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CASE REPORT

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Isolated cleft of the posterior mitral valve leaflet with mitral valve prolapse

Hadi A*, Fkhar S, Soukrat S, Elkarimi S and Elhattaoui M

Department of Cardiology, University Hospital of Mohamed VI-Marrakech, Morocco

Abstract

Isolated cleft of posterior mitral valve leaflet is a very rare cause of congenital mitral regurgitation. We present a 56-year-old woman referred for an echocardiogram by her physician for evaluation of a cardiac murmur. The echocardiogram showed normal left ventricular sizes and function; an isolated cleft of the posterior mitral valve leaflet with posterior leaflet prolapse causing severe mitral regurgitation. The patient was treated surgically with excellent outcome.

Keywords: *echocardiogram, cardiac murmur, congenital mitral regurgitation*

Introduction

Cleft mitral valve leaflet is an uncommon congenital cause of mitral regurgitation. Mitral valve insufficiency in congenital heart disease occurs most frequently in patients with partial or complete atrioventricular septal defects due to a cleft in the anterior leaflet of the mitral valve. Several series of patients with isolated anterior cleft have been studied, especially in pediatric populations [1,2] but isolated clefts in the posterior mitral valve (IPLCMV) was published only in a few earlier case reports. Clefts are slit-like holes or defects of the leaflet, hypothesized to be a result of incomplete expression of an endocardial cushion defect, which most commonly involves the anterior mitral valve leaflet. We report the case of a 56-year-old woman with an isolated midline cleft of the posterior mitral leaflet associated with posterior leaflet prolapse.

Case report

A 56-year-old woman was referred by her primary physician for evaluation of a cardiac murmur. She was presented with dyspea II (NYHA) and chronic palpitations. There was no family history of Marfan syndrome or tall stature. Clinical examination revealed a woman with a normal length and weight. She had not evident sign of elasticity. Cardiovascular examination revealed normal pulses, blood pressure, and jugular venous pressure. The apex was not displaced. Auscultation revealed an apical grade 5 over 6 high frequency holosystolic murmur irradiating to the axilla. Heart sounds were irregular, no third heart sound. Rest of the examination was normal. The electrocardiogram showed atrial fibrillation with left ventricular hypertrophy without ST segment or T wave changes.

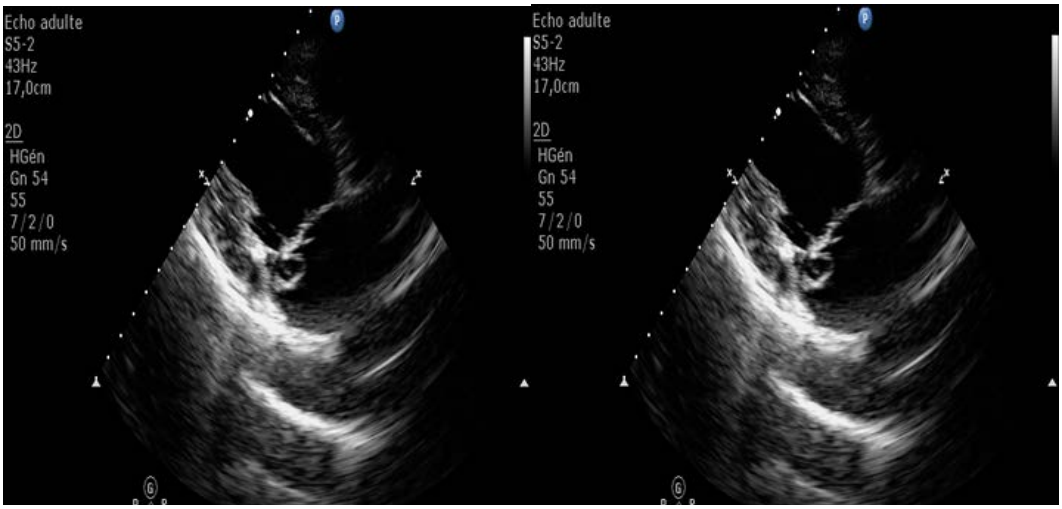

Figure 1. *Parasternal long axis view showing a cleft of posterior leaflet.*

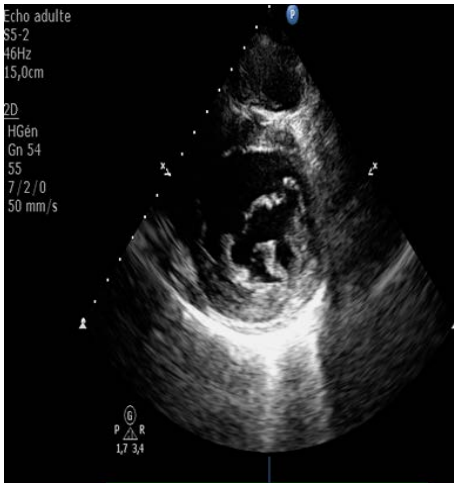

Figure 2. *Parasternal long axis view showing P2 prolapse of mitral valve P2.*

Figure 3. *Parasternal short axis view at mitral level showing a cleft of mitral valve with a 'V' shaped posterior cleft.*

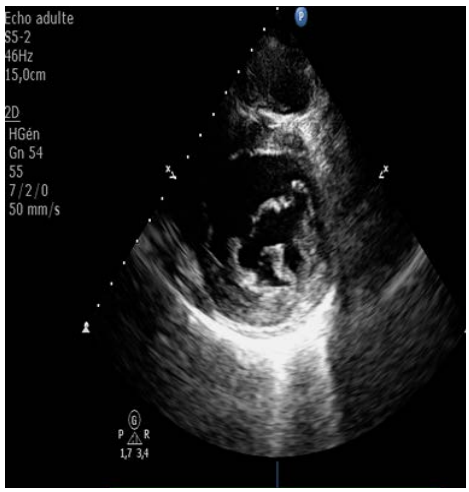

Figure 4. *Paarasternal short axis view,Trileaflet apparence of cleft mitral mitral.*

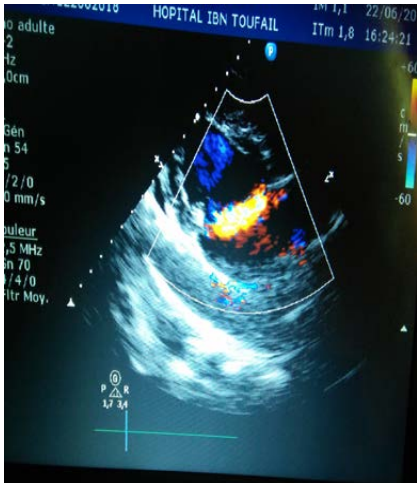

Figure 5. *Parasternal long axis view showing a severe eccentric mitral regurgitant jet through the posterior mitral leaflet.*

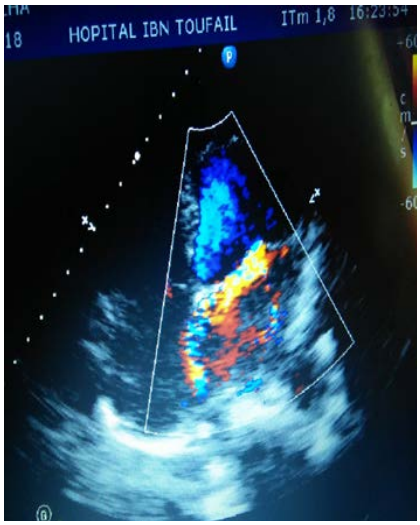

Figure 6. Apical four chamber view showing a severe eccentric mitral regurgitant jet through the posterior mitral leaflet.

Transthoracic echocardiographic examination revealed a situs solitus, laevocardia and concordant atrio-ventricular and ventriculo-arterial connections. There was severe mitral regurgitation with normal systolic left ventricular function and dilated LV and left atrial chambers evidenced by normal shortening fraction, ejection fraction and Surface of regurgitation orifice=100 mm^2. There was prolapse of P2 and cleft of posterior mitral leaflet in the parasternal long axis image (Figures 1 and 2). From the parasternal short axis images, a cleft in the middle of the posterior leaflet (P2) was identified (Figures 3 and 4). The posterior leaflet was bisected by the midline cleft in two equal parts giving the mitral valve appearance of a trileaflet structure (Figure 4). Color Doppler examination revealed a single jet of mitral regurgitation arising from the midline cleft extending postero-superiorly (Figures 5 and 6). The number and location of the papillary muscles and chordal structures were normal. There was no dilatation of the aortic root or the ascending aorta. There were no other cardiac anomalies known to accompany a mitral cleft such as an atrioventricular septal defect. Quadrangular resection of the posterior leaflet and ring annuloplasty was performed successfully in our patient

Discussion

Cleft of the posterior mitral valve leaflet is an extremely rare congenital cardiovascular malformation. A few cases have been reported in the literature [3-8]. A retrospective echocardiographic adult patient study led by Christophe` A Wyss [9] identified twenty-two subjects from 19,320 adults with cleft of the posterior mitral leaflet. Of these, 4 patients had associated heart disease. In 13of the 22 patients, the cleft was in segment P2. Only two cases of Marfan phenotype and an isolated posterior mitral cleft are reported [3,4]. Isolated mitral clefts, which occur in the absence of an atrioventricular septal defect, are rare and mostly affect the anterior leaflet. In an echocardiograpic study involving more than 13000 children, only 10 isolated mitral clefts were identified with all involving the anterior leaflet [10]. This entity is distinct from atrioventricular septal defect clefts [11]. Most reports of clefts of the posterior mitral leaflet have been anecdotal. The cleft can be clearly imaged by the two-dimensional echocardiography. Three-dimensional echocardiographic reconstructions can be very useful in anatomical localization of the papillary muscles and chordal attachments. Mitral valve cleft can cause mitral regurgitation and left ventricular outflow tract obstruction in the presence of abnormal chordal attachments [7]. Regurgitation from Cleft mitral can lead to important physiological and anatomical changes within the cardiac system. Regurgitation results from blood flow directly through the cleft itself or from malcoaptation from accessory chordae with or without papillary muscle distortion. Significant chronic mitral regurgitation elevates left atrial filling pressures and leads to chamber enlargement and eccentric left ventricular hypertrophy. An isolated cleft can be sutured without additional resection of the leaflet. However, direct suture of the isolated posterior clefts should be preferred over valve reconstruction [12]. If direct suture is not feasible, mitral valve repair is preferable to mitral valve replacement in adults [13].

Conclusion

The IPLCMV is an extremely rare cause of mitral insufficiency. Early recognition of this rare clinical entity and possible co-existent anomalies can identify the patients who would benefit from surgical intervention before compensatory left ventricular remodeling and contractile dysfunction develop.

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Correspondence: Hadi A, Department of Cardiology, University Hospital of Mohamed VI-Marrakech, Morocco, Tel: +212641524330;
E-mail: abdelalizate@hotmail.fr

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