

Title: ATYPICAL VEGETATION IN CONGENITAL HEART DISEASE

Category: Valvular Heart Disease

Abstract

CASE HISTORY: A tall statured 17 Y 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.

CLINICAL EXAMINATION: 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.

LAB. INVESTIGATIONS: His initial laboratory data showed significant leukocytosis and elevated inflammatory markers and microscopic hematuria. Blood, respiratory and urine cultures were negative.

CHEST X RAY: Chest imaging showed pulmonary edema like picture/ARDS.

ECHOCARDIOGRAPHY : 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 chordi 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 vs replacement of mitral and aortic valves.

SURGICAL DATA: 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 prosthesis.

Literature review : We did a literature review about native TV Endocarditis with atypical vegetation at ventricular side and if there any relationship to connective tissue disorder like Marfan syndrome , We found that, 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 CHD.

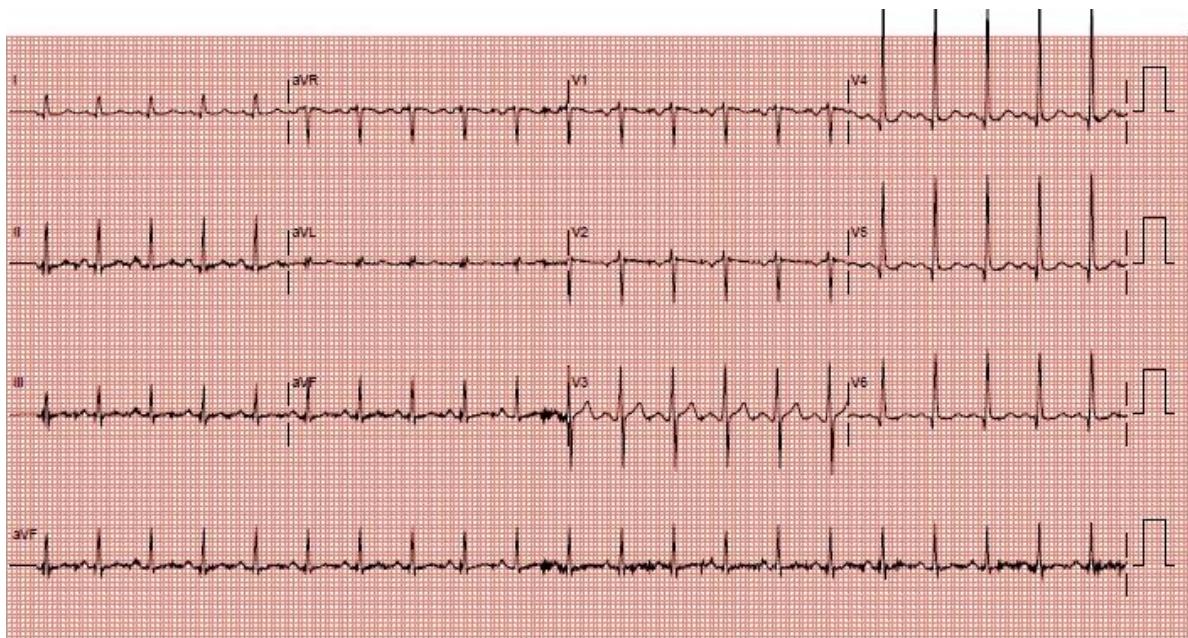
Persistent fever associated with pulmonary events, anemia, and microscopic hematuria is known as 'tricuspid syndrome', and should alert for TVE, Early Echo is recommended in such patients.

Differential Diagnosis: 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.

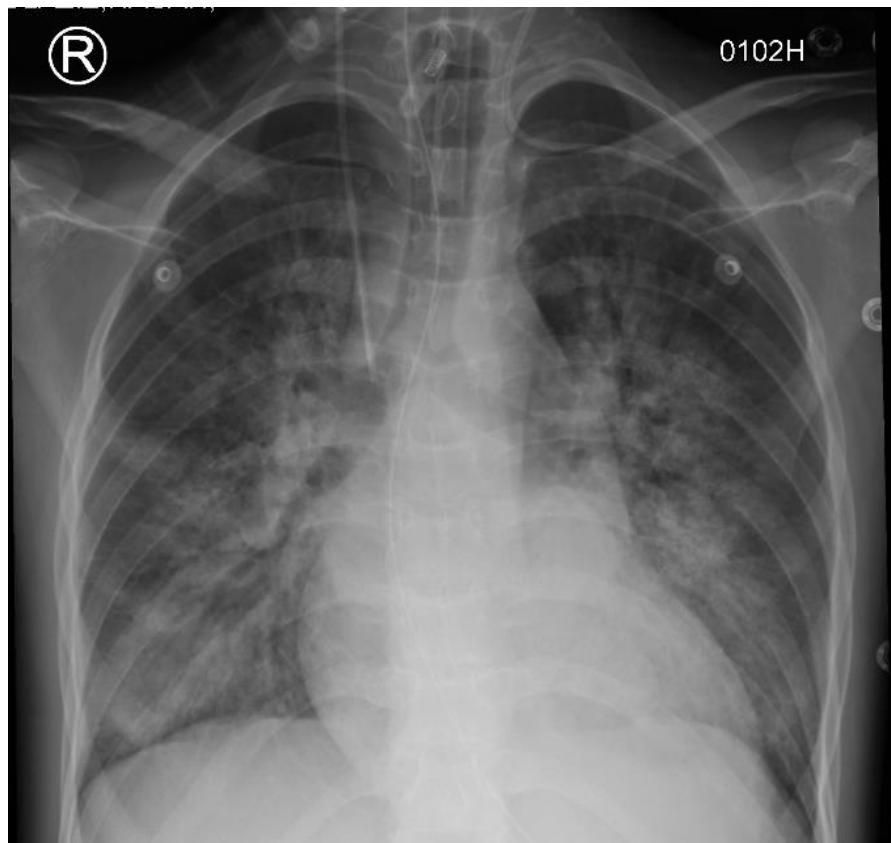
Echocardiography is the mainstay of assessment of Marfan's syndrome which may include aortic valve with Annuloaortic ectasia, especially with dilatation of aortic root, is found in 60% to 80% of adult cases which can cause severe AR or may progress to aortic root dissection.

Also Mitral valve may suffer from MVP which is less benign than the common type of MVP identified in the general population. Flail leaflet is an independent predictor of progression of MR and MV-related clinical events.

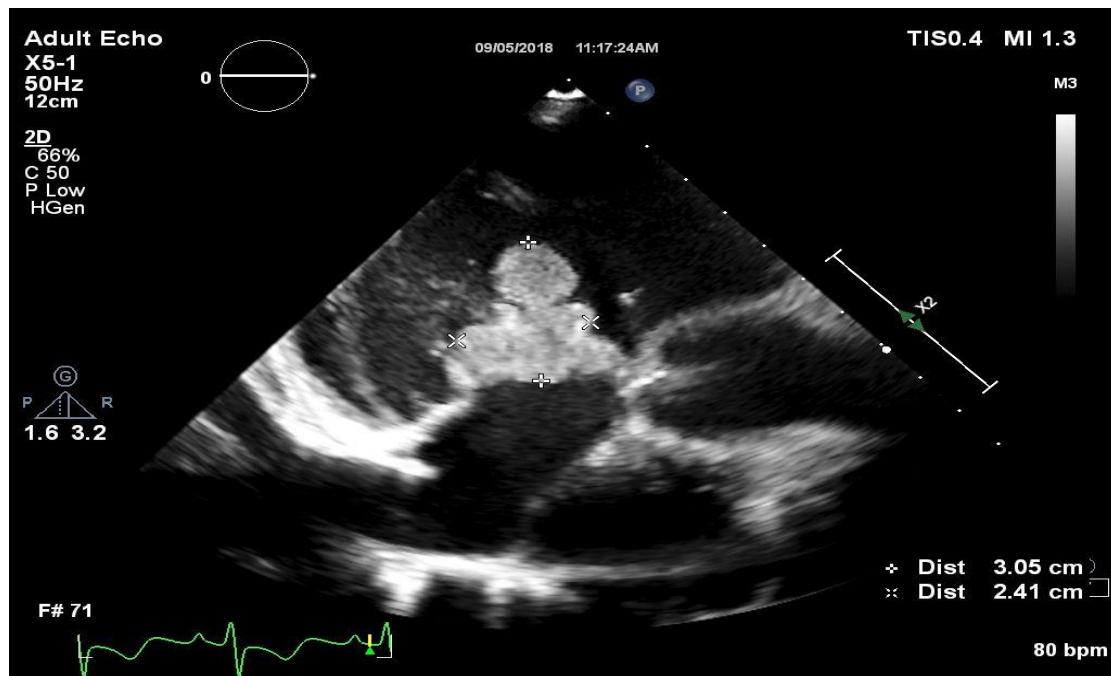
Back to our case, we found no single case report in the literature with combination of such rare findings



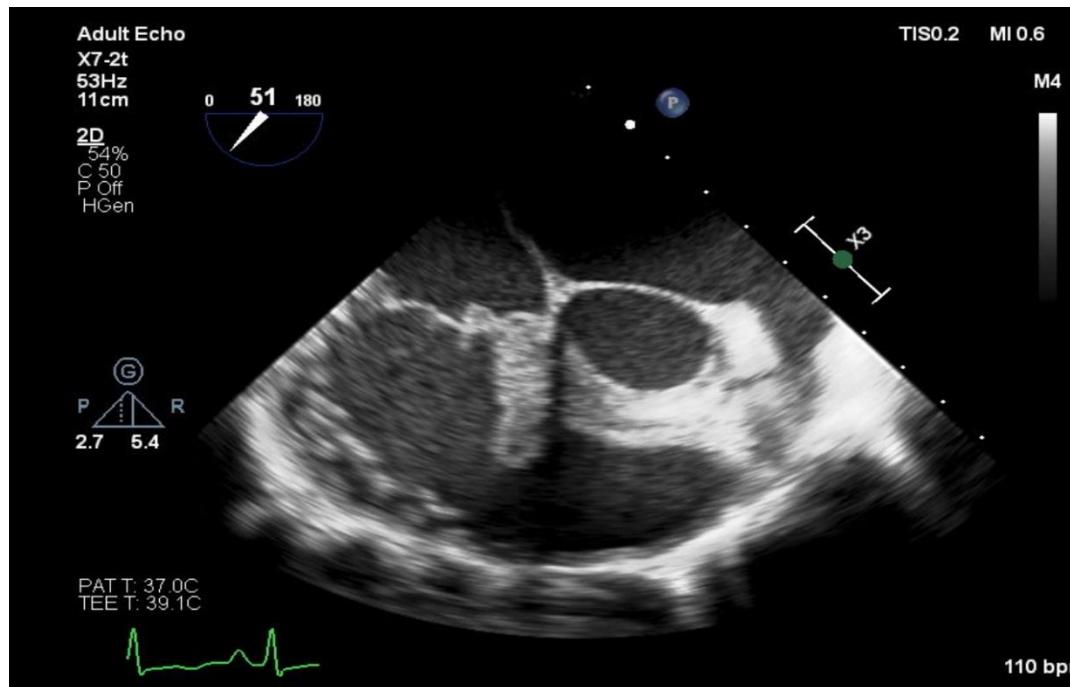
ECG



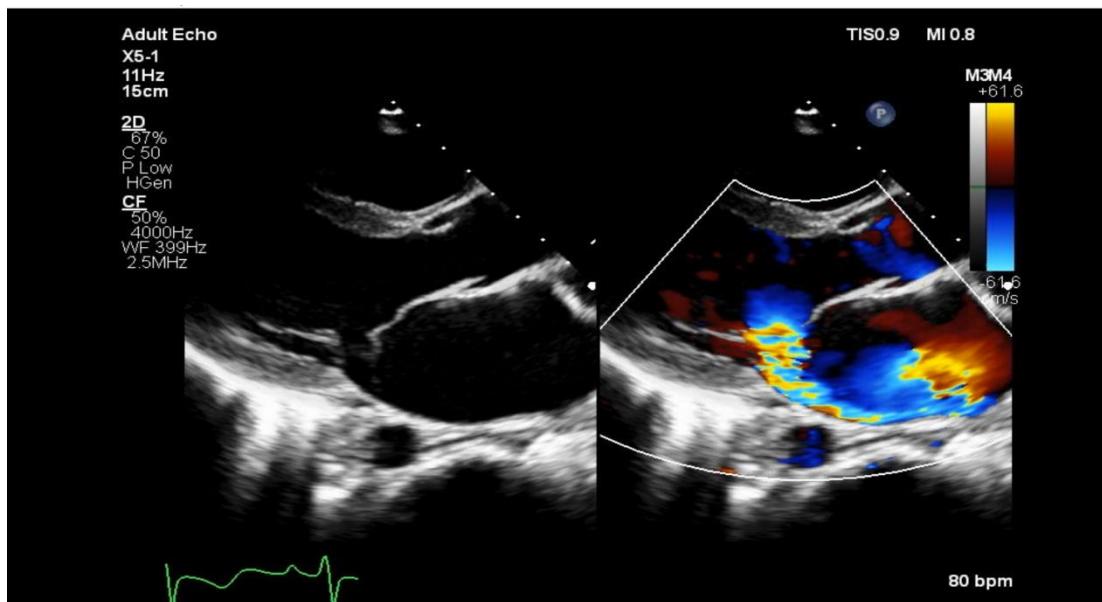
Chest X Ray



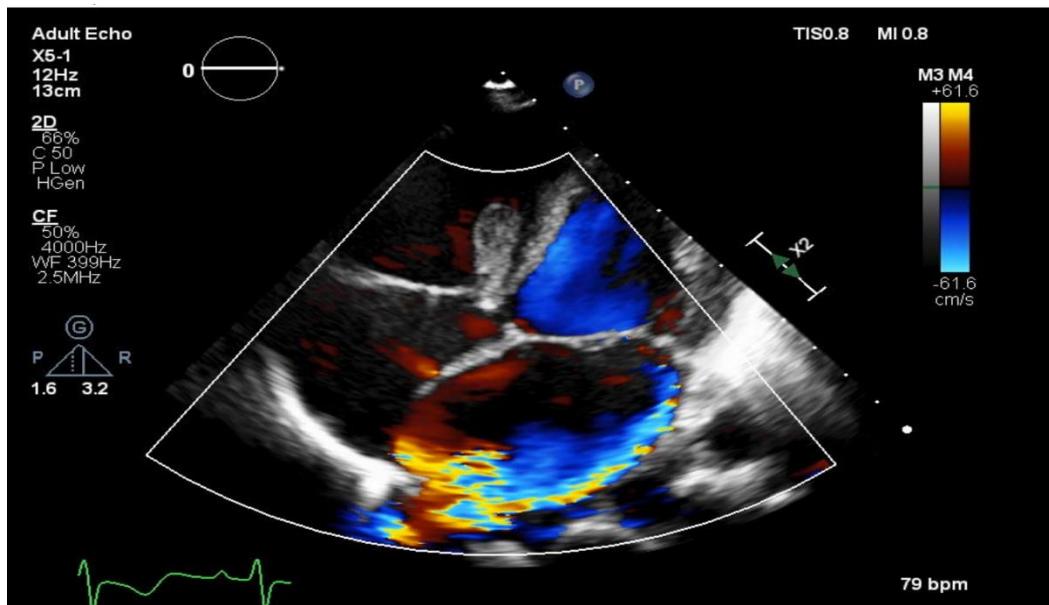
Bulky mass (vegetation) attached to ventricular side of TV annulus(TTE)



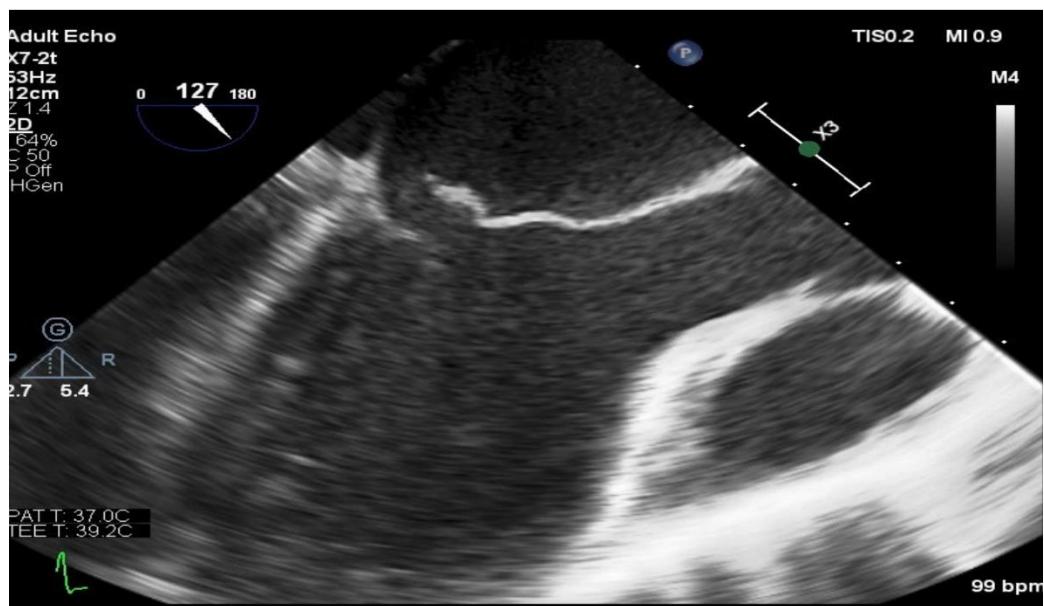
Vegetation by TEE



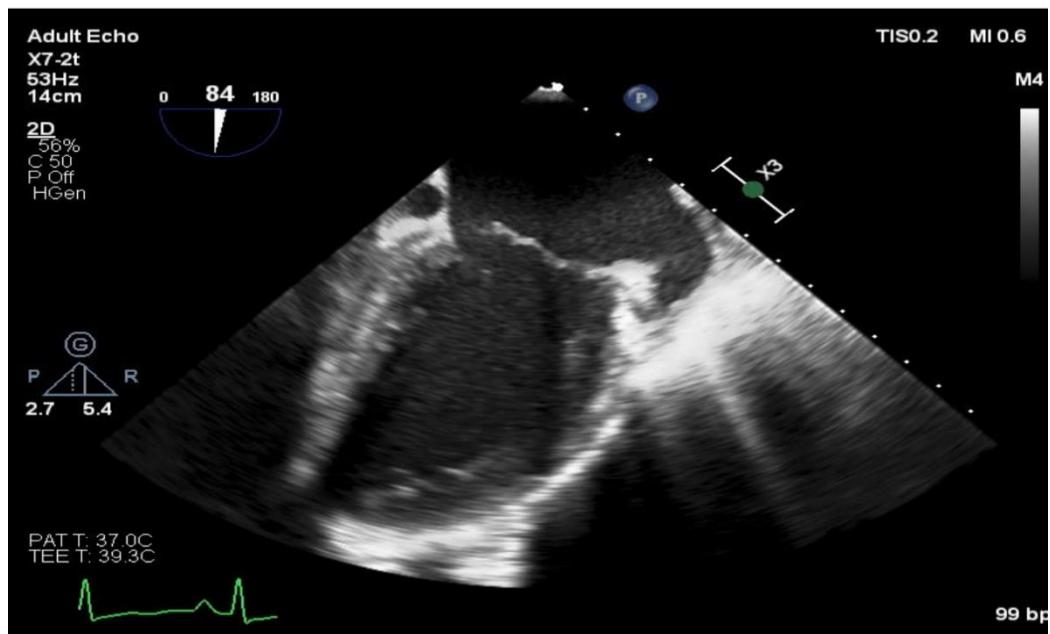
Flail AML with severe MR



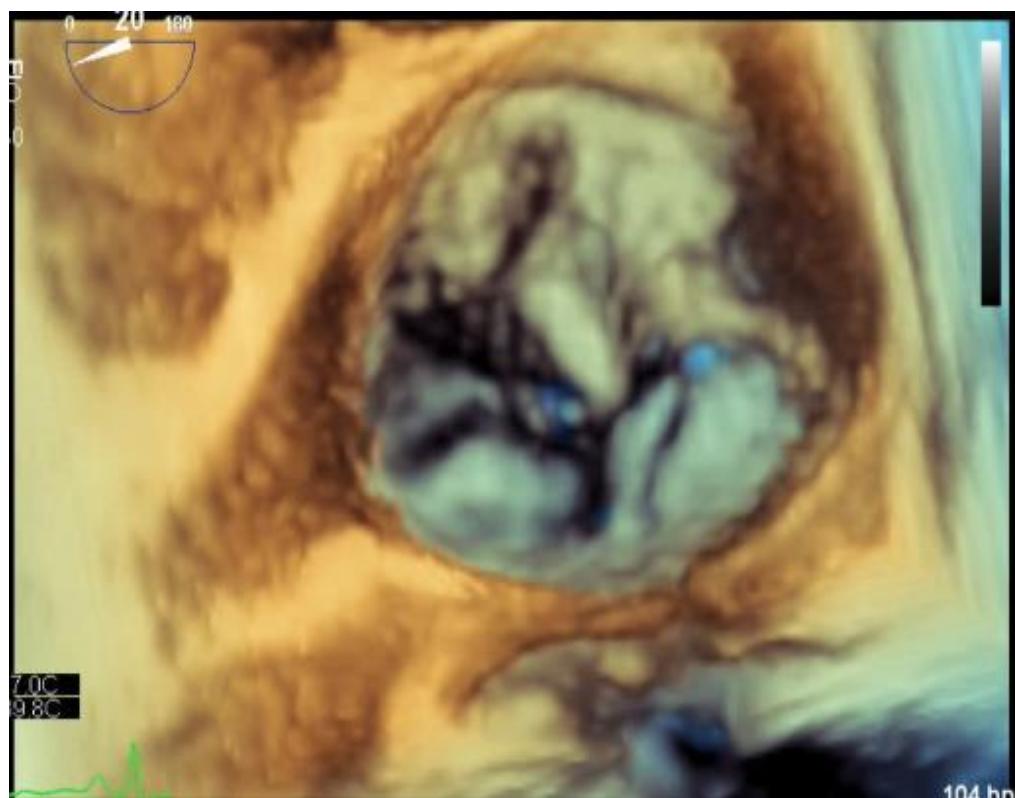
Severe eccentric MR,small ASD



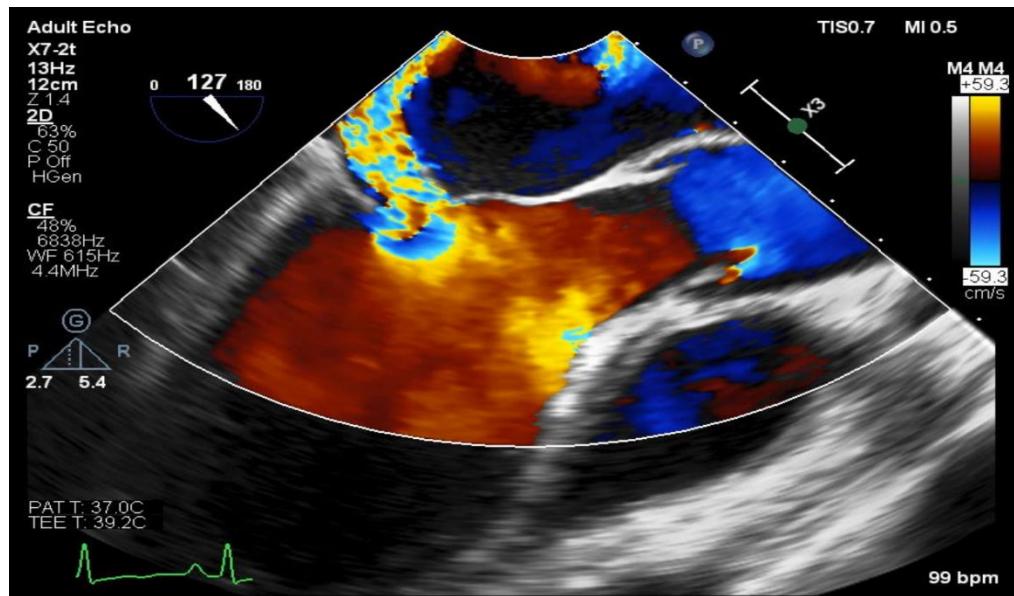
Flail AML (A2 scallop)



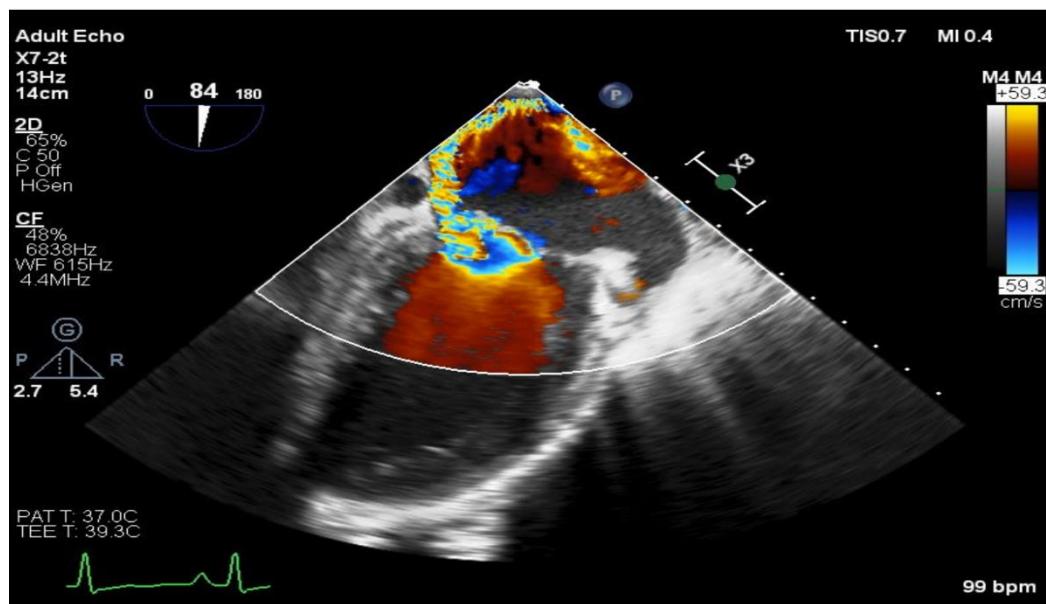
Flail AML (A3 scallop)

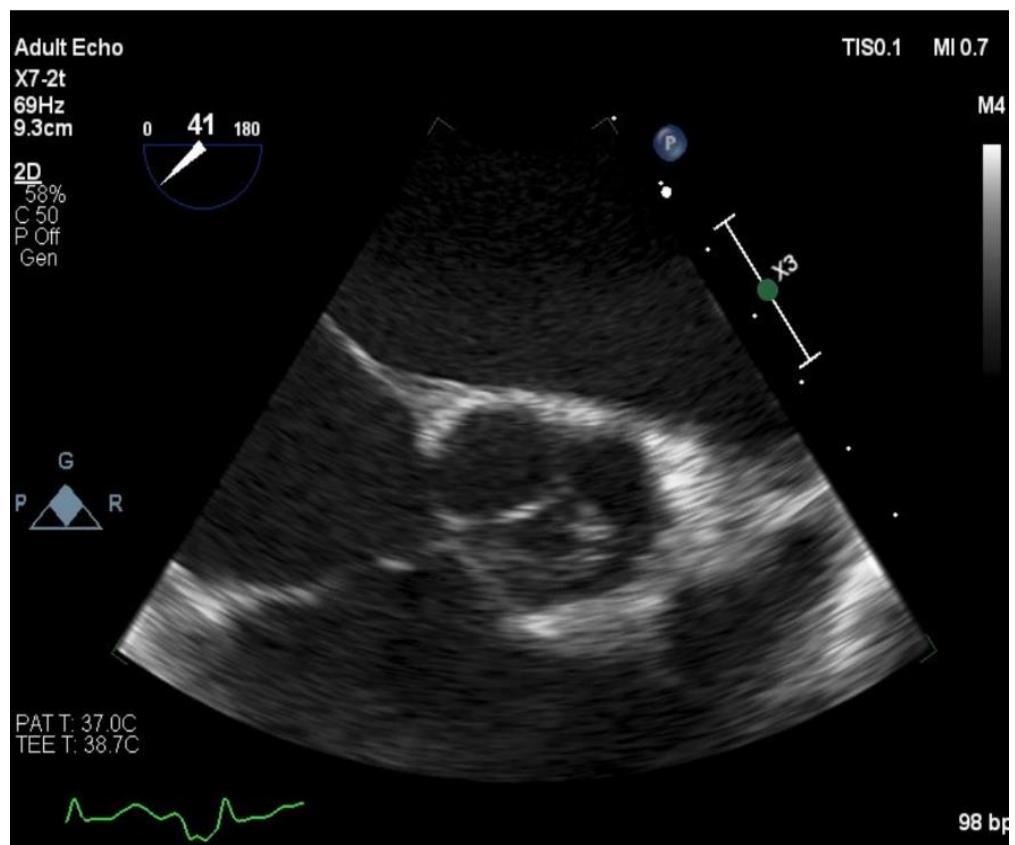


Ruptured A2 and A3 chordi by 3D TEE

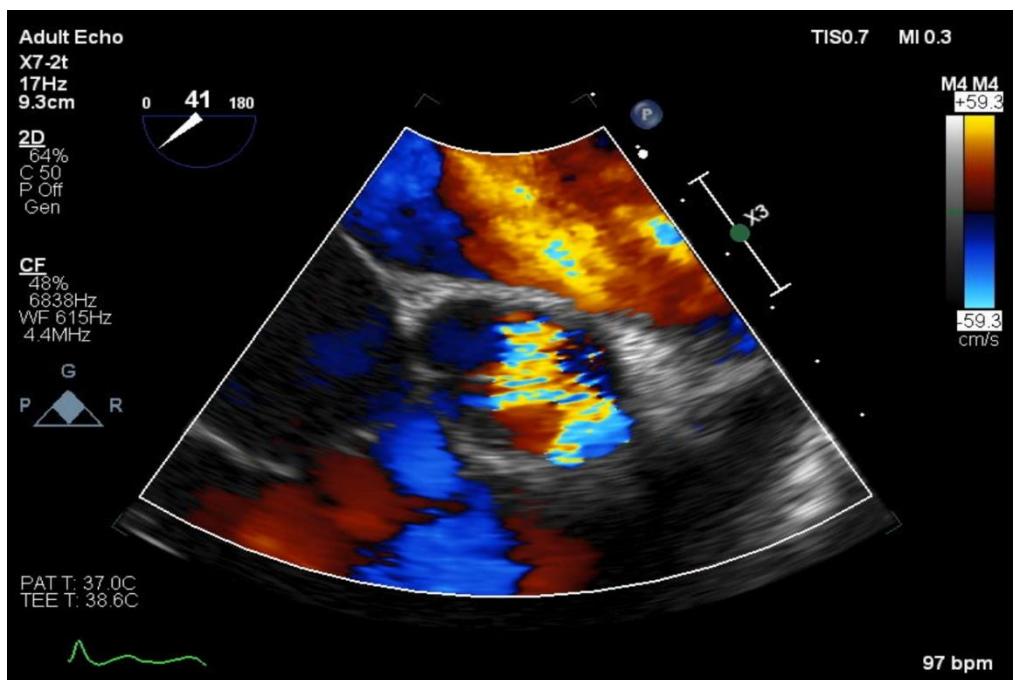


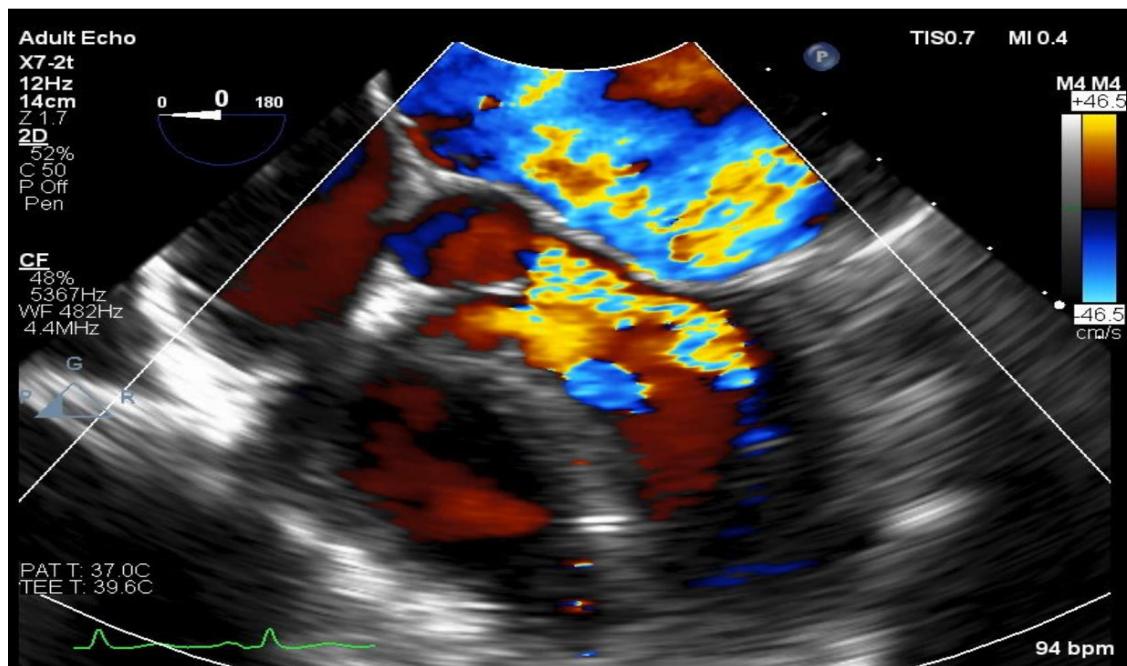
Eccentric severe MR



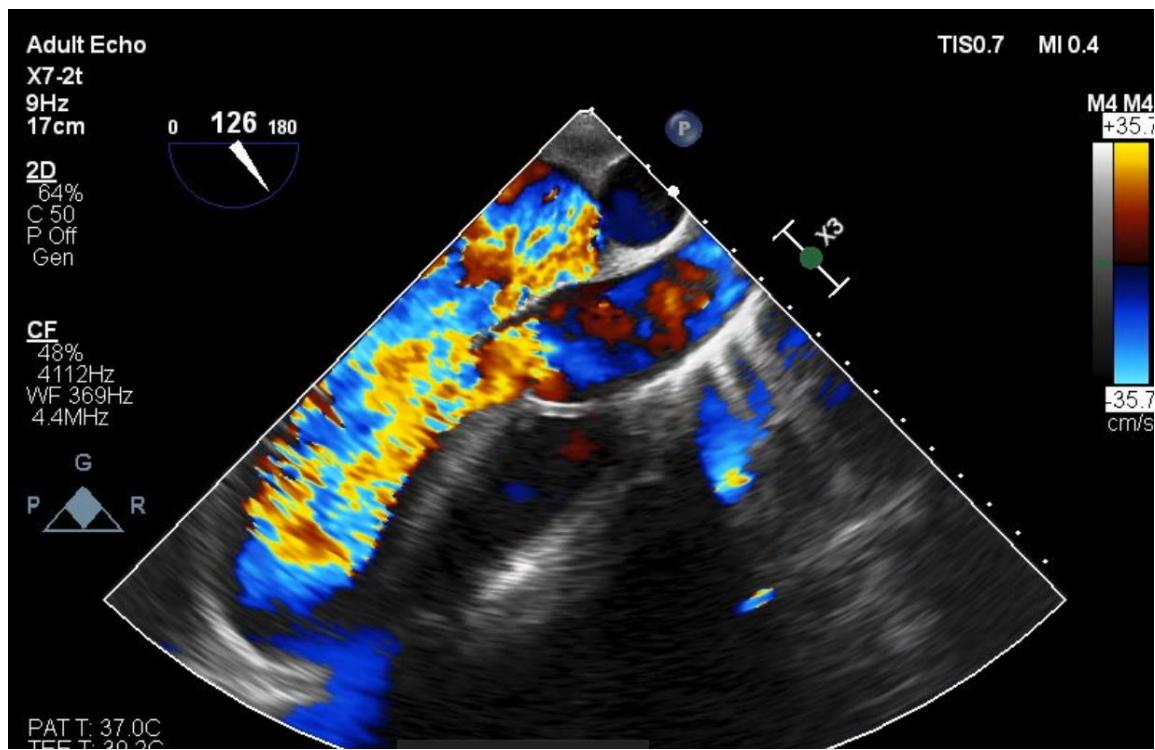


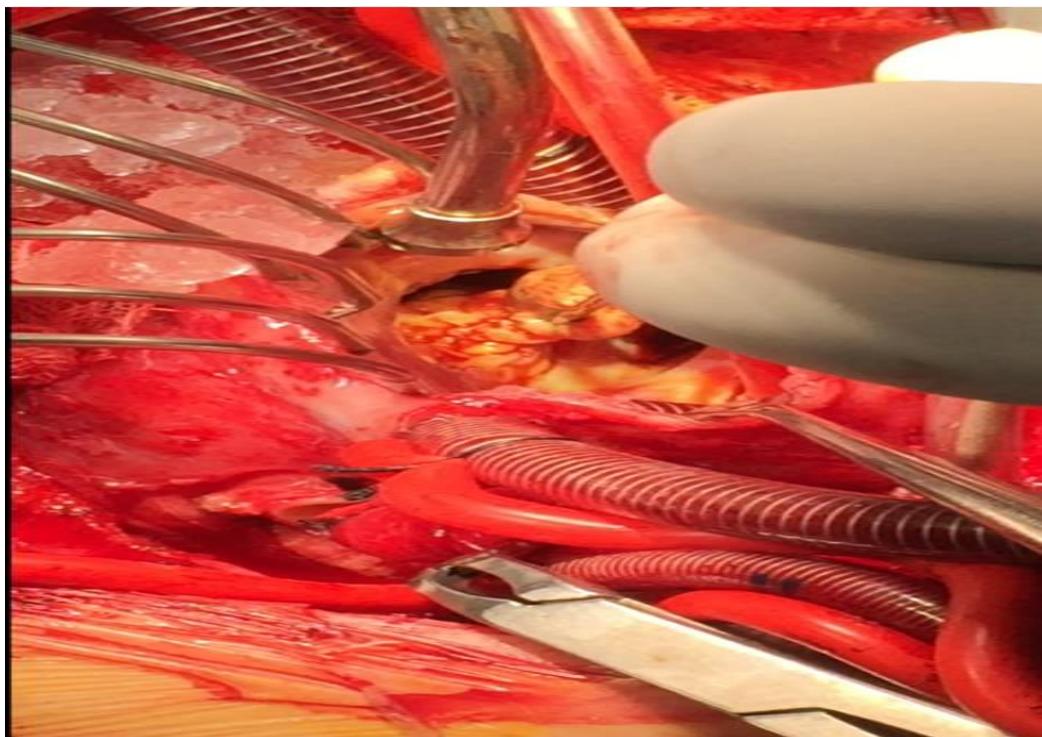
Retraction of Aortic RCC with severe AR



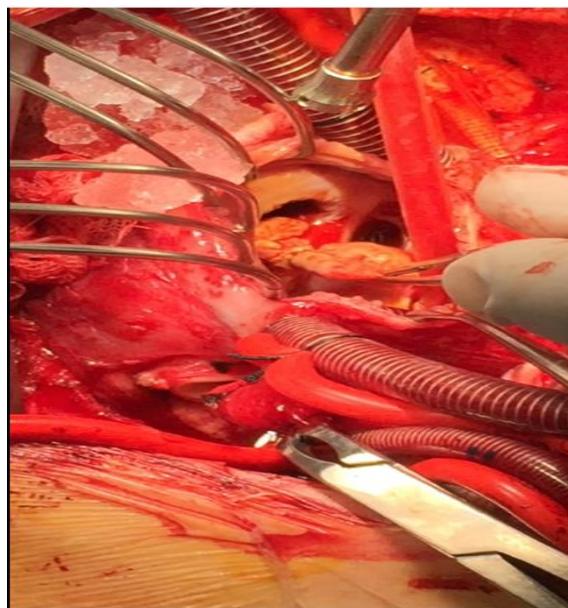


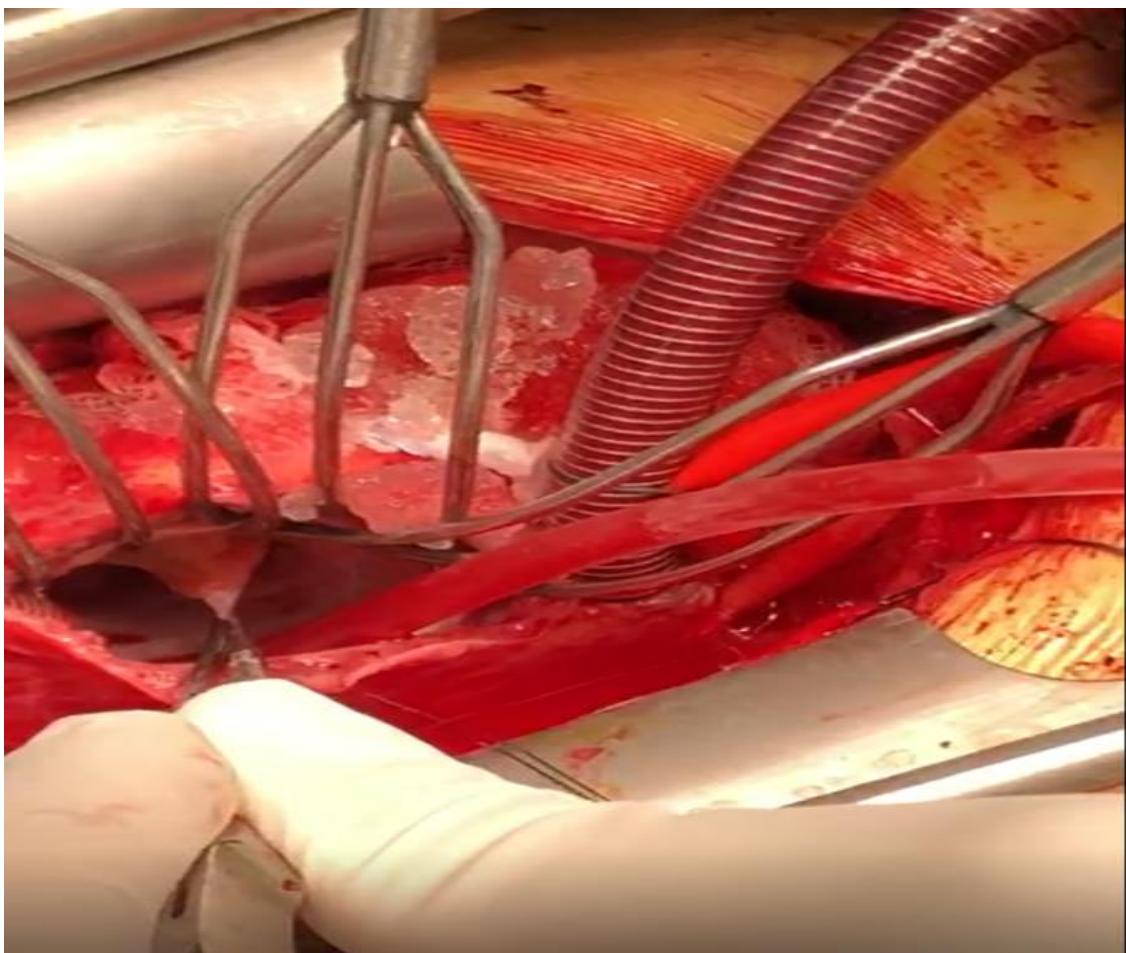
Severe AR





Surgical removal of vegetation





Ruptured A2 chord of MV