

An Isolated Anterior Mitral Leaflet Cleft: Two Case Reports

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March 07, 2024

Abstract

The isolated anterior mitral leaflet cleft (AMLC) is a very rare anomaly. Echocardiographic imaging is the mainstay diagnostic tool. The isolated AMLC is a surgically correctable anomaly. Valve repair should be preferred over valve replacement in suitable patients.

INTRODUCTION

The anterior mitral leaflet cleft (AMLC) is a rare congenital anomaly and is often associated with other congenital heart diseases. It may be seen with ostium primum atrial septal defect, ventricular septal defect, tetralogy of Fallot, transposition of the great arteries, double outlet right ventricle, tricuspid atresia and double inlet left ventricle. The isolated AMLC is a very rare anomaly. Echocardiographic imaging is the mainstay diagnostic tool for demonstrating the cleft, identifying other congenital heart anomalies, and determining the severity of mitral regurgitation. Mitral cleft is a surgically correctable anomaly. Valve repair should be preferred over valve replacement in suitable patients. (1-3)

In this case report, two cases with isolated AMLC are presented with their echocardiographic diagnosis and follow-up.

CASE PRESENTATION

A 28-year-old female patient was admitted to the cardiology outpatient clinic with the complaint of shortness of breath for two years. Her medical history was not significant. Cardiac auscultation showed a 3/6 grade holosystolic murmur on the left sternal border. Her electrocardiogram (ECG) revealed no significant abnormalities. The transthoracic echocardiography (TTE) had revealed severe mitral regurgitation from the anterior leaflet, left atrial enlargement, mild tricuspid insufficiency, a normal systolic function (LVEF %60), an intact interatrial septum, and elevated systolic pulmonary artery pressure (33 mmHg). The transesophageal echocardiography (TEE) showed a cleft that was between the mitral valve A1 and A2 scallops, and severe regurgitation flow was detected from the cleft region and mild regurgitation flow was detected from the coaptation line. Systolic inversion of pulmonary venous flow was also found. (Video 1, Video 2) The patient was evaluated by the heart team and underwent mitral valve repair with directed cleft suture. Postoperative TTE revealed very good results of mitral valve repair and a mild pericardial effusion. The patient was discharged.

A 19-year-old female was referred to our outpatient clinic with suspected congenital heart disease. At admission the patient was asymptomatic. Her medical history was not significant. Cardiac auscultation showed a 2/6 grade holosystolic murmur on the mitral area. Her ECG showed normal sinus rhythm. A previous TTE had revealed a moderate mitral regurgitation from the anterior leaflet, a conserved systolic function (LVEF %60), an intact interatrial septum, and normal systolic pulmonary artery pressure. The TEE revealed thickening of the mitral valve A2P2 and A3P3 scallops, a cleft between the A2A3 scallops and

moderate-severe mitral regurgitation from the cleft and moderate mitral regurgitation from the coaptation line. (regurgitant volume 36 mL/beat) (Video 3, 4) The follow-up of the asymptomatic patient continues.

DISCUSSION

AMLC without congenital heart disease is diagnosed very rare. It characterizes by a cleft on the anterior mitral valve leaflet that is not associated with an ostium primum atrial septal defect or atrioventricular septal defect. It is thought to develop as a result of incomplete expression of the endocardial cushion defect. The mitral annulus is usually in a normal position. The prevalence of AMLC is unknown. (4, 5)

In patients with isolated AMLC, if mitral regurgitation is not severe, patients can remain asymptomatic for years and the diagnosis can be made incidentally. Patients with AMLC tend to have more severe regurgitation than patients with posterior mitral cleft. (5)

Echocardiography is the main diagnostic tool in patients with suspected or known congenital mitral valve disease. Echocardiography provides information about valve anatomy and morphology, mitral regurgitation severity, and presence of other congenital heart disease. The diagnosis of mitral cleft using 2D TTE is particularly challenging. It may not be possible in every patient to obtain a high-resolution mitral valve image and to clearly visualize the anterior and posterior leaflets in the parasternal short axis image. 3D TEE imaging provides the most valuable information in the evaluation of mitral apparatus anatomy, determination of the presence of cleft, quantitative evaluation of mitral regurgitation, whether the valve is suitable for repair and the valve can be seen anatomically with “surgeon’s view”. (1, 2, 4-8)

When feasible, surgical repair is the first option of the treatment, it consists of a direct suture of the cleft with or without insertion of a prosthetic ring. (5, 9, 10)

CONCLUSION

Isolated AMLC is rare. Echocardiography is the main diagnostic tool in demonstrating cleft, detecting and grading mitral regurgitation, and evaluating suitability for surgical repair.

Source of Funding

None.

Consent

The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

Conflict of Interest/Disclosure

None.

Data Availability

The data used to support the findings of this case report are included within the article.

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Video Legends:

Video 1: The 3D TEE image showed a cleft that was between the mitral valve A1 and A2 scallops.

Video 2: The TEE image showed severe mitral regurgitation from the cleft region.

Video 3: The 3D TEE image showed a cleft between the A2A3 scallops.

Video 4: The TEE image showed double outlet moderate severe mitral regurgitation from the cleft and the coaptation line.