

Posterior Mitral Valve Leaflet Hypoplasia: An Unusual Presentation of Mitral Regurgitation

Background:

Posterior mitral valve leaflet (PMVL) hypoplasia or agenesis is an extremely rare congenital abnormality.

Case:

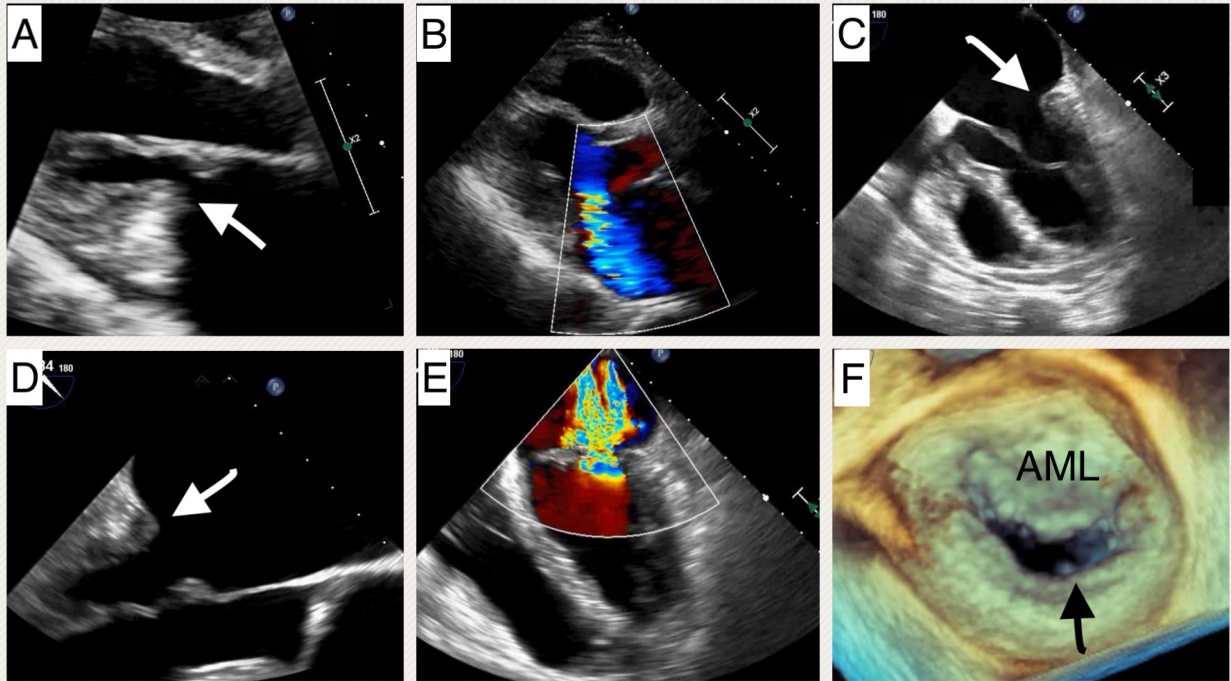
72 year old male with diastolic heart failure, permanent atrial fibrillation, chronic kidney disease, COPD and mitral regurgitation (MR) was referred to our clinic for undergoing evaluation for percutaneous mitral valve repair. Transthoracic echocardiogram demonstrated an eccentric posteriorly directed MR and normal ejection fraction. Following this a transesophageal workup was performed that demonstrated a large anterior mitral leaflet and absent PMVL and hypoplastic posteromedial papillary muscle. The anterior mitral leaflet had degenerative changes with prolapse of the A2-A3 scallops and severe posteriorly directed MR.

Discussion:

PMVL hypoplasia is a rare congenital abnormality of the mitral valve and its prevalence is estimated to be 1:8800. It is usually detected in early life, however, in our patient this was not detected until the seventh decade likely due to lack of symptoms from MR. The patient had a large anterior mitral leaflet that likely had good coaptation with the posterior rim of the mitral annulus and therefore had minimal regurgitation. Progressively with age and degenerative changes, the patient developed severe mitral regurgitation. Transesophageal echo with multiplanar 3D imaging helped identify this rare congenital abnormality. This anatomy is unsuitable for percutaneous mitral valve repair and therefore surgery was recommended.

Conclusion:

PMVL is a rare congenital abnormality and results in severe MR and requires careful evaluation.



- A. 2D transthoracic echocardiography image - parasternal long axis - White Arrow indicates the hypoplastic / absent PMVL.
- B. 2D transthoracic echocardiography image – parasternal long axis – showing a posteriorly directed jet.
- C. 2D transesophageal echocardiography image – four chamber – white arrow indicating hypoplastic / absent PMVL.
- D. 2D transesophageal echocardiography image – white arrow indicating hypoplastic / absent PMVL.
- E. 2D transesophageal echocardiography image – four chamber – showing the severe, eccentric mitral regurgitation.
- F. 3D transesophageal echocardiography image – Mitral valve – black arrow indicating hypoplastic / absent PMVL.