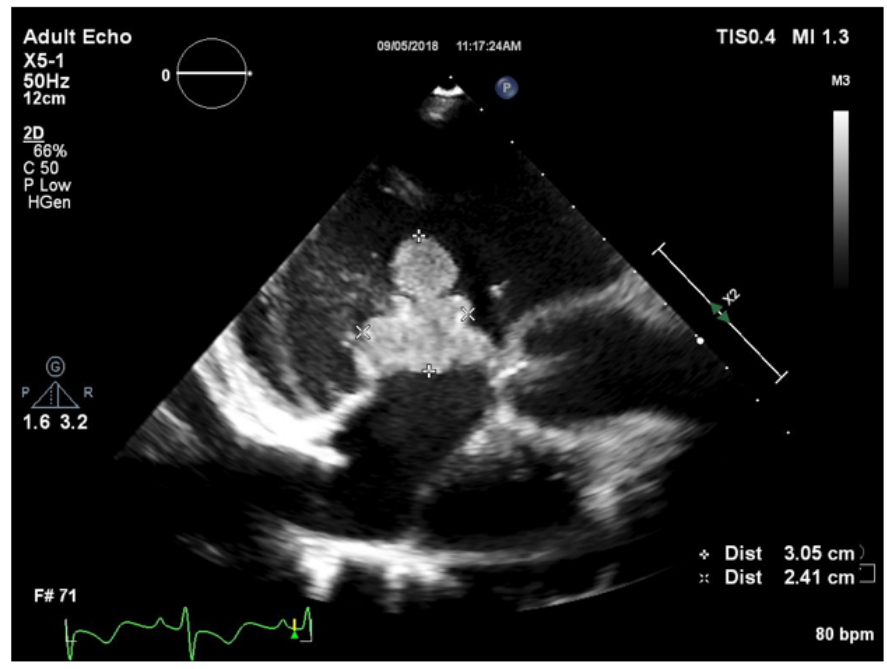

Figure 1: *Bulky mass (vegetation) attached to ventricular side of TV annulus.*

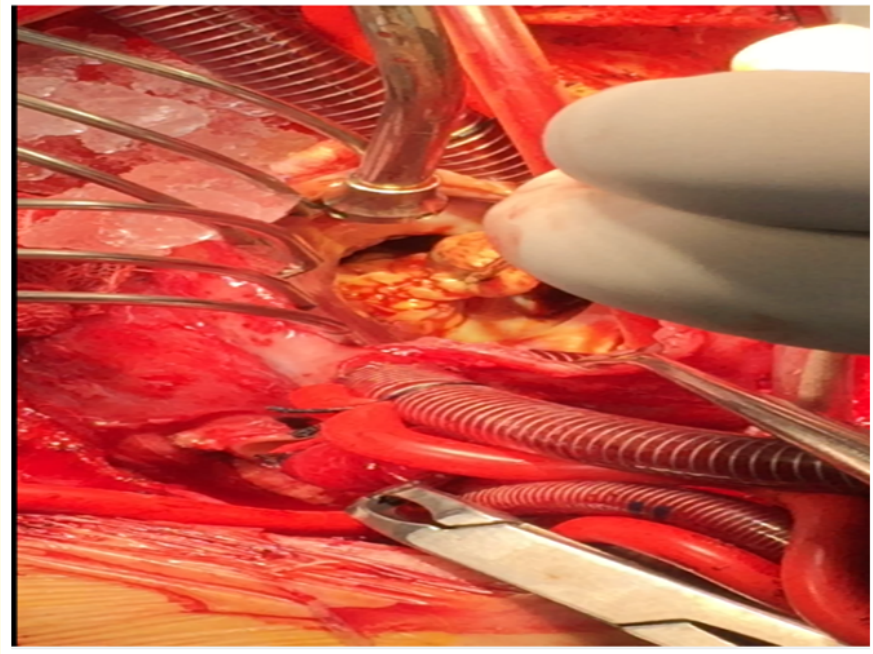

Figure 2: *Surgical removal of vegetation.*

Conclusion

Atypical infective endocarditis can have peculiar presentation, in real life when the same patient has multiple valvular lesions which are diagnosed acutely, it is not mandatory to be of same etiology and you have to search for other causes which may be not related to the same etiology.

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