

Mitral valve cleft associated with secundum atrial septal defect: case report and review of the literature

Cleft della valvola mitrale associato a difetto del setto interatriale tipo ostium secundum: descrizione di un caso e revisione della letteratura

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ABSTRACT: *Mitral valve cleft associated with secundum atrial septal defect: case report and review of the literature. Shi-Min Yuan, A. Shinfeld, Ehud R.*

Mitral valve cleft associated with secundum atrial septal defect (ASD) is uncommon. We report a 39-year-old male patient manifesting symptoms of congestive heart failure 3 months before admission. Echocardiography showed typical mitral valve prolapse and a large ASD of the secundum type. He was diagnosed as severe mitral regurgitation and ASD. At operation, severe mitral valve prolapse with additional degenerative leaflets and a middle-sized cleft in the anterior leaflet were noted. A large ASD of a mixed cen-

tral and inferior vena cava type was found. Mitral valve repair was impossible. The mitral valve was replaced with an ATS prosthesis. The ASD was repaired with a pericardial patch. Three slow arrhythmias, including nodal rhythm, sinus bradycardia and atrial fibrillation, complicated his early postoperative course. The literature of this entity was reviewed, and the etiology of the postoperative slow arrhythmias was discussed.

Keywords: atrial septal defect, secundum, mitral valve cleft, mitral valve replacement, slow arrhythmia.

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Introduction

A cleft anterior mitral valve leaflet is usually an integral part of ostium primum atrial septal defect (ASD) and atrioventricularis communis [1]. The entity of a mitral valve cleft associated with a secundum type of ASD is apparently infrequent [1, 2]. We present a mitral valve cleft in the anterior leaflet associated with a large secundum ASD in an adult patient.

Case Report

A 39-year-old man was admitted to the hospital due to cough and shortness of breath on exertion for 3 months and peripheral edema for one month. A grade 4/6 pansystolic murmur was audible along left sternal border at the third intercostal space, and a grade 3/6 pansystolic murmur at apex radiating to left axilla. An electrocardiogram revealed a sinus rhythm with a heart rate of 99/min, a P-R interval of 256 ms, a QRS duration of 92 ms, an axis deviation of -20° , and an rSr' pattern in V₁ and V₂, and a qRS pattern in V₆, suggestive of first degree atrio-ventricular block and incomplete right bundle branch block. Echocardiography showed the following: diastolic dimension of the left ventricle was 6.1 cm, left atrial diameter was 6.7 cm, right atrium area was

39 cm², and mitral valve prolapse in the anterior and posterior leaflets with a cleft in the anterior leaflet (fig. 1), and an ASD of secundum type extending 5 cm in size. He was diagnosed as severe mitral regurgitation and ASD, and was operated on April 3, 2006.

At operation, severely dilated left ventricle, left and right atria were noted. The pathologic change of the mitral valve was annulus dilation with severely prolapsed and distorted anterior and posterior leaflets as well as a middle-sized cleft in the anterior. Additional degeneration of the mitral valve included myxomatous and fibrosed chordae tendinae, with thickening and creasing of the cusps and fibrosed chordae tendinae. An ASD of a mixed central and inferior vena cava type measuring 5 × 5 cm was found. A thin strand of fibrotic tissue covered the defect with multiple communications as small needle holes on the membrane. The mitral valve was stiff and was poorly mobile. Valve competence under repair that was evaluated by repeated injections of a bolus of saline into the left ventricle during cardiac arrest was unsatisfactory. After a number of vain attempts, the mitral valve was replaced with a 33-mm ATS prosthesis. The ASD was repaired with a pericardial patch. Three slow arrhythmias, including nodal rhythm, sinus bradycardia, and atrial fibrillation with a ventricular response of 60-70/min

complicated his early postoperative course. He was discharged on the 13rd postoperative day. At 1.5-month follow-up, his echocardiography showed the valve prosthesis in the mitral position and the patch repair of ASD (fig. 2), and his heart rhythm and heart rate recovered to normal.

Discussion

Mitral valve cleft associated with secundum ASD is uncommon [1-4]. It has been stated that patients of concurrent mitral valve cleft and secundum ASD with abnormal superior vectors on electrocardiogram are likely to have clefts on the anterior mitral leaflets, and those with normal vectors are likely to have clefts on the posterior [3]. A right bundle branch block with a frontal plane axis greater than 90° correctly identifies the secundum ASD, whereas a left axis greater than -30° indicates an ostium primum defect [4]. It is of interest that an electrocardiogram typical of an ostium primum syndrome, such as prolonged P-R interval, left axis deviation, and counter clock-wise rotation of the frontal QRS loop, can also occur in the secundum type ASD [2].

Among 16 articles that have been reported since 1960, a total of 42 cases were involved [1-3, 5-17]. Four cases in the report by Liebman and Nadas [5] were atrioventricular valve cleft associated with secundum ASD. There were no concurrent mitral and tricuspid clefts. The number of the patients with a mitral cleft was less than 3. The case mentioned by Neville in the discussion of the report by Messmer *et al.* [8] might be repeatedly reported by Piffarré *et al.* [7]. We disagree with Davies *et al.* [14], who put 5 cases of mitral valve defects associated with secundum ASDs reported by Hynes *et al.* [4] and 2 other cases of scalloped mitral leaflets with secundum ASDs reported by Messmer *et al.* [8] into this entity. Obviously, a scalloped mitral leaflet is not a mitral cleft. Mitral valve defect may include mitral valve dysplasia, stenosis, insufficiency, prolapse, and cleft, etc [18]. It's not at all certain that mitral valve defect must be mitral cleft. But mitral cleft should be included in the domain of mitral valve defect.

Based on the literature, patients with this entity aged from 0 to 52 years with a mean of 18.8 years (n=25). Of them, 14 (56%) were younger than 18 years, and 11 (44%) were older than 19 years. Among the 25 cases whose sexes could be traced, 20 (80%) were females, and 5 (20%) were males. The female-to-male ratio was 4:1. Mitral clefts were located in the anterior leaflet in 22, in the posterior leaflet in 16, in both the anterior and posterior leaflets in 1, and cleft location was unspecified in 3 cases. Two clefts in one leaflet occurred in 1 case [10]. Complete cleft was noted in 3 cases [7, 14]. Associated pathological changes were ruptured

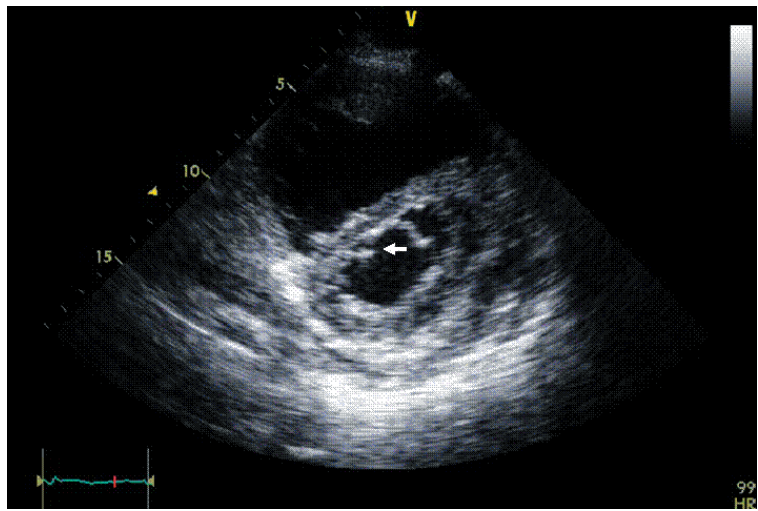


Figure 1. - Parasternal short axis view recorded in diastole showing a cleft (arrow) in the anterior leaflet of the mitral valve, with slight thickening and calcification in the mitral annulus.

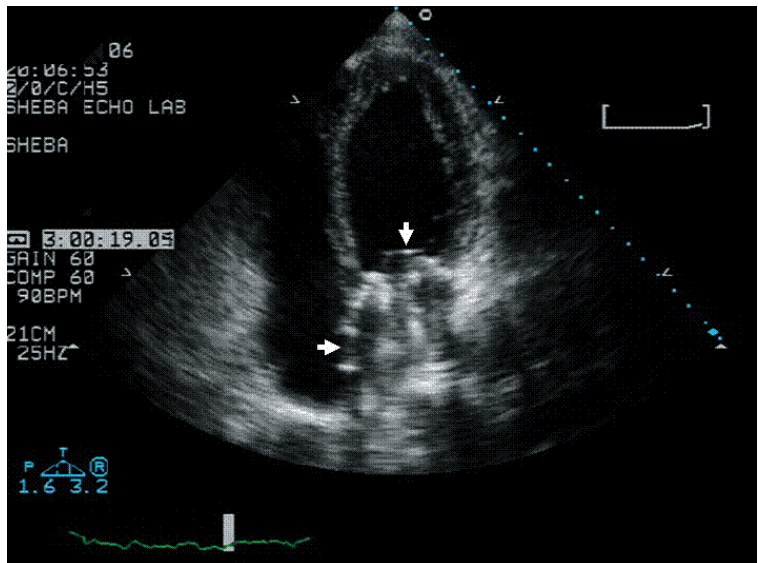


Figure 2. - Four chamber view showing an ATS prosthesis in the mitral position (downward arrow), and the ASD (rightward arrow) repaired with a pericardial patch.

chordae tendinae (n=2) [3, 10], mild myxoid degeneration at the free edges of the cleft (n=1) [14], and elongation of the leaflet and thickening of the chordae tendinae (n=1) [14]. Only one mitral cleft occurred after trauma [12]. Concurrent disorders of this entity included tricuspid cleft [3], trisomy 3p [15], and Down's syndrome and patent ductus arteriosus with or without ventricular septal defect [16]. The secundum ASDs were sutured in 19 (45.2%), repaired with a patch in 4 (9.5%), while closure method was unspecified in 19 cases (45.2%). Cleft repair methods included direct suture (including the case of both leaflet clefts with only anterior cleft repaired) (n=34), suture and annuloplasty (n=1), cleft without repair (including the case of both leaflet clefts with only anterior cleft repaired) (n=4), mitral valve replacement (n=2), and unspecified [5]. The reasons of unrepaired cleft were the cleft connected throughout its entire free margin to a separate papillary muscle by thin chordae [10], the valve was fully competent on testing [10], and clefts were too small [10, 16]. The follow-up was as following: 1 patient (2.4%) died, 1 (2.4%) had worsened mitral

insufficiency due to subacute bacterial endocarditis 5 months after operation [3], and the remainder (95.2%) were uneventful.

The indication of mitral valve replacement for patients with mitral valve regurgitation is rather limited, especially in young patients. Günther and colleagues [19] proposed that mitral valve replacement is reserved for patients with severely dysplastic valves or after failed repair [19]. Abdel-Rahman and associates [20] closed mitral valve replacement in a 16-year-old girl with mitral valve insufficiency and additional leaflet degeneration, when mitral valve repair became impossible. Indications for mitral valve replacement in associated mitral valve cleft and secundum ASD were reported to be ruptured chordae tendinae [11] and mitral endocarditis [9], and severe degeneration and distortion of the leaflet in the present case. This is the third reported patient of this entity who had indication for and underwent mitral valve replacement. Of this entity, majorities of the clefts could be repaired, some small clefts could be left unrepaired, and only a few were underwent mitral valve replacement, when mitral valve repair is impossible.

Postoperative slow arrhythmias after surgical repair of ASD and of mitral valve repair have been stated in the literature. de Salle *et al.* [21] evaluated the frequency and the type of arrhythmias following the surgical correction of 147 cases of ASDs not associated with other congenital heart malformations. The nodal and the coronary sinus rhythms are the most frequently encountered after surgery, attaining 53.8% of all arrhythmias, while atrial fibrillation or flutter represented about 22%. Lancelin *et al.* [22] reported in a series of 300 cases whose ASDs were closed, arrhythmias were found in 60%. These were usually slow supraventricular arrhythmias caused by substitution (51%). Arrhythmias are most common during the first week (56%), and are usually of the slow type. The slow type of arrhythmias occurred in the high ASDs, and the fast type especially amongst elderly patients. The factors influencing arrhythmias were age, cardiac enlargement, and the mean pulmonary arterial pressure. Bolens and Friedli [23] noted electrophysiologic disturbances might result from the ASD itself or from surgery. Closure of ASD improves atrioventricular conduction, decreases atrioventricular nodal refractory periods and improves sinus node function, probably by suppressing right-sided heart volume overload. Kernis *et al.* [24] demonstrated that independent predictors of early atrial fibrillation after mitral valve operations were lower left ventricular ejection fraction, larger left atrial size, mitral valve replacement, and non-ischemic etiology of mitral valve regurgitation. Early atrial fibrillation was more frequent in patients with than without left atrial size ≥ 50 mm. These independent predictors might give explicit explanations to early postoperative slow arrhythmias of the present patient.

Riassunto

Il cleft della valvola mitrale associato al difetto del setto interatriale (DIA) tipo ostium secundum è infrequente. Riportamo un paziente maschio di 39

anni che ha manifestato sintomi di scompenso cardiaco 3 mesi prima del ricovero ospedaliero. L'ecocardiografia ha evidenziato un prollasso valvolare mitralico tipico ed un ampio DIA tipo ostium secundum. Era associata una insufficienza mitralica severa. All'intervento, si riscontrava un prollasso valvolare mitralico severo con lembi valvolari degenerati e un cleft nel tratto medio del lembo anteriore mitralico, ed anche un ampio DIA misto centrale e tipo vena cava inferiore. Essendo impossibile la riparazione della valvola, si è proceduto ad una sostituzione valvolare mitralica con una protesi ATS, e ad una riparazione del DIA con un patch di pericardio. Il decorso postoperatorio precoce è stato complicato da tre episodi di bradiaritmia, uno di ritmo giunzionale, uno di bradicardia sinusale, ed uno di fibrillazione atriale. La letteratura di questa associazione è stata rivista, e l'eziologia delle bradiaritmie postoperatorie è stata discussa.

Parole chiave: difetto del setto interatriale tipo ostium secundum, cleft del lembo anteriore mitralico, sostituzione valvolare mitralica; bradiaritmie.

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