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Case Report

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Isolated Cleft of the Posterior Mitral Leaflet in 8 Year Old Patient

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Introduction

Cleft of the mitral valve is a rare cause of the congenital mitral regurgitation. Isolated cleft is uncommon finding and is more often found associated with some form of endocardial cushion defect. Cleft of the septal leaflet is found much more often than the defect in the posterior leaflet. Mitral clefts can extend to the mitral annulus or they can be less "deep" and show only minor regurgitation. Clinical importance of the isolated cleft of the mitral valve (ICMV) is that it can be successfully corrected by surgery and that it can have good long-term results.

It is found that ICMV is dominantly prevalent in females. Color Doppler echocardiography should be able to diagnose the ICMV and to distinguish it from the mitral valve defect associated with atrioventricular septal defect. Transesophageal echocardiography is an useful tool for the assesment of congenital mitral regurgitation and the actual width of the cleft as well as the regurgitational jet can be obtained this way [1-3]. Di Signi et.al describe left axis deviation on ECG in one third of the patients with ICMV. (Di Signi: Isolated Cleft Mitral Valve) [4].

Patients with mitral valve cleft associated with another cardiac anomalies usually present earlier than those with ICMV [3].

Medical history in severe mitral regurgitation can reveal dyspnea within exertion, chest pain, failure to thrive and frequent infections of the respiratory tract.

Operative treatment of ICMV associated with mitral regurgitation can usually be carried out without the neccessity for mitral valve replacement. Operative suturing of the isolated cleft is a safe and efficacous procedure without midterm morbidity and mortality. Sometimes the lack of valve tissue that is a result of the retraction of the cleft's edges can be challenging for surgeon. It is previously reported that some surgical corrections of the ICMV require an additional pericardial patch to achieve a good cleft suture [5].

Case Presentation

In our case we present the 8-year-old female patient that was admitted to our hospital for the planned operative treatment due to mitral regurgitation. Medical history revealed dyspnea with exertion as well as sporadical pain in the precordial region. First symptoms seemingly occured at the age of 5. Diagnosis of moderately severe mitral regurgitation was brought 2 years before the surgical treatment and our patient was on a regular cardiologic follow-up with prescribed ACE inhibitors and diuretics. Physical exam showed asthenic body type and systolic ejection click in the precordial projection of mitral valve. Preoperative echocardiography (Figure 1) showed enlargement of the left heart cavities, predominantly left atrium (dimensions 28x38x23 mm): Anulus of the mitral valve was cca 25 mm in diameter and the prolapse of the posterior leaflet was present. Significant mitral regurgitation with two regurgitational jets was noted. The first regurgitational jet sank along the interatrial septum IAS towards the entrance to pulmonary vein inflow tract, but without reverse flow in pulmonary veins. The second central jet reached 2/3 of the LA. According to the analysis of the first regurgitational jet, only moderately severe regurgitation could be described, but summation of the two mentioned jets directed us towards the diagnosis of severe MR.

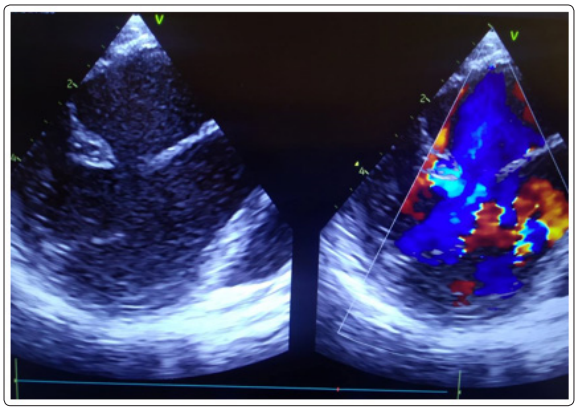

Figure 1: Preoperative echocardiography

Preoperative X ray as well as the ECG showed no relevant abnormalities. Intraoperatively, dilatation of the left heart cavities was confirmed. Mitral valve was found to be dysmorphic. We found cleft of the posterior mitral leaflet between P2 and P3 segment. Edges of the cleft were extremelly thickened. Posteromedial comissure presented as dysmorphic and thinned. The mitral valve was repaired by direct suturing of the cleft and by placing one supportive paracommissural suture at the posteromedial

commissure. The repaired mitral valve was tested with water and it presented as competent with minimal residual regurgitation. Comparision of the transesophageal echocardiogram before and after the repair showed significantly reduced mitral regurgitation: from severe to practically minimal. Postoperative course went without any complications. The girl was dismissed home on the sixth postoperative day.

Discussion

ICMV happens to be an uncommon cause of mitral regurgitation. However, in congenital mitral regurgitation it is very well recognised [6]. Our presented case had the less common site of ICMV: the posterior mitral leaflet. At the age of 8, she already had dilated left heart cavities and was without any doubt significantly symptomatic. More optimal treatment would definitely require an earlier surgical procedure. The girl was followed up in the previous period as a moderate regurgitation and when mitral regurgitation is mild to moderate, surgical repair is not urgent [2]. Surgery for ICMV should be done early, because the results are good in general and the risk of future cardiac dysfunction is lessened [2,3]. Our expectations at the moment of admission were directed towards the future postoperative reduction of cardiac symptoms and towards the relief of the dilated left atrium as well as the prevention of pulmonary hypertension advancement. We were also ready to change the course of the operation if there would be need for mitral valve replacement.

Preoperative transthoracic echocardiography showed moderate to severe mitral regurgitation but couldn't describe the exact cause of valve insufficiency. Preoperatively it seemed that the regurgitation was probably due to the extreme posterior leaflet prolapse. Nevertheless, transesophageal echocardiograhy (TEE) showed severe mitral regurgitation and intraoperatively we found dysmorphic valve with ICMV. Intraoperative TEE is useful for diagnosing mitral regurgitation due to a cleft between P2 and P3 with P2 prolapse [7]. Luckily it was possible to repair the valve with reasonably short cross-clamp time and without the need for mitral valve replacement. However, it is possible that our patient is going to be a valve replacement candidate in the adulthood due to extremely dysmorphic mitral valve and the chance of future worsening of now minimal residual regurgitation.

Conclusion

ICMV is well recognised as a cause of regurgitation in congenital mitral disease and as such should be evaluated promptly. It is important that surgery for congenital mitral regurgitation can be performed in early childhood because it correlates with better prognosis and lessens the risk of possible cardiac dysfunction in the future.

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