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Unusual rheumatic mitral valve stenosis

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R heumatic mitral valve stenosis can be adequately evaluated by two dimensional echocardiography; including the pattern Rof valve involvement and calcification, severity of stenosis, sub-valvular apparatus thickening and associated mitral regurgitation. Suitability of mitral valve for Balloon Mitral Valvuloplasty (BMV) is based on these echocardiographic findings However, three dimensional echocardiography allows en face visualization of the mitral valve and more accurate measurement of the Mitral Valve Area (MVA). A 42-year-old woman presented with gradually progressive dyspnea II-III 2 months before with no orthopnea nor paroxysmal nocturnal dyspnea. She has background of rheumatic heart disease since childhood however, she was not compliant on penicillin therapy. She was referred to our clinic for trans-esophageal echocardiography consultation before considering BMV. On examination: The patient was dyspneic with a heart rate of 100 beats/min and blood pressure of 110/70 mmHg. Loud S1 and normal S2 heart sounds were present on auscultation. An electrocardiogram was performed, which showed normal sinus rhythm. Two dimensional transthoracic echocardiography (2D TTE) revealed thickened mitral valve leaflets, the posterior leaflet is fixed while the anterior one is pliable, no calcifications could be seen. Mildly thickened subvalvular apparatus was present. Both commissures are closed but non calcific. There was mild central valvular regurgitant jet. Mean diastolic gradient was 8mmHg at resting heart rate of 110 beats/min. MVA was calculated using QLAB software (Philips IE33, QLAB 10.4), it was estimated as 1.2 cm². Willkins score was calculated as 7/16. Other heart valves were normal except for mild tricuspid valve regurgitation. Pulmonary artery systolic pressure was 45 mmHg. Normal LV systolic function was present. By 2D trans-esophageal echocardiography (2D TEE), left atrial thrombus was ruled out. Mild mitral valve regurgitation was confirmed. Upon three dimensional trans-esophageal echocardiography (3D TEE); surprisingly an abnormal cleft bisecting anterior leaflet at A2 scallop was clearly seen. The cleft was extending from the free edge of the anterior leaflet and reaching the annulus. It was directed towards Left Ventricular Outflow Tract (LVOT) not towards the Inter-Ventricular Septum (IVS). Moreover, the inter-atrial septum showed multiple fenestrations at the site of fossa ovalis, these small holes were clearly seen from both right and left atrial persepectives. Upon color Doppler, left to right flow was seen through the holes. So, we present an unusual case of rheumatic mitral valve disease, was assessed as severe stenosis with favorable score for BMV and abnormally central insignificant valve regurgitation. However, 3D TEE revealed an abnormal cleft anterior leaflet and Patent Foramen Ovale (PFO) with left to right flow. Now, our patient is not suitable for BMV and she was referred for surgery. Upon reviewing the literature, the association of mitral valve stenosis and left to right shunt at the atrial level is named as Lutembacher syndrome. In addition to these findings, our patient had an isolated cleft anterior leaflet of the mitral valve, so we have a novel association between mitral valve stenosis, PFO with left to right shunt and cleft anterior leaflet of the mitral valve.

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