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

Shi-Min Yuan, Amihay Shinfeld, Ehud Raanani

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 central 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.

Monaldi Arch Chest Dis 2007; 68: 48-51.

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 leaflet associated with a large secundum ASD in an adult patient.

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.

Monaldi Arch Chest Dis 2007; 68: 48-51.
complicated his early postoperative course. He was discharged on the 13th 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 Pi-farré 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 chordae tendineae (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 tendineae (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] chose 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 tendineae [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 underwент 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 atioventricular conduction, decreases atioventricular 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.

**References**

1. Salomon J, Aygen M, Levy MJ. Secundum type atrial septal defect with mitral valve disease. Chest 1970; 58: 540-2.
2. Hara M, Char F. Partial cleft of septal mitral leaflet associated with atrial septal defect of the secundum type. Am J Cardiol 1966; 17: 282-5.
3. Snow NJ, Ankeney JL. Congenitally cleft atrioventricular valves associated with secundum atrial septal defects. J Thorac Cardiovasc Surg 1976; 72: 925-8.
4. Hynes KM, Frye RL, Brandenburg RO, McGoon DC, Titus JL, Giuliani ER. Atrial septal defect (secundum) associated with mitral regurgitation. Am J Cardiol 1974; 34: 333-8.
5. Liebman J, Nadas AS. The vectorcardiogram in the differential diagnosis of atrial septal defect in children. Circulation 1966; 22: 956-75.
6. Billig DM, Hallman GL, Bloodwell RD, Cooley DA. Surgical treatment of atrial septal defects in patients with angina pectoris. Ann Thorac Surg 1968; 5: 566-8.
7. Pifarre R, Dieter RA, Hoffman FG, Neville WE. Atrial septum defect and cleft mitral valve. Ann Thorac Surg 1968; 6: 373-6.
8. Messner BJ, Hallman GL, Cooley DA. Congenital mitral insufficiency. Results in unusual lesions. Ann Thorac Surg 1970; 10: 450-61.
9. Cohn LH, Hancock EW, Griep RB, Shumway NE. Congenital cleft mitral valve associated with ostium secundum atrial septal defect. Circulation 1971; 44 (Suppl II): 152.
10. McElroy MT, English TA, Ross DN. The congenitally cleft posterior mitral valve leaflet. An antecedent to mitral regurgitation. Ann Thorac Surg 1973; 16: 281-92.
11. Goodman DJ, Hancock EW. Secundum atrial septal defect associated with a cleft mitral valve. Br Heart J 1973; 35: 1315-20.
12. Murray GF, Wilcox BR. Secundum atrial septal defect and mitral valve incompetence. Ann Thorac Surg 1975; 20: 136-43.
13. Kerin NZ, Edelstein J, Louridas G. Prolapsing mitral valve leaflet syndrome. A spectrum that includes cleft posterior mitral valve. Cathet Cardiovasc Diagn 1976; 2: 77-85.
14. Davies RS, Green DC, Brott WH. Secundum atrial septal defect and cleft mitral valve. Ann Thorac Surg 1977; 24: 28-33.
15. Suzuki M, Ishikawa S, Otaki A, Sakata K, Kawashima O, Otani Y, Morishita Y. Secundum atrial septal defect associated with mitral valve cleft: report of a case with chromosomal syndrome of trisomy 3p. *Surg Today* 1996; 26: 734-6.

16. Fraisse A, Massih TA, Bonnet D, Sidi D, Kachaner J. Cleft of the mitral valve in patients with Down’s syndrome. *Cardiol Young* 2002; 12: 27-31.

17. Kirali K, Mansuroğlu D, Özcan Y, Bozbuğa NU, Tuncer A, Toker ME, Şişmanoğlu M, Yakut C. Mitral clefts and interatrial septum defects: 15-year results. *Asian Cardiovasc Thorac Ann* 2003; 11: 135-8.

18. Mitral valve defect. Available at: [http://www.shilohgtf.com/Mitral_Valve_Defect.htm](http://www.shilohgtf.com/Mitral_Valve_Defect.htm). Accessed on January 30, 2007.

19. Günther T, Mazzitelli D, Schreiber C, Wottke M, Paek SU, Meisner H, Lange R. Mitral-valve replacement in children under 6 years of age. *Eur J Cardiothorac Surg* 2000; 17: 426-30.

20. Abdel-Rahman U, Wimmer-Greinecker G, Matheis G, Klesius A, Seitz U, Hofstetter R, Moritz A. Correction of simple congenital heart defects in infants and children through a minithoracotomy. *Ann Thorac Surg* 2001; 72: 1645-9.

21. de Salle P, Gonen M, Lecron J, Jaumin P, Tremouroux J. Troubles du rythme survenant après fermeture chirurgicale de la communication interauriculaire. *Acta Cardiol* 1975; 30: 239-49.

22. Lancelin B, Crepieux A, Diebold B, Abbou B, Goujon J, Apoil E, Pauly-Laubry C, Maurice P. Les troubles du rythme après fermeture des communications inter-auriculaires. A propos de 300 cas. *Arch Mal Coeur Vaiss* 1977; 70: 1283-91.

23. Bolens M, Friedli B. Sinus node function and conduction system before and after surgery for secundum atrial septal defect: an electrophysiologic study. *Am J Cardiol* 1984; 53: 1415-20.

24. Kernis SJ, Nkomo VT, Messika-Zeitoun D, Gersh BJ, Sundt TM 3rd, Ballman KV, Scott CG, Schaff HV, Enriquez-Sarano M. Atrial fibrillation after surgical correction of mitral regurgitation in sinus rhythm: incidence, outcome, and determinants. *Circulation* 2004; 110: 2320-5.