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Case Report

Isolated congenital cleft mitral valve: a rare cause of mitral regurgitation presented as poor weight gain in children

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ABSTRACT

The anterior mitral leaflet cleft is very rare congenital lesion often encountered in association with other congenital heart defects. The isolated anterior leaflet cleft is a rare anomaly and is usually the cause of mitral valve regurgitation which is correctable. Children with poor feeding exhibit weight loss, growth retardation and sometimes heart failure. Clinically echocardiography is the first investigation of choice for evaluation of mitral valve disease providing useful information about valve anatomy and hemodynamic parameters. A case of 6-month-old male child with chief complaint of poor feeding and difficulty in breathing especially in early morning is being presented in this case report. A pan-systolic murmur of grade 4/6 heard on apex and back side of chest wall. Echocardiography revealed an isolated anterior mitral leaflet cleft producing moderate-severe mitral regurgitation. Isolated cleft of mitral valve is a very rare defect that has to be identified and diagnosed. All children presented with poor weight gain or failure to thrive should be screened with TTE to rule out congenital heart diseases.

Keywords: Anterior mitral leaflet cleft, Mitral valve regurgitation, Heart failure

INTRODUCTION

The anterior mitral leaflet cleft is an unusual congenital lesion first described in 1954.[1] Generally this lesion is encountered in association with other congenital heart defects (viz. ventricular septal defect, tetralogy of Fallot, transposition of great arteries, double-outlet right ventricle, tricuspid atresia and double-inlet left ventricle).[2] The isolated anterior leaflet cleft is quite a rare anomaly in which some anatomic data are specific. Unlike endocardial cushion defect, mitral annulus remains in normal position, cleft pointed towards left ventricular outflow tract and mitral and tricuspid valves similar to normal subjects attached to the interventricular septum at different levels (tricuspid valve junction is lower than mitral valve junction).[1] Mitral cleft is usually the cause of isolated valve regurgitation (MR). This lesion is important because it is often correctable. Cleft suture and eventually annuloplasty are better than valve replacement. Sometimes direct cleft suture is not possible because of the lack of valvular tissues and in such cases glutaraldehyde-treated autologous pericardium may be used. Clefts, defined as slit-like holes or defects are hypothesized as being a result of incomplete expression of an endocardial cushion defect often involves the anterior mitral valve leaflet with a pediatric incidence of 1:1340.[3] This lesion is uncommon in adults and is responsible for 33% of congenital mitral valve regurgitation.[4] Nevertheless, if atrio-ventricular junction is normal and MR is mild, patients may be asymptomatic for years. Cleft is the main determinant of MR, however, annular dilatation and restricted motion of anterior leaflet coexist contributing to MR. Regurgitation degree is a consequence of interactions between leaflets, accessory chordal attachment, papillary muscles, left atrium and left ventricle free wall.[5] Echocardiography is the first-choice technique for evaluation of suspected or known mitral valve congenital abnormalities providing useful

information about valve anatomical and morphological details and the mechanism of MR and its quantitative evaluation. Sometimes, preoperative cleft diagnosis is difficult because of the position, dimensions and shape of the lesion.[6] In such patient's three-dimensional echocardiography (3DE) may be useful. The 3DE enables precise assessment of mitral valve pathology as it provides a structural display in three dimensions from every perspective. While its utility has been extensively documented in acquired mitral valve disease, the data on its incremental value in congenital mitral valve pathology are meager.

CASE REPORT

A case of 6-month-old male child weighing 4.5 kg in between (-2 to -3 Z-Score) and recorded birth weight 2.7 kg was brought to the hospital in critical condition with chief complaint of poor feeding and difficulty in breathing while wakeup in early morning since birth. A pan -systolic murmur of grade 4/6 was heard on apex and also on back side of the chest wall. After carrying out TTE showed the mitral dysplasia, anterior mitral leaflet cleft, and moderate-severe MR with a pan-systolic jet originating centrally and then directed towards lateral wall of the left atrium (jet orifice area 0.35 cm2, jet/left atrium ratio 0.42) figure 1-2. A mild left ventricle enlargement (end-diastolic diameter 41 mm, end-systolic diameter 36 mm) with a conserved systolic function (LVEF) 60%, an intact atrial and ventricular septum and normal systolic pulmonary artery pressure.

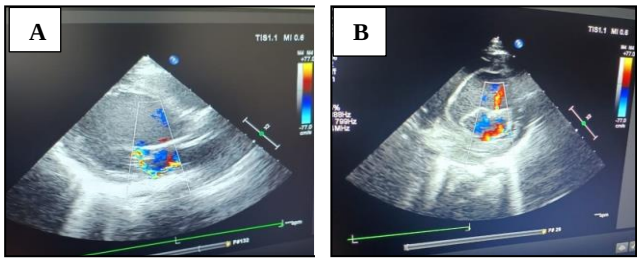

Figure 1 (A and B): PLAX and PSAX view showing centrally origin of mitral regurgitation jet and then directed towards lateral wall of the left atrium.

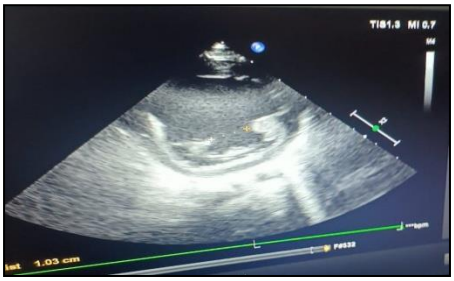

Figure 2: In PSAX view at mitral valve level cleft width in AML was calculated about 10.03 mm.

DISCUSSION

Observation presented in this communication revealed that the echocardiography is a must for the diagnosis of the patients suffering from cardiac complications. The importance of identifying and interpreting a mitral disease justifies the need to perform some echocardiographic evaluations in echo-labs with suitable experience and competence. Information reporting comparisons of echocardiography with any other imaging technique in the evaluation of isolated mitral cleft are rare.

Cardiac magnetic resonance (CMR) has the potential role of correctly identifying this congenital lesion. Nevertheless, CMR is not extensively available and its diagnostic utilization is not justifiable because of its cost. It is reported that the prognosis of repaired mitral cleft is usually excellent with reoperation rate of ~ 3.1% and with a significant survival improvement.[7] Wherever technically feasible mitral repair is preferable to valve replacement.[1] At the same time morbidity and late mortality is shown to be not statistically different with an overall survival probability of 67±7% at 5 years after repair versus 73±9% after replacement.[7]

Real-time 3DE (RT3DE), both TTE and TEE imaging can be highly sensitive in diagnosis of cleft valves providing accurate pathoanatomic definition including width and depth of the cleft, degree of fibrosis and edge retraction, presence of accessory chordate and origin and mechanism of regurgitant jet in addition to characterizing associated congenital malformations. Furthermore, RT3DE imaging also allows visualization of mitral valve en face either from the left atrium or left ventricle and provides a view of the valve similar to that seen intra-operatively by the cardiac surgeon.[7]

TEE is the best technique to characterize mechanism and morphological details of mitral cleft. Assessment of MR by TEE appears to correlate closely with the angiographic degree of mitral regurgitation. Satisfactory results have been obtained in 93% of surgical mitral repair performed on the basis of transesophageal evaluation.[6]

TEE showing the immediate results of repaired valve and the possible mechanism of suboptimal repair is very useful in intra-operative monitoring of repaired mitral valve while TTE is mandatory during follow-up for the evaluation of the repaired valve providing residual functional and anatomical data.

CONCLUSION

Isolated cleft of mitral valve is a very rare defect that has to be known in order to be diagnosed. All children presented with poor weight gain or failure to thrive should be screened with TTE to rule out congenital heart diseases.

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