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A comparison of quality of life in mitral valve replacement and mitral valve repair patients

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ABSTRACT

Objectives: Mitral valve replacement and repair are the surgical treatment methods in mitral valve disease. Although both treatments have some superiorities against each other in some certain fields, their effects on the improvement of the quality of life of patients is another factor that needs to be taken into consideration. In this study, we aimed to investigate the effects of these treatment methods on the improvement of the quality of life.

Patients and methods: Between January 2006 and June 2009, a total of 40 patients (27 females, 13 males; mean age 48.2±14.8 years; range 16 to 74 years) with similar etiologies and EuroSCORE values were included in this prospective, single-center study. All patients were divided into two groups as mitral valve replacement (group 1, n=22) and mitral valve repair (group 2, n=18). The Short Form-36 was used to assess the improvement in the quality of life.

Results: Seven of eight domains including physical function, role limitation due to physical function, social function, role limitation due to emotional problems, energy, mental health, and general health perception in the Short Form-36 were significantly improved in both groups. Pain scores were significantly improved in group 2, whereas no significant improvement was seen in group 1.

Conclusion: Our study results show a significant improvement in the quality of life following both treatments. However, mitral valve repair seems to be superior to mitral valve replacement in terms of pain scores.

Keywords: Mitral valve repair; mitral valve replacement; quality of life.

Mitral valve diseases are very common in both developed and developing countries and considerably affect mortality and morbidity.[1-5] The optimal timing for the surgical treatment of valve disease is before the permanent myocardial damage and serious symptoms occur.[6] Mitral valve diseases include both mitral valve stenosis and regurgitation. In addition, structural valve disease became surgically treatable after the introduction of the cardiopulmonary bypass system. Replacement of the damaged valve was the first choice in surgical treatment, since it is easier to perform and the results are more predictable. However, mitral valve repair (MVr) has become more popular, since mitral valve replacement (MVR) is associated with a high risk of thromboembolism, endocarditis, and left ventricular dysfunction.[7-9] Furthermore, it is necessary to use anticoagulants after MVR. In addition, MVr has a lower risk of thromboembolism and valvular infections, and requires less reoperation.[10] It is more popular thanks to these advantages.[10,11] On the other hand, although repair is more popular today, MVR still has more advantages in certain conditions. Mitral valve replacement is in favor in patients with ischemic valvular disease, as MVr takes more time and, therefore, the duration of cardiopulmonary bypass machine use is longer.[12] Furthermore, MVr needs a great knowledge about the mitral valve morphology and structure, and an adequate experience. Nonetheless, previous studies have shown no significant difference between MVR and MVr in terms of long-time survival and mortality rates in patients with ischemic mitral valve disease.[13,14]

Although there are some new advanced biomaterials designed for valve production, they are still far from being the ideal materials. Neither mechanical nor biological valves are as ideal, as the patient's own valve. Although biological valves function much

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more like the natural valve of the human heart, they still tend to become deteriorated and need a replacement in a shorter time of period, compared to the mechanical valves.[15] Therefore, we can suggest that there is no prosthetic valve which provides an excellent hemodynamic properties without causing early and late mortality risks and not easy to supply and cannot be used by all surgeons.[16] Both of these two techniques have some superiority against each other. The primary goal is to heal the disease and to protect the functional capacity in the treatment of the patients with mitral valve disease. Therefore, one of the major outcomes of the treatment is to improve the quality of life (QoL) in these patients.

In the present study, we aimed to investigate the effects of MVR and MVr on the improvements in the QoL.

PATIENTS AND METHODS

A total of 40 patients (27 females, 13 males; mean age 48.2±14.8 years; range 16 to 74 years) who underwent MVR and MVr at Dr. Siyami Ersek Thoracic and Cardiovascular Training and Research Hospital, Istanbul, Turkey between January 2006 and June 2009 were included in this prospective, single-center study. All patients had a similar etiology, diagnosis, and EuroSCORE values. The Short Form-36 (SF-36), which is a questionnaire with 36 questions concerning the QoL and health care, was used to assess QoL of the patients at baseline and at six months of surgery. Face-to-face interview method was used to make sure that the patient fully understood the questions in the SF-36 scale. All patients were divided into two groups as group 1 (MVR, n=22) and group 2 (MVr, n=18). The patients who underwent only mitral and tricuspid valve interventions were included, and those who underwent double valve replacement or additional procedures (coronary artery bypass grafting or ascending aorta aneurysm repair) were excluded. Data including age and gender, diabetes mellitus, tobacco use, hypertension, cerebrovascular stroke, renal insufficiency, EuroSCORE, and myocardial infarction were recorded. Following parameters were investigated in the preoperative echocardiograms: stenosis (pure stenosis, predominant stenosis, predominant insufficiency, pure insufficiency) and diastolic diameter of the left ventricle, ejection fraction (EF), left atrium diameter, and mean pulmonary artery pressure. Cross-clamp and cardiopulmonary bypass times were also recorded. The mean body surface area was calculated.

The study was approved by the local Ethics Committee and conducted in accordance with the principles of the Declaration of Helsinki.

Surgical technique

A written informed consent was obtained from each patient. All operations were performed by three different surgical teams at a single-center, using the same surgical principles. Median sternotomy was used in both groups. Right atriotomy and transseptal incision were performed in tricuspid annuloplasty patients. The mitral valve was reached via only the left atriotomy in patients who were treated for only mitral valve dysfunction. The valve was excised and Teflon-pledged sutures (2.0 polyester, Ethicon Inc., Somerville, NJ, USA) were used to implant the mitral valve. Different techniques were used to repair the mitral valve such as commissurotomy, quadrangular resection, and sliding annuloplasty, posterior annuloplasty, and cleft repair.

Statistical analysis

Statistical analysis was performed using the Number Cruncher Statistical System (NCSS, LLC, Kaysville, Utah, USA). Descriptive data were expressed in mean + standard deviation. The Student's t-test was used to compare normally distributed variables, while the Mann-Whitney U test was used to compare abnormally distributed variables between the two groups. The Wilcoxon sign test was used to analyze intra-group variables. The Fisher's exact and Fisher-Freeman-Halton tests were carried out to compare qualitative data. A p value of <0.05 was considered statistically significant.

RESULTS

A total of 22 patients underwent MVR, while 18 patients underwent MVr. None of the patients died after the operation until sixth month of the postoperative period. Baseline EF was 55.86% and 60.28%, while baseline EuroSCORE was 2.5 and 2.78 in group 1 and group 2, respectively (Table 1). Mitral valve disease had rheumatic and degenerative etiology in 29 and 11 patients, respectively. Thirty patients were classified as the New York Heart Association (NYHA) Class II, while the remaining 10 patients were classified as the NYHA III. There was no

Table 1
Baseline echocardiography findings and EuroSCORE values

	Group 1 Mean±SD	Group 2 Mean±SD	*p*
Ejection fraction, mean (%)	55.9±8.6	60.3±5.0	0.049
Pulmonary pressure	41.1±10.6	39.3±12.3	0.619
Left atrium size (mm)	48.6±7.6	43.9±9.4	0.085
EuroSCORE	2.5±2.2	2.8±2.2	0.693

SD: Standard deviation.

statistically significant difference in age, gender, mean body surface area, cardiopulmonary bypass and cross-clamp time, pulmonary artery pressure, left atrial volume, EuroSCORE, etiology, NYHA classification, left ventricular internal dimension in systole (LVIDs) enlargement, cerebrovascular disease, myocardial infarction, revision and hypertension between the groups.

When the preoperative findings were evaluated, EF and tobacco use were found to be significantly higher in group 2 ($p<0.05$). Although a higher Class III NHYA classification was found in group 2, it was not statistically significant ($p>0.05$). Severe heart failure was found to be higher in group 1, although it did not reach statistical significance ($p>0.05$). Baseline rhythm was atrial fibrillation in nine patients in group 1 and three patients in group 2 (Table 2).

Table 2
Details of the mitral valve disease according to study groups

	Group 1		Group 2	
	n	%	n	%
Diagnosis				
MVF-moderate	3	13.6	10	55.6
MVF-severe	8	36.4	2	11.1
MS-moderate	5	22.7	4	22.2
MS-severe	3	13.6	1	5.6
MVF+MS	3	13.6	1	5.6
Etiology				
Rheumatic	18	81.8	11	61.1
Degenerative	4	18.2	7	38.9
NYHA Class				
II	19	86.4	11	61.1
III	3	13.6	7	38.9
Cardiac rhythm				
Atrial fibrillation	9	40.9	3	13.6
Sinus	13	59.1	15	83.3
LVIDs				
Increased	3	13.6	0	0
Normal	19	86.4	18	81.8

MVF: Mitral valve failure; MS: Mitral stenosis; NYHA: New York Heart Association; LVIDs: Left ventricular internal dimension in systole.

In addition, seven of eight domains of the SF-36 including physical function, role limitation due to physical function, social function, role limitation due to emotional problems, energy, mental health, and general health perception were significantly improved after the treatment in both groups ($p<0.05$). The increases in these domains were also similar in both groups ($p>0.05$) (Table 3).

There was no significant increase in terms of pain scores in group 1 ($p>0.05$), although pain scores significantly increased in group 2 ($p<0.01$). The increase was not significantly different between two groups ($p>0.05$). There was a significant increase in terms of general health perception scores and energy in both groups ($p<0.01$), while the increase was similar in both groups ($p>0.05$). The social function scores increased in both group 1 ($p<0.05$) and group 2 ($p<0.1$), although the increase was statistically significant in group 2 only. However, there was no significant difference in the social function scores between the two groups.

DISCUSSION

Chronical mitral valve disease is related with significant mortality and morbidity.[17,18] Thus, one of the primary goals of MVR or MVr is to reduce the mortality and morbidity rates. Although both methods have some superiority to each other, there are no strict rules about the order of the method that needs to be selected in a specific type of the disease. In this study, we investigated the effects of both treatment on the QoL of the patients using the SF-36 scale.

Table 3
Results of the Short Form-36 scale

	Group 1		Group 2		
	Mean±SD	Median	Mean±SD	Median	*p*
Physical function					
Preoperative	45.5±25.4	40	34.2±27.4	22.5	0.127
Postoperative	81.1±13.8	80	75.0±27.3	82.5	0.901
p	0.001		0.001		
Role limitation due to physical function					
Preoperative	22.7±40.8	0	15.3±35.5	0	0.445
Postoperative	78.4±38.8	100	72.2±41.9	100	0.844
p	0.002		0.001		
Pain					
Preoperative	65.5±29.7	68	61.6±25.7	58	0.528
Postoperative	75.9±24.9	84	82.9±14.4	79	0.556
p	0.102		0.005		
General health perception					
Preoperative	34.0±22.6	27.5	29.6±25.3	23.5	0.414
Postoperative	70.0±16.5	72.0	65.3±23.8	68.5	0.614
p	0.001		0.001		
Energy					
Preoperative	33.9±22.1	32.5	34.4±18.4	35	0.712
Postoperative	63.2±16.2	65	67.2±22.9	72.5	0.223
p	0.001		0.001		
Social function					
Preoperative	55.7±22.4	50	41.7±27.5	37.5	0.063
Postoperative	74.4±17.9	75	77.1±26.2	87.5	0.402
p	0.012		0.001		
Role limitation due to emotional problems					
Preoperative	42.4±25.6	33.3	35.2±31.3	33.33	0.302
Postoperative	68.2±30.0	66.67	57.4±33.9	66.67	0.294
p	0.006		0.009		
Mental health					
Preoperative	48.4±18.9	44	50.7±17.7	50	0.540
Postoperative	70.7±17.5	74	72.2±21.7	76	0.567
p	0.001		0.001		

SD: Standard deviation.

It is not always possible to prevent mitral valve regurgitation after MVr. Soon or later, regurgitation is expected to recur. In the presence of a significant level of calcification or fibrosis, it is reasonable to perform a MVR operation to decrease the need for reoperation. In a recent meta-analysis, MVR and repair were compared in terms of multiple factors and it was found that MVr was related to an increased reoperation risk in patients with progressive rheumatic mitral valve disease.[19] It was also reported that there was no significant difference between MVR and MVr in terms of the survival rates in patients with ischemic mitral valve disease.[20-22] However, 30 days of mortality reduced in patients who underwent MVr. In another study, MVr was found to be related to a longer survival, and valve replacement was a risk factor in terms of long-term mortality.[23] Based on our findings, we can conclude that the method to be chosen for patients with mitral valve disease needs a multi-factorial assessment including age, etiology, comorbidities, additional cardiac pathologies, and severity of the mitral valve disease. Previous studies

also showed that MVr was related with a lower incidence of in-hospital mortality, longer survival, increased left ventricle functions, and a lower risk of valve-related complications.[24-26] Accordingly, MVr is more popular, particularly in patients in whom the ventricular function should be preserved carefully.[27,28] The primary goal of mitral valve operation is to heal the disease, while protecting the functional capacity. Of note, the main goal of all treatment modalities should be to improve the QoL of patients.

Furthermore, we found significantly improved pain scores in group 2 ($p<0.01$), while no statistically significant improvement was seen in group 1 ($p>0.05$). This finding is also consistent with the finding in a previous study.[29] In the aforementioned study, however, Goldsmith and Patel[29] compared preoperative SF-36 results with third-month results. In another study, prognosis was reported to be poor in patients with an EF <50% and left ventricle dysfunction.[6] In addition, Goldsmith and Patel[29] found no significant improvement in the QoL of the patients with an EF value lower than 50%. Our study included only three patients with an EF value lower than 50%, and a significant improvement was seen in the QoL of these three patients.

The major limitations of the present study include being a single-center study, performing all operations by three different surgical teams, a small sample size, and not having mechanical and biological valve replacements in two different groups.

In conclusion, our study results show a significant improvement in the quality of life following both treatments. However, mitral valve repair seems to be superior to mitral valve replacement in terms of pain scores.

Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

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Original Article Open Access

Cardiac Myxomas: Clinical spectrum, investigation findings, and surgical treatment based on our 25-year-experiences

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ABSTRACT

Objectives: The aim of this study was to describe the clinical spectrum, investigation findings, and surgical treatment of cardiac myxomas.

Patients and methods: Between November 1991 and July 2016, 229 patients (99 males, 130 females; mean age 37.1 years; range 7 to 63 years) with a primary or recurrent intra-cardiac myxoma underwent surgical excision at our institute. The diagnosis was made by transthoracic and transesophageal echocardiography. The basic surgical principle of complete wide excision was applied, and either unicameral (right or left atrial approach) or bicameral (both atria opened) surgical approach was used. Myxomas originating from the valve or valve annulus required a special attention. Postoperative echocardiograms were obtained in all patients before discharge. All patients were followed at three and 18 months, and five years postoperatively.

Results: The most common presenting symptoms were dyspnea and palpitation. There were 197 left atrial, 27 right atrial, two left ventricular, and one each of right ventricular, right ventricular outflow tract and multiple myxomas. There were three early death events; however no late mortality was seen. During follow-up, all patients were in the New York Heart Association Class I and echocardiography showed good ventricular functions with normal pulmonary artery pressure. Seven patients developed sporadic recurrence of myxoma. The valves were competent in the patients who underwent valve repair.

Conclusion: Based on our experience, we recommend the left atriotomy approach for left atrial myxomas and the right atriotomy approach for dumbbell-shaped left atrial and right atrial myxomas. Biatrial approach should be used in large and unusually located left atrial myxomas, while individualized approaches should be performed for others. To prevent recurrence, the surgical excision must include a substantial portion of normal endocardium near the base of implantation. The early mortality is commonly seen due to coronary embolism, and the late survival of patients after myxoma excision is usually excellent.

Keywords: Biatrial approach; cardiac tumor; left atrium; myxoma; recurrence.

Primary cardiac tumors are uncommon and represent only 5 to 10% of all neoplasms of the heart and pericardium.[1] About 80% of primary cardiac tumors are benign and, of these, more than half are myxomas.[2,3] The incidence of cardiac myxomas is between 0.0013 to 0.005%.[4] The majority of the data originate from small series of patients in developed countries. In this article, we report our 25-year experiences and describe the clinical spectrum, investigation findings, and surgical treatment of cardiac myxomas.

PATIENTS AND METHODS

Between November 1991 and July 2016, a total of 229 patients (99 males, 130 females; mean age: 37.1 years; range, 7 to 63 years) underwent complete and wide excision of primary or recurrent intra-cardiac myxomas at our institute.

Operative technique

All patients were referred for surgery and conventional median sternotomy approach was used. The cardiopulmonary bypass was established by aortic and bicaval cannulation. The myocardial protection was achieved by antegrade root cardioplegia. A special care was taken to avoid a forceful manipulation of the heart before the aorta was cross-clamped. Various unicameral (either right or left atrial approach) or bicameral (both atria opened for big tumors, particularly for the left atrial tumors) approaches were used.

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Statistical analysis

Statistical analysis was performed using SPSS version 10.0 software (SPSS Inc., Chicago, IL, USA). The Fisher's exact and chi-square tests were used to analyze statistical significance between variables. For continuous variables, the unpaired Student t-test was used. A p value of less than 0.05 was considered statistically significant.

RESULTS

Clinical findings

The common presenting symptoms were dyspnea and palpitation. One patient who had a huge right atrial myxoma developed hypoxia and cyanosis due to right-to-left shunting through atrial septal defect. Detailed clinical presentation is described in the Table 1. The laboratory findings showed an elevated erythrocyte sedimentation rate (ESR) in 171, anemia in 103, eosinophilia in 55, elevated WBC count in 39, and elevated serum globulin levels in all patients. A familial occurrence of the disease was excluded in the first-degree relatives. Initial chest radiography showed cardiomegaly with a cardiothoracic ratio of 55 to 70%, pulmonary congestion in 84, and left atrial enlargement in 47 patients. Electrocardiography showed left atrial enlargement in 79, right atrial enlargement in 21, right axis deviation in 44, left axis deviation in nine, right ventricular hypertrophy in 42, left ventricular hypertrophy in 11, and right bundle branch block in 16 patients. Two-dimensional transthoracic and transesophageal echocardiography were done in all patients and was the main diagnostic tool (Figures 1, 2). Of 229 patients, there were 197 left atrial, 27 right atrial, two left ventricular, and one each of right ventricular, right ventricular outflow tract, and multiple myxoma. These myxomas ranged from 3 to 16 cm in size at their greatest diameter. Preoperative right ventricular systolic pressure (RVSP) was less than 30 mmHg in 29 patients, between 30 and 60 mmHg in 55 patients, and higher than 60 mmHg in 56 patients. In the remaining patients, RVSP was unable to be calculated. Mild right ventricular (RV) dysfunction was present in 10 patients, and moderate and severe RV dysfunction were seen in three and two patients, respectively. Isolated mild left ventricular (LV) dysfunction was present in two patients, while mild to moderate biventricular dysfunction was seen in three and global hypokinesia was detected in one patient. The remaining patients had good biventricular functions.

Table 1
Clinical characteristics of patients

	n	%
Cardiac symptoms		
Dyspnea	199	86.9
Palpitation	128	55.9
Chest pain	64	27.9
Syncope	46	20.1
Orthopnea/paroxysmal nocturnal dyspnea	12	5.2
Cyanosis	1	0.4
Embolic symptoms		
Central nervous system	12	5.2
Peripheral	1	0.4
Coronary	1	0.4
Systemic symptoms		
Fever	54	23.6
Fatigue	43	18.8
Weight loss	32	14
Others		
Pedal edema	19	8.3
Miscelleneous	9	3.9
Auscultation		
Mid diastolic murmur	153	66.8
Tumor plop	114	49.8
Pansystolic murmur	71	31.0
Loud pulmonary second sound	62	27.1
Other signs		
Edema	9	3.9
Hepatomegaly	7	3.1
Clubbing	4	1.7

In addition, there were few associated lesions such as mild mitral regurgitation (MR) in 117, moderate MR in 21, severe MR in six, mild to moderate mitral stenosis in four, mild tricuspid regurgitation (TR) in 49, moderate TR in 10, severe TR in five and ostium secundum type of atrial septal defect (ASD-OS) in one patient. Coronary angiography was performed in patients above 40 years of age to rule out coronary artery lesions. One patient showed recanalized obtuse marginalis, while another patient had right coronary artery disease. In two patients, angiogram showed tumor blush from the right coronary artery and from the left coronary artery in another patient (Figure 3).

Operative findings

Complete excision of the tumor with a cuff of surrounding tissue was the basic principle of excision (Figures 4, 5a, b). Myxomas originating from the posterior mitral annulus required quadrangular

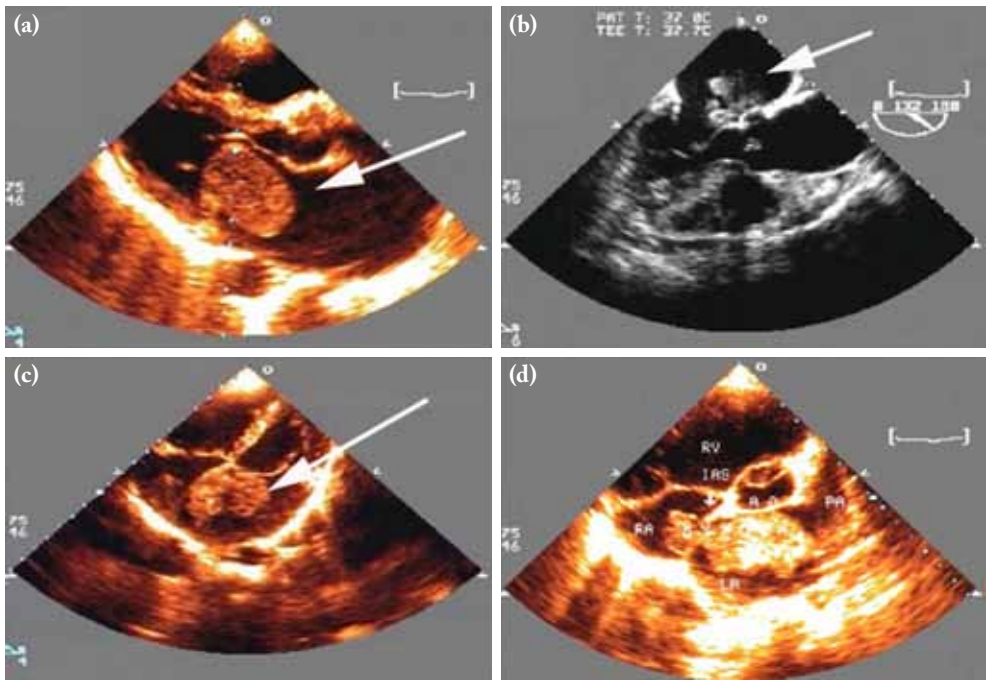

Figure 1. Transthoracic and transesophageal echocardiography images showing **(a)** a typical left atrial myxoma attached to the fossa ovalis of interatrial septum (arrow); **(b)** a multi-headed left atrial myxoma attached to the fossa ovalis of interatrial septum (arrow); **(c)** a left atrial myxoma attached to the interatrial septum and mitral valve-mimicking vegetation (arrow); and **(d)** a dumbbell-shaped myxoma, primarily in left atrium.

resection and prosthetic ring annuloplasty in six patients. Mitral valve repair was required in another 13 patients and repair was performed with anterior mitral leaflet chordal shortening and ring annuloplasty using prosthetic ring in seven patients and commissural annuloplasty in the remaining six patients. Myxomas arising from the tricuspid annulus were shaved off and modified DeVega's annuloplasty was done. In a patient with a myxoma arising from the right ventricular out flow tract involving the pulmonary valve leaflet required excision of the two leaflets. Details of surgical findings, approach, and technique are described in Table 2. Weaning from the cardiopulmonary bypass was done in a usual way. Transesophageal or epicardial echocardiography was done in the patients with valve regurgitation.

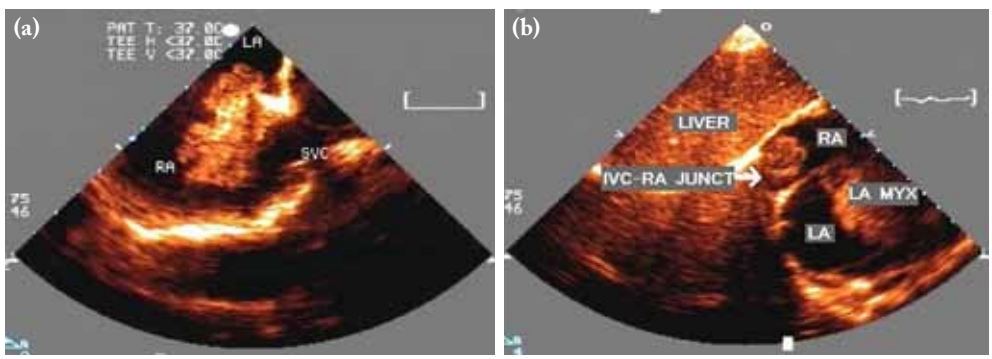

Figure 2. Transesophageal echocardiography images showing **(a)** a right atrial myxoma attached to the fossa ovalis of interatrial septum; and **(b)** a right atrial myxoma at inferior vena cava-right atrial junction.

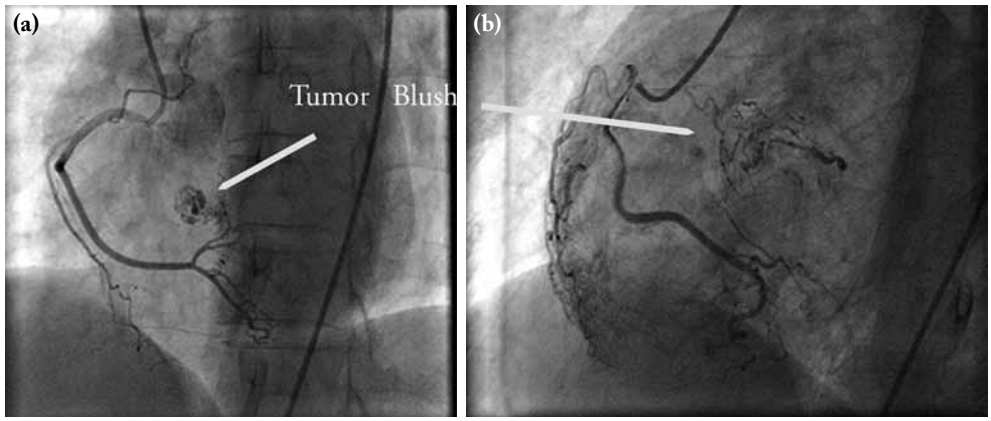

Figure 3. **(a, b)** A right-sided coronary angiogram showing tumor blush from right coronary artery (arrow).

Postoperative outcomes

There were three early mortalities in our entire series due to acute severe left ventricular dysfunction and multi-organ failure, probably secondary to coronary embolism. There was no late mortality. One patient developed convulsion postoperatively and cranial computed tomography (CT) showed an old parietal lobe infarct. The patient was recovered and discharged uneventfully. One patient also developed cerebral hypoxia due to cyanosis caused by right-to-left shunting across the ASD, secondary to right atrial outflow obstruction caused by a huge right atrial myxoma. The patient required prolonged ventilation; however, he recovered and was uneventfully discharged. All patients underwent transthoracic echocardiography on the day of surgery in the intensive care unit and before discharge to evaluate the ventricular functions, valve repair status, and possibility of residual tumors. Echocardiography revealed no residual myxoma in all patients (n=229), good biventricular functions in 224 patients, mild to moderate right ventricular dysfunction in two patients, and severe left ventricular dysfunction in three patients. Three patients also developed mild to moderate MR, one patient developed severe MR requiring mitral valve repair, and one patient developed severe TR, which was medically managed. Postoperatively, the right ventricular systolic pressure was less than 30 mmHg in 87 patients, between 30 and 60 mmHg in 41 patients, and higher than 60 mmHg in two patients. None of the patients had pulmonary embolism. The mean length of intensive care unit stay was 6 (range: 2 to 9) days and the mean length of hospital stay was 9 (range: 5 to 18) days. Follow-up was done at three months, one and five years, and 6 to 17 years. A total of 226 patients survived after the operation, and 203 of them (90%) attended to follow-up at three months. However, the remaining 23 patients (10%) were lost to follow-up. Of the patients, 40.3% (n=66) attended to the follow-up at one year, 30.5% (n=69) at five years, and 24.3% (n=55) between 5 to 17 years. All patients were in the New York Heart Association (NYHA) Class I and echocardiography showed that all patients had good ventricular functions with normal pulmonary artery pressures. However, there were six patients with moderate MR, one with severe MR, and two with moderate to severe TR during follow-up. One patient developed right parietal area infarct leading to left hemiparesis. The patient with severe mitral regurgitation was advised mitral valve replacement. There were seven patients with a recurrent myxoma (Figure 5c) in our entire series, and the mean time to recurrence was 7.2 (range: 2 to 13) years. Apart from one case of multiple myxomas, the remaining six patients developed recurrence at the same site or adjacent area, probably due to inadequate excision or tumor sidling. In a patient with multiple myxomas, a left ventricular myxoma which was developed was thought to be due to multi-centric foci. The location at the initial surgery, recurrence period and site of recurrence are described in Table 3.

DISCUSSION

Although primary tumors of the heart are rare, myxomas are the most frequent benign primary heart

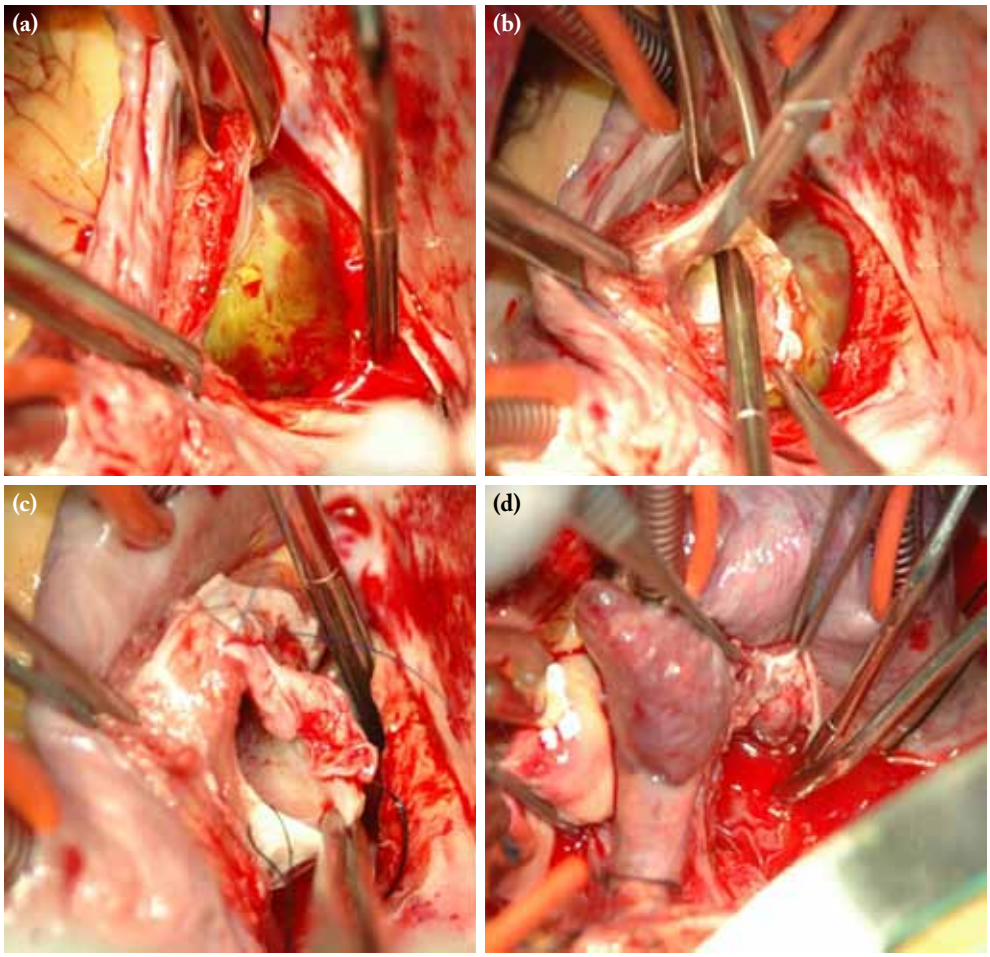

Figure 4. An intraoperative view of left atrial myxoma excision through left atriotomy approach. **(a)** Left atrial approach showing myxoma attached to the interatrial septum; **(b)** left atrial myxoma along with interatrial septum excised; **(c)** a septal defect closure using an autologous untreated pericardial patch; and **(d)** a completed septal closure and checking for suture line leakage.

tumors, which account for 0.3% of open heart surgery worldwide. The clinical presentation in the majority of the patients includes significant hemodynamic symptoms related to the blood flow obstruction and embolic phenomena. Cardiac myxomas can be challenging to diagnose, due to their rare occurrence and varying clinical presentation. The mean age was 37.1 years at the time of diagnosis in our study,

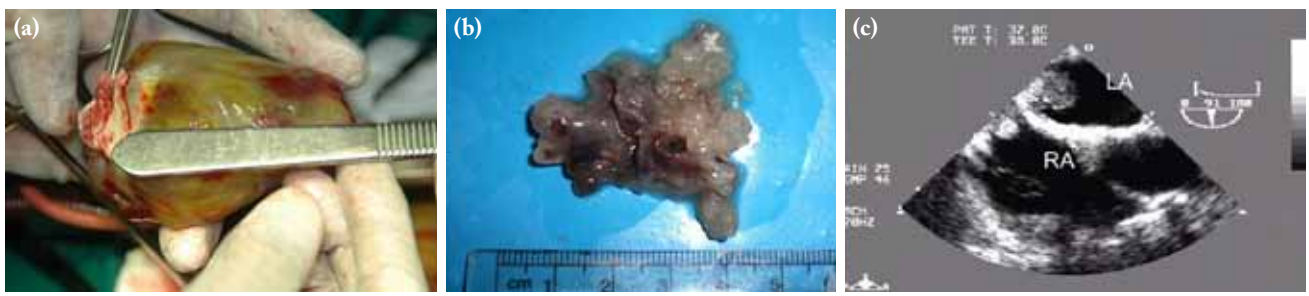

Figure 5. An excised myxoma and postoperative echocardiography showing **(a)** an excised left atrial myxoma; and **(b)** an excised multi-headed myxoma. **(c)** Postoperative transesophageal echocardiography showing a recurrent biatrial myxoma.

Table 2
Operative findings and approaches and techniques applied

LA myxoma			**RA myxoma**			**Others**		
Site of attachment	n	%	Site of attachment	n	%	Site of attachment	n	%
Fossa ovalis (IAS)	170	86.3	Fossa ovalis	23	85.2	LV apex	2	50
LA roof	17	6.6	IVC-RA junction	3	11	RV-IVS	1	25
PML annulus	6	3.0	Tricuspid annulus	1	3.7	Conal septum & PV cusps	1	25
Pulmonary veins	3	1.3				LA, RA, RVOT	1	25
Base of LAA	1	0.5						
Approach			**Approach**			**Approach**		
LA	145	73.6	RA	27	100	LA & aortic	2	50
RA	27	13.7				RA & PA	2	50
Biatrial	25	12.7				PA	1	25
Technique			**Technique**			**Technique**		
WE + direct closure	155	58.4	WE + pericardial patch closure	23	85.2	WE	3	75
WE + pericardial patch closure	57	28.9	Excision & fulguration	4	14.8	WE + pericardial patch closure	1	25
Excision & fulguration	23	11.7						
WE + Dacron patch closure	2	1.0						
Additional procedures			**Additional procedures**			**Additional procedures**		
Mitral annuloplasty	19	9.6	Tricuspid annuloplasty	10	37.0	Pulmonary valvectomy	1	25
CABG & mitral annuloplasty	1	0.5						

LA: Left atrium; RA: Right atrium; IAS: Interatrial septum; LV: Left ventricle; IVC: Inferior vena cava; IVS: Interventricular septum; PML: Posterior mitral leaflet; PV: Pulmonary valve; RVOT: Right ventricular outflow tract; LAA: Left atrial appendage; PA: Pulmonary artery; WE: Wide excision; CABG: Coronary artery bypass grafting.

which is significantly lower than in most Western countries where the mean age is 50 to 55 years.[5-7] In addition, dyspnea was the most common symptom in our patients as the obstructive pathology. Patients may also present with cyanosis and cerebral hypoxia produced by right-to-left shunting secondary to the right ventricular inflow obstruction caused by a huge right atrial myxoma associated with patent foramen ovale or ASD, as in one of our patients. A higher risk of embolization has been also reported and events occur in 30 to 45% of the patients.[5-10] In our series, the embolization was less

Table 3
Recurrence data

Location at first surgery	Recurrence period (years)	Site of recurrence
Right atrium (IAS, fossa ovalis)	13	Biatrial (IAS)
Left atrium (IAS)	13	Biatrial & extending to right atrium wall
Right atrium (Lower end of fossa ovalis)	9	LA (IAS)
Left atrium (IAS)	7	RA (IAS)
Multiple (LA, RA, RVOT) IAS & RVOT	5	Left ventricle apex
Left atrium (IAS)	2	Roof of left atrium & IAS
Biatrial (IAS)	2	Biatrial (IAS)

IAS: Interatrial septum; LA: Left atrium; RA: Right atrium; RVOT: Right ventricular outflow tract.

frequent than the Western series. In our series a total of 14 patients developed embolization preoperatively and three patients developed tumor embolization postoperatively. Of 14 patients, 12 patients had central nervous system (CNS) embolization and one each had peripheral arterial and coronary artery embolization. Of three patients with postoperative tumor embolization, two developed in the early postoperative period and one patient developed in the late postoperative period. All these patients had CNS embolization. Anemia and elevated ESR are non-specific markers of several diseases and are significantly more common in our study. The presence of systemic symptoms was positively correlated with elevated ESR. Systemic symptoms, anemia, and elevated ESR could be due to the systemic effects of interlukin-6, the cytokine implanted in generating a generalized inflammatory response in patients with myxomas. There were also elevated serum globulin levels in all patients with reversal of albumin-globulin ratio in our series.

Echocardiography is currently the most main diagnostic modality available for imaging cardiac tumors. It is non-invasive and allows a preoperative diagnosis with accuracy and can quantitate the tumor size, shape, attachment, and mobility.[11,12] It can also screen the other chambers of the heart for additional tumors. In addition, transesophageal echocardiography has an increased sensitivity and specificity for the diagnosis, particularly in patients with poor transthoracic echo window.[11,12] In our series, the two-dimensional transthoracic and transesophageal echocardiography were successful primary tools for the diagnosis of cardiac tumors. Magnetic resonance imaging (MRI) and CT are not the first-line diagnostic tools for myxomas, although more and more cases are diagnosed by these technologies. These modalities are helpful to detect benign and malignant tumors, when transthoracic and transesophageal echocardiography offer limited tissue characterization and confident distinction between thrombi.[11-14] In addition, prolapse through the mitral valve orifice on CT is a reliable discriminative finding indicating a myxoma,[11-14] while the absence of both first-pass and delayed contrast enhancement on MRI is suggestive of a thrombus.[11-14]

Cardiac CT is also useful to detect metastases in suspected malignancies, particularly when coupled with 18 F-fluorodeoxyglucose (FDG) positron emission tomography (PET). However, if a mass has a typical echocardiographic appearance and is located as a left atrial myxoma, additional images with CT or MRI are unnecessary. Surgical excision is the treatment and must be undertaken immediately to avoid the complications, such as systemic embolization and valvular obstruction or incompetence.

The first successful surgery of a myxoma was performed by Crafoord in Stockholm, Sweden in 1955, on cardiopulmonary bypass.[1-19] Since then, many approaches have been described in the literature, such as left atriotomy, right atriotomy and biatrial approach.[11-19] In our series, the approaches used for left atrial myxomas are left atriotomy in 73.6% (n=145), right atriotomy in 13.7% (n=27), and biatrial approach in 12.7% patients (n=25). Based on our findings, we found that the left atriotomy approach was much more convenient, simple, rapid, and safe approach for the excision of the left atrial myxomas. The right atriotomy approach was used for dumbbell-shaped left atrial myxomas. The biatrial approach was used in large and unusually located left atrial myxomas. The right atrial myxoma was approached through right atriotomy, while other types of myxoma were approached either through right atrium and pulmonary artery for the right ventricular outflow tract myxomas and left atriotomy and transaortic approach for the left ventricular myxomas. A wide excision with direct closure of the defect was also done in 58.4%, pericardial patch closure of the defect in 28.9%, excision and fulguration of the raw area in 11.7%, and the use of a Dacron patch in 1% patients of left atrial myxomas. For the right atrial myxoma excision, a pericardial patch was used in 85.2% patients and excision with fulguration was done in 14.8% patients. Myxomas attached to the chamber wall apart from the interatrial septum, such as the right ventricular septum, left ventricular apex, and tricuspid valve annulus require wide excision alone or excision with fulguration. Myxomas attached to the pulmonary valve require excision of the valve leaflet, while those attached to the posterior mitral leaflet can be excised with the quadrangular resection of the leaflet with myxoma, followed by prosthetic ring annuloplasty. Myxomas attached to the anterior mitral leaflet require treatment according to the involvement of the leaflet area. If a small portion of the leaflet is involved, myxoma can be shaved off the leaflet, or a small portion of the leaflet can be excised, and the defect can be repaired with an autologous pericardial patch. However, if the major portion of the leaflet is involved, it may require mitral valve replacement. To prevent recurrence, there

is a consensus that surgical excision must include a substantial portion of the normal endocardium near the base of implantation.[16-19] Recurrence can be due to inadequate resection, intraoperative implantation, embolization or multi-centric growth.[5,6] In our series, the recurrence rate was 3.1%, which is comparable to the international standard of 5%.[16,17,19] The recurrence was mainly found in left atrial myxomas, excised through right atriotomy approach, probably due to inadequate excision or intraoperative implantation. In one case of multiple myxomas, recurrence was seen at a different location, probably due to the multi-centric growth. There were also three early mortalities in our entire series, due to acute severe left ventricular dysfunction and multi-organ failure, probably secondary to coronary embolism. There was no late mortality or pulmonary embolism.

In conclusion, cardiac myxomas are the most common benign cardiac tumors in adults and the left atrium is the commonest chamber involved. Transthoracic and transesophageal echocardiography is the optimal diagnostic modalities for myxomas. The diagnosis of a cardiac myxoma is an indication for an urgent surgery, due to the high risk of sudden death from a thromboembolism or valvular obstruction. Therefore, we recommend the left atriotomy approach for the left atrial myxoma excision in view of convenience, simplicity, and safety and right atriotomy approach for dumbbell-shaped and right atrial myxomas. The biatrial approach can be used in large and unusually located left atrial myxomas, while individualized approaches can be used for the treatment of for other types of myxoma. To prevent recurrence, the surgical excision must include a substantial portion of the normal endocardium near the base of implantation. The early mortality is most commonly due to coronary embolism and the late postoperative survival is usually excellent.

Declaration of conflicting interests

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Cardiovascular Surgery and Interventions

Case Report Open Access

Ethylenediaminetetraacetic acid-dependent pseudothrombocytopenia in complex cardiac surgery

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ABSTRACT

Pseudothrombocytopenia is an unusual hematological disorder, which develops in response to ethylenediaminetetraacetic acid-dependent anti-platelet autoantibodies in blood, leading to platelet clumping. It is only an *in vitro* phenomenon, which presents with low platelet counts in routine hematology analysis. The definitive diagnosis should be established to avoid a delay in surgery and unnecessary blood transfusion in patients undergoing cardiovascular surgery. Herein, we present a 51-year-old female case with ethylenediaminetetraacetic acid-dependent pseudothrombocytopenia, who underwent a successful aortic root and valve surgery, and discuss perioperative management of this rare disorder.

Keywords: Cardiac surgery; ethylenediaminetetraacetic acid-dependent; ethylenediaminetetraacetic acid; pseudothrombocytopenia; pseudothrombocytopenia.

Pseudothrombocytopenia is a rare hematological disorder of platelet clumping related to ethylenediaminetetraacetic acid (EDTA)-dependent antiplatelet autoantibodies in blood.[1-4] The EDTA is a safe anticoagulant for a complete blood count analysis. However, this agent may induce the clumping, which causes the automatic hematology analyzers to undercount platelets, thereby, resulting in low platelet counts. Pseudothrombocytopenia is only an *in vitro* effect, which does not cause any hemostatic complications, as all platelet functions and coagulation tests are normal.[5] It can be seen in some patients with autoimmune diseases, malignancies, chronic liver diseases, viral infections, and cardiovascular diseases. In addition, pregnant women and healthy individuals may rarely present with this disorder.[5] The diagnosis of pseudothrombocytopenia is of unique clinical importance to avoid a delay in surgery and unnecessary blood transfusion in patients undergoing cardiac surgery.

Although EDTA-dependent pseudothrombocytopenia has been previously described in patients undergoing cardiac surgery,[2-4] its clinical features and management approaches still remain controversial for cardiac surgeons. Herein, we present a case of pseudothrombocytopenia who underwent aortic root replacement, mitral valve replacement, and tricuspid valve repair and discuss perioperative management of this entity.

CASE REPORT

A 51-year-old female was referred to our hospital with progressive dyspnea due to valvular heart disease. Her medical history revealed chronic renal failure and hypertension. She was also on medical treatment for hypertension with angiotensin-receptor blockers. On admission, her vital signs were stable. Physical examination revealed aortic, mitral, and tricuspid diastolic murmur. In the biochemical analysis, low platelet counts (65,000/mm^3) were observed. There was no skin lesion such as petechiae, ecchymosis, or purpura. Other than thrombocytopenia, biochemical and serological test results were normal without any indicator of an infection, inflammatory disease or coagulation disorder. Electrocardiography revealed sinus rhythm with a left-axis deviation. A chest X-ray showed cardiomegaly with enlarged left chambers. Echocardiography revealed an ejection fraction of 45%

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with a severe aortic and mitral valve regurgitation and moderate tricuspid valve regurgitation. Pulmonary artery pressure was 40 mmHg. It also showed enlarged left cardiac chambers, mild pericardial effusion, and an aneurysm of the ascending aorta. Thoracic computed tomography revealed an aortic annulus of 30 mm, sinus of Valsalva of 43 mm, sinotubular junction of 36 mm, and ascending aorta of 51 mm in size. Coronary angiography demonstrated no abnormality of coronary arteries. The patient was decided to undergo the Bentall procedure with mitral valve replacement and tricuspid valvuloplasty. Preoperatively, she was referred to a hematology consultant. EDTA-dependent pseudothrombocytopenia was diagnosed with a peripheral blood smear which showed platelet clumping (Figure 1). Clumping was not observed after analysis of heparinized blood sample. Surgery was decided based on the discretion of the consultant hematologist.

The operation was performed with a median sternotomy and systemic heparinization. Cardiopulmonary bypass was initiated through the cannulation of the ascending aorta and both vena cava. Cardiac arrest was established with antegrade infusion of isothermic blood cardioplegia through the coronary ostia. First, right atriotomy incision was performed and, using transseptal incision, the mitral valve was explored. It was degenerated and replaced with a No. 29 bileaflet mechanical valve, preserving the posterior mitral leaflet. Then, the Bentall procedure with a No. 21 mechanical aortic valve conduit was performed. Finally, the tricuspid valve annuloplasty was made using a No. 29 flexible annuloplasty ring. The valve showed a good coaptation on saline test. The operation was completed uneventfully. Cardiopulmonary bypass and aortic cross-clamp times were 169 and 137 min, respectively. She was transferred to the ward on postoperative Day 1 with a platelet count of 74,000/m^3. No platelet suspension was delivered postoperatively.

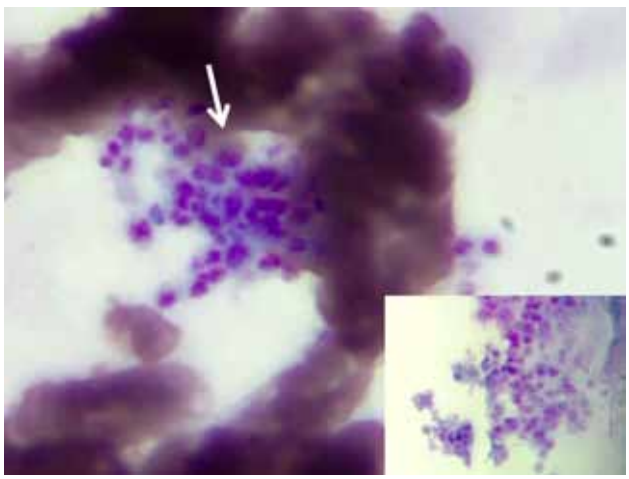

Figure 1. Hematoxylin and Eosin stained EDTA-blood smear showing platelet clumping (arrow, inset) (H-E x 40).
EDTA: Ethylenediaminetetraacetic acid.

The patient was discharged home with a favorable outcome on postoperative Day 6. The platelet count ranged between 65,000 and 87,000/mm^3. Routine anticoagulation was delivered, including early delivery of low-molecular-weight heparin and warfarin with an international normalized ratio of ranging between 2.5 and 3.5. At four months of follow-up, she is still disease-free.

DISCUSSION

Acquired platelet disorders are mainly classified into disorders of the platelet count and function.[1] The etiology of low platelet count includes decreased production by bone marrow and increased peripheral destruction due to immunological causes, non-immunological disorders, hemodilution, and pseudothrombocytopenia. The prevalence of EDTA-dependent pseudothrombocytopenia is reported to be between 0.1% and 2% among hospitalized patients and up to 17% in patients with isolated thrombocytopenia.[3,5] The EDTA is an anticoagulant, which is frequently used for hematological tests, and may induce platelet clumping. However, some other anticoagulants such as heparin, oxalate, hirudin, citrate, or abciximab may also cause pseudothrombocytopenia, although their actual incidence is still unclear.[5]

The underlying mechanism of platelet clumping in pseudothrombocytopenia includes an immunological process which was first described by Shreiner and Bell in 1973.[6] The authors proved that EDTA caused a new type of platelet agglutinin, which was active at 37 °C, as well as at room temperature. The EDTA-dependent antiplatelet antibodies recognize and activate different receptors, such as glycoprotein IIb-IIIa and thrombospondin, resulting in platelet clumping in the *in vitro* setting. Previously, Bizzaro[1] reported antiplatelet antibodies in 83% of the patients with pseudothrombocytopenia.[1] These antibodies were majorly immunoglobulin (Ig) M or IgG. In addition, a small number of patients had an IgA class.[1] Although

platelet clumping occurs during hematological tests, the number and function of the platelets can be normal in patients with pseudothrombocytopenia in the *in vitro* setting.[1] Previous reports also showed that low platelet counts were not associated with an increased risk of bleeding in the perioperative period of cardiac surgery.[2-5]

Although the cardiac surgical practice is itself associated with hemorrhagic complications, EDTA-dependent pseudothrombocytopenia presents with a benign course.[2-4] Preoperatively, patients usually have low platelet counts in the routine blood tests. Due to platelet clumping, conventional automated hematology analyzers count each clump as one and do not show the actual number of platelets. Therefore, the visual assessment of blood smears for clumping is considered as the gold standard for the diagnosis of this phenomenon.[1-5] Unawareness of this entity may lead to a delay in cardiac procedures or unnecessary transfusion in the perioperative period. These may, eventually, lead to severe complications, particularly in high-risk patients with a critical coronary or valve disease.

In the literature, pseudothrombocytopenia in cardiovascular operations has been described in few reports; however, it is still an unusual entity for cardiac surgeons.[2-4] The first report by Dalamangas et al.[3] described an uneventful coronary revascularization and aortic valve replacement using cardiopulmonary bypass. Then, Wilkes et al.[2] presented a patient with anticoagulant-induced pseudothrombocytopenia who underwent a successful coronary artery bypass grafting. The authors confirmed the low platelet count and clumping with microscopic examination, and concluded that clumping was associated with both EDTA and citrate on hematological analysis in the postoperative period. On the other hand, Nair et al.[4] reported that EDTA-dependent pseudothrombocytopenia could be easily diagnosed by repeating platelet counts in citrate and heparin-anticoagulated blood samples. These reports confirm that any type of anticoagulant can be associated with clumping. To the best of our knowledge, our case is the first report of a complex cardiac surgery with a prolonged cardiopulmonary bypass time in pseudothrombocytopenia.

The diagnostic approaches in such cases include blood smears which are simple and valuable tests to show an abnormality of clumping or hemostasis. Laboratory tests such as thromboelastography, which shows specifically the function of platelets, can be also used to reveal an abnormality of coagulation related to platelets. It is a simple and rapid tool for the diagnosis of hemostatic disorders; however, the availability of this tool is limited. Beyond the diagnostic tests, clinical experience is also important for hemostasis in complex and prolonged cases. In our case, if the duration of the procedure prolonged, which means coagulation can be affected from the cardiopulmonary bypass time, we could use platelet suspensions after weaning from cardiopulmonary bypass. Indeed, there is no known drawback of using platelets in prolonged and complex cardiac surgery procedures.

In conclusion, ethylenediaminetetraacetic acid-dependent pseudothrombocytopenia is a rare disorder of platelet clumping during hematological testing. It is a benign phenomenon and does not pose an increased risk for perioperative bleeding. The definitive diagnosis of pseudothrombocytopenia prevents unnecessary testing for thrombocytopenia, a delay in surgery, and unnecessary platelet transfusions.

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Cardiovascular Surgery and Interventions

Case Report Open Access

Combined double-chambered right ventricle with double-chambered left ventricle: a rare anomaly

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ABSTRACT

Double-chambered right ventricle with double-chambered left ventricle is a very rare congenital anomaly. An 18-year-old female presented with dyspnea, chest pain, and palpitation. Diagnosis was made using transthoracic echocardiography and was confirmed by angiography. Surgical excision of the anomalous muscle bundles in the right ventricular outflow tract along with excision of the fibrous band from the left ventricle was performed. The postoperative course was uneventful and repeated echocardiogram before discharge showed a right ventricular outflow tract gradient of 8 mmHg, no gradient across the left ventricular cavity, and improved biventricular functions. The management strategy is dependent on presence of symptom, associated anomalies and type of pathology.

Keywords: Angiography; double-chambered left ventricle; double-chambered ventricle; echocardiography; infundibular stenosis; subaortic stenosis.

Double-chambered ventricle is a rare congenital cardiac anomaly, where the ventricular chamber is wholly or partially partitioned usually by abnormal muscular ridges or fibrosis.[1-9] Unlike double-chambered right ventricle (DCRV), division of the left ventricle (LV) is a rare anomaly.[1-5] Previously, the description of the double-chambered left ventricle (DCLV) was given by Gerlis et al.[1] and Kay et al.[2]

Herein, we describe a case of DCRV and DCLV in an 18-year-old female.

CASE REPORT

An 18-year-old female patient presented with dyspnea, chest pain, and palpitation. The physical examination revealed a Grade 4/6 ejection systolic murmur at the left upper sternal border. Chest X-ray showed a cardiothoracic ratio of 50% with reduced pulmonary blood flow. Electrocardiogram showed right ventricular hypertrophy. Transthoracic echocardiography revealed levocardia and DCRV with a gradient of 70/58 mmHg associated with a ledge of muscle tissue in the LV cavity below the mitral valve (Figure 1a) with 30 mmHg gradient across the constriction with figure of 8 appearance in apical 4 chamber view. No regional wall motion abnormality and improved biventricular functions were observed. Cardiac catheterization revealed a right atrial pressure of 21/0 mmHg with a mean of 10 mmHg, a right ventricular pressure of 157/10 mmHg, a right ventricular outflow tract of 31/10 mmHg, a pulmonary artery of 33/11 mmHg, a pulmonary artery wedge of 17/6 mmHg, a LV apex of 162/11 mmHg, a LV outflow tract of 152/76 mmHg, an aortic pressure of 141/83 mmHg, and a femoral artery pressure of 157/77 mmHg. The gradient between the right ventricular body and the outflow tract was 126 mmHg, while the gradient across the fibrous ridge between the LV apex and outflow was 10 mmHg. Left ventricular angiography showed a ledge of tissue in the mid-LV cavity below the mitral valve (Figure 1b), separating the LV with a hypertrophied distal chamber and thin-walled proximal chamber. Right ventricular angiography revealed the presence of anomalous septal and parietal bands, producing DCRV with a good-sized main pulmonary artery and confluent branch pulmonary arteries. The surgical procedure was performed through a conventional median sternotomy and cardiopulmonary bypass was established by aortic

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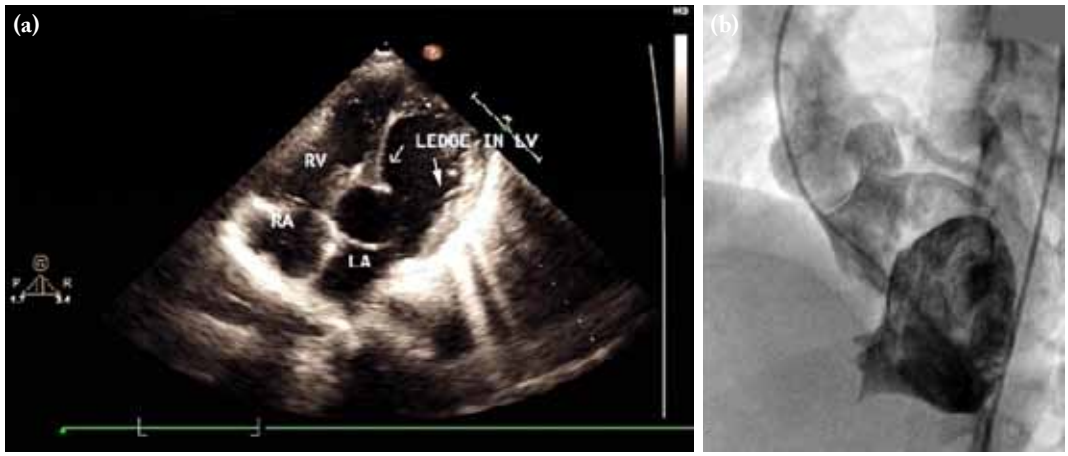

Figure 1. A preoperative echocardiogram and angiogram view. **(a)** Transthoracic echocardiogram showing ledge of tissue from both the wall of the left ventricle, more from the septal wall, producing double-chambered left ventricle. **(b)** Left ventriculography showing double-chambered left ventricle with possibly closed apical ventricular septal defect.

RA: Right atrium; RV: Right ventricle; LA: Left atrium; LV: Left ventricle.

and bicaval cannulation. The myocardial protection was provided with cold blood antegrade cardioplegia. After cross-clamping the aorta, the right and the left atrium were opened, and the anatomy was visualized. The intraoperative findings showed a muscular band in the LV cavity below the mitral valve (Figure 2a), and there was a tight fibrous ring with anomalous septal and parietal bands in the right ventricular cavity. The ledge of the muscular tissue in the LV cavity was accessed and excised through a left atrial-mitral valve route, and the fibrous ring and muscle bundles in the right ventricle was accessed and excised through the right atrial-tricuspid valve route. Weaning from the cardiopulmonary bypass was performed in a usual fashion. Intraoperative transesophageal echocardiography showed no gradient across the mid LV cavity, trivial mitral regurgitation, and an-8-mmHg gradient across the right ventricular outflow tract with improved biventricular functions. The postoperative course was uneventful, and transthoracic echocardiography before discharge revealed a widely opened right ventricular outflow tract and mid-LV cavity (Figure 2b) with a right ventricular outflow gradient of 8 mmHg and no gradient in the mid-cavity of the LV with improved biventricular functions. The histopathology examination of the resected tissue from the LV cavity showed a fibrous tissue with normal myocardium.

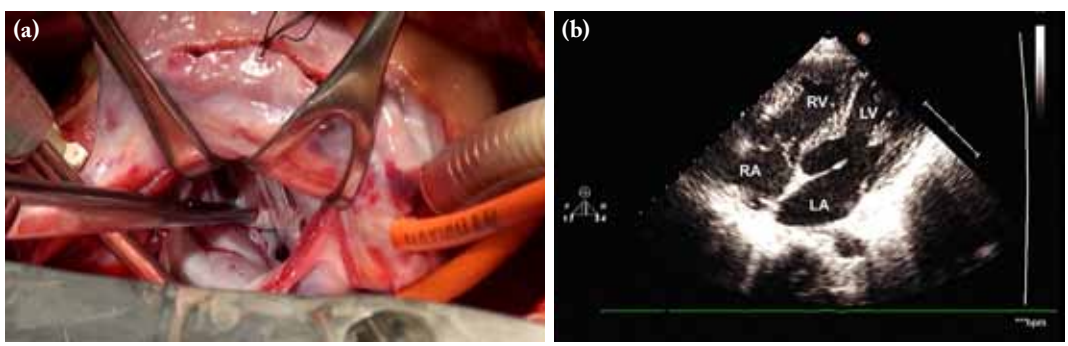

Figure 2. Surgical procedure image and post operative echocardiogram showing. **(a)** An intraoperative view obtained from the surgeon's side, showing muscle tissue in the left ventricle with fibrosis over, producing double-chambered ventricle. **(b)** A postoperative transthoracic echocardiogram view showing a widely opened left ventricular cavity.

RA: Right atrium; RV: Right ventricle; LA: Left atrium; LV: Left ventricle.

Figure 3. Types of double-chambered left ventricle.
Type 1: The left ventricular cavity is divided into two chambers and both the chambers communicate through multiple orifices between the smooth-walled chamber and the left ventricle proper.
Type 2: The left ventricular cavity is divided by the hypertrophied muscular wall with an apical portion having prominent and enlarged branching sinusoids lined by thickened endocardium, more commonly seen with endocardial fibroelastosis.
Type 3: The left ventricle is completely divided into two portions and the both chambers lie side by side.

DISCUSSION

Double-chambered left ventricle is a very rare congenital cardiac anomaly.[1] This pathology is rarer than DCRV, as described by Kay et al.[2] In addition, the association of DCLV with DCRV is extremely rare. Although the etiology of DCLV is less known, it is thought to be congenital and non-progressive.[1,2,4,5] Various theories for DCLV have been suggested, including post-inflammatory pathologies, defects or hypoplasia of the myocardial wall, and division by a viable myocardium with fibrous opening and the endocardial fibroelastosis.[1,2,4,5] Embryologically, DCLV is caused by the failure of regression of the fetal trabeculations.[4] Gerlis et al.[1] classified DCLV into three types (Figure 3): Type 1, where the LV cavity is divided into two chambers and both chambers communicate through multiple orifices between the smooth-walled chamber and the left ventricle proper. In type 2, the LV cavity is divided by the hypertrophied muscular wall with an apical portion, having prominent and enlarged branching sinusoids lined by thickened endocardium, more commonly seen with endocardial fibroelastosis. In type 3, the LV is completely divided into two portions and both chambers lie side by side.

Double-chambered left ventricle can be differentiated from the relatively more common congenital aneurysm and diverticula.[1] It is best differentiated from LV aneurysms and pseudoaneurysms, by the fact that the double-chambered ventricles exhibit normal contractile motions during systole.[1] Ventricular aneurysms also lack complete layering of the ventricular wall, thereby, expanding slightly due to the increased pressure during systole.[1]

In general, DCLV is incidentally detected in the course of an evaluation for other cardiovascular abnormalities. The differentiation between DCRV and DCLV is evident, as both have a distinct pathophysiology. The former is more common and often presents with murmur and exertional dyspnea. Several studies have found that DCRV is associated with septal defects, tetralogy of Fallot, and transposition of the great arteries.[5-7] Conversely, the latter one is commonly asymptomatic. In addition, DCRV is often caused by a progressive thickening of the right ventricular septum due to the presence of anomalous muscle bundles, which results in a pressure gradient and formation of two chambers in series. In contrast, the chambers of a DCLV are in parallel and present less of a pressure gradient, as both contract synchronously.[4,6,7] Similarly, in our case, echocardiographic gradient was 30 mmHg; however, catheterization gradient was only 10 mmHg. The DCLV etiology is less well-known, although the anomaly is thought to be congenital and non-progressive. As this is an extremely rare finding, no definite data regarding the prognosis, outcomes, and potential complications, such as risk of an embolism, in the DCLV are available. It is often believed that

DCLV poses little risks to the patient, and treatment is usually guided by the presence of other associated abnormalities.[2,5]

Furthermore, the transthoracic and transesophageal echocardiography and computed tomography can aid in the detection of the double-chambered ventricles.[2-5] Transthoracic echocardiography in short-axis inverted views and apical four-chamber views show a transverse muscle band traversing the LV dividing the LV into two halves, producing a typical "figure-of-eight" appearance, indicating a double-chambered.[8] However, cardiac magnetic resonance imaging allows an improved delineation of this condition, thanks to its higher spatial resolution and the ability for tissue characterization, particularly regarding the differentiation between fibrosis and normal myocardium, which would not be easily achievable with echocardiography[8] or computed tomography.[3]

In the literature, several treatment strategies have been described.[2,4,5] In asymptomatic patients without any associated anomaly or non-significant gradient across the DCLV, clinical observation and follow-up are recommended. However, in symptomatic cases with or without gradient across the DCLV or with an associated lesion, surgical excision is indicated. There are several surgical techniques in the literature.[2,4,5] For type 1 and type 2 cases, excision of the obstructive shelf is recommended, while accessory chamber exclusion is indicated for type 3 disease. Due to the rarity of DCLV, on the other hand, few data exist on the treatment approaches and outcomes. Surgical excision of the accessory chamber with interposition patch reconstruction and cardiac transplant are two valid options.[4,5]

Although our case did not fit to any of the types described by Gerlis et al.,[1] it had a muscular self-like projections producing DCLV which was associated with DCRV. As it was associated with DCRV, presentation was delayed and required excision of an abnormal muscle bridge in the LV cavity along with coring out of the right ventricular cavity. The histopathological examination of the resected tissue showed a fibrous tissue with normal myocardium and no evidence of endocardial fibroelastosis. The etiology probably might be abnormal muscular hypertrophy or poor regression of the fetal trabeculations.

In conclusion, double-chambered right ventricle is a rare congenital anomaly; however, the association of this pathology with double-chambered left ventricle is extremely rare. As in our case, both anomalies are responsible for the clinical presentation. The diagnosis can be achieved by transthoracic echocardiography and can be confirmed by angiography and cardiac magnetic resonance imaging. The treatment modalities vary according to symptoms, associated lesions, and type of the pathology.

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Cardiovascular Surgery and Interventions

Case Report Open Access

Surgically revascularized intercoronary communication: an extremely rare case of an open-ended circulation pattern

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ABSTRACT

Coronary artery abnormalities are congenital deformities including the origin, tract, and structure of the coronary arteries. Intercoronary communicating vessels are the conduits between two or more coronary arteries enabling either one- or two-way blood flow and they are exceptionally rare. In this article, we present a 54-year-old male case of coronary artery abnormality with typical chest pain and exertional angina and discuss surgical revascularization in the light of literature data.

Keywords: Coronary vessel abnormality; intercoronary communication; revascularization; surgery.

Intercoronary communication (ICC), which is extremely rare, was first described by Cheng in 1972.[1] This type of communication between normal coronary arteries is congenital with a diagnostic coronary angiography incidence of 0.05%.[2]

Two types have been described: communication between the left anterior descending (LAD) and posterior descending artery (PDA) in the distal interventricular groove, and communication between the circumflex artery (Cx) and right coronary artery (RCA) in the posterior atrioventricular groove.[3] The communication between the LAD and RCA in the distal PDA branch is extremely rare.[3] Herein, we report a 54-year-old male case of coronary artery abnormality with typical chest pain and exertional angina and discuss surgical revascularization in the light of literature data.

CASE REPORT

A 54-year-old male patient was admitted to our cardiology outpatient clinic with complaints of typical chest pain and exertional angina. Physical examination and laboratory analysis showed no pathology. His medical history revealed that he was a current smoker, which was a risk factor for heart disease. Transthoracic echocardiography demonstrated an ejection fraction (EF) of 60%, left ventricular diastolic relaxation abnormality (grade 1), left atrial dilation (4.0 cm), and minimal mitral regurgitation (1° MR). Myocardial perfusion scintigraphy showed reversible hyperperfusion in the apical, basal, lateral and inferolateral walls, and hypokinesia in the apical, lateral and inferolateral walls. Coronary angiography showed a severe lesion (80 to 90%) in the LAD before the second diagonal branch and a severe lesion in the proximal branch of the RCA (70 to 80%). In addition, a communication was detected between the RCA and LAD artery branches (Figure 1). This communication competed between two arteries in both diastoles.

A written informed consent was obtained from the patient. He, then, underwent elective surgery with median sternotomy through aorto-atrial cannulation under cardiopulmonary bypass. Antegrade cold blood cardioplegia was applied in on-pump surgery. Intraoperatively, ICC was clearly detected. Three-vessel (RCA-saphenous vein, second diagonal artery-saphenous vein, and LAD artery-left internal mammary artery) coronary artery bypass grafting (CABG) was performed. In the postoperative period, the patient was hemodynamically stable, and he was uneventfully discharged with acetylsalicylic acid

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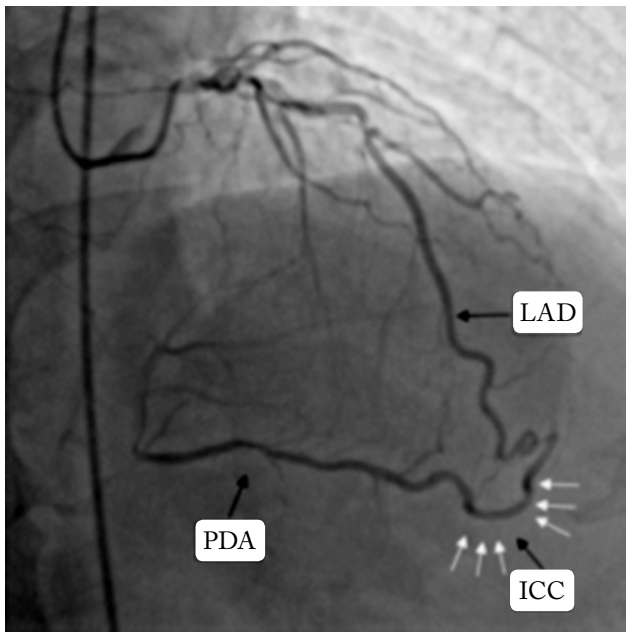

Figure 1. A coronary angiographic view showing intercoronary communication between the LAD artery and RCA in the distal PDA branch.

LAD: Left anterior descending; RCA: Right coronary artery; PDA: Posterior descending artery; ICC: Intercoronary communication.

300 mg daily after one week of surgery. At three months, he is still uneventful.

DISCUSSION

Intercoronary communication is congenital in origin, and it has been suggested that a defective embryological development allowed the existing intercoronary channel to remain prominent.[4]

As it is a congenital malformation, it can be misdiagnosed as collateral circulation secondary to obstructive coronary artery disease.[5] Intercoronary collaterals are tortoise, typically smaller than 1 mm with a very meandering course. Histologically, they are composed of endothelium supported by weak collagens, and muscular and elastic fibers.[2,3] On the other hand, intercoronary communications are typically larger in size (≥1 mm), single, and straight vessels and are arterioles which carry the muscular structure of the epicardial vessels.[2,3]

It is well-established that the main goal of CABG is to treat reversible ischemia due to coronary artery stenosis. In case of an ICC, which is extremely rare, treatment, intervention, and follow-up are of utmost importance.

On the other hand, the functional value of this type of abnormality still remains to be elucidated. These connections may play a protective role for myocardium, if the coronary artery obstruction develops in one of the two connecting vessels.[6] This abnormality, also known as an open-ended coronary circulation pattern, is considered to be protective for the myocardium against occlusive coronary damage.[6] However, some authors have suggested that one-way flow, as confirmed by coronary angiography, may result in insufficient perfusion, thereby, leading to coronary steal phenomenon and myocardial ischemia.[7]

In our case, two-way flow was maintained through contrast injection into the right or left coronary artery. It was evident that two-way intercoronary blood flow, which led to a competition between two arteries in both diastoles, was seen and that it was not the communication between the collaterals between the RCA and LDA artery branches. We believe that the absence of isolated RCA stenosis induced competition phenomenon, rather than steal phenomenon in our case in whom the abnormality originated from an atherosclerotic process. In addition, coronary angiography showed severe atherosclerotic plaques in the RCA and LAD artery, while myocardial perfusion scintigraphy revealed reversible ischemic regions. Based on these imaging findings, both arteries were bypassed and only the left internal mammary artery was used to avoid sternal instability. On the other hand, it is still controversial that which arteries are bypassed, which grafts are selected, and how the selected grafts affect the ICC. Based on our experience, we recommend that the atherosclerotic processes should be investigated, vessels with ICC should be examined, and steal/competition phenomena should be further evaluated.

In conclusion, the uncommon presence of an open-ended coronary circulation pattern is known in patients with and without coronary artery narrowing. In the presence of obstructive coronary artery disease, it can be confused with collateral circulation, although both share distinct anatomical, histological, and functional structures. Although rare, it may cause an incorrect diagnosis and treatment, if left unrecognized; therefore, clinicians should be careful.

Declaration of conflicting interests

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Cardiovascular Surgery and Interventions

Case Report Open Access

Pulmonary sinus of Valsalva aneurysm: A rare entity

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ABSTRACT

Pulmonary sinus of Valsalva aneurysm is rare. An intrinsic weakness of the wall and increased hemodynamic stress is the main causative factor. Surgery has a definite role. Herein, we describe a case of pulmonary sinus of Valsalva aneurysm who was successfully treated in our clinic.

Keywords: Pulmonary sinus of Valsalva aneurysm; pulmonary valve; sinus of Valsalva aneurysm.

Pulmonary artery sinus of Valsalva aneurysm is an unusual lesion,[1-4] which can be associated with congenital heart diseases, pulmonary artery hypertension, pulmonary valve stenosis, connective tissue diseases (such as Marfan syndrome), and vasculitis. To the best of our knowledge, true pulmonary sinus of Valsalva aneurysm with a dilated pulmonary artery has not been reported in the literature. Herein, we describe a case of pulmonary sinus of Valsalva aneurysm who was successfully treated in our clinic.

CASE REPORT

A five-month-old boy presented with a complaint of recurrent respiratory tract infection. Chest X-ray showed a dilated pulmonary artery (Figure 1). Echocardiography showed a large perimembranous ventricular septal defect (VSD), large-sized patent ductus arteriosus (PDA), a dilated main pulmonary artery and branches. Cardiac catheterization showed a right ventricular systolic and mean pulmonary artery pressure of 67 mmHg and 43 mmHg, respectively, and a left-to-right shunt at a ratio of 2.1:1 and a pulmonary vascular resistance index of 2.4 Woods units. Angiography showed a dilated pulmonary artery with an aneurysm of the anterior sinus of Valsalva (Figure 2). The patient underwent a successful closure of VSD and PDA with plication of the sinus of Valsalva aneurysm (Figure 3, 4). The post operative course in the hospital was uneventful and was discharged in stable condition on sixth postoperative day.

DISCUSSION

In the literature review, true pulmonary sinus of Valsalva aneurysm is rare and unusual lesion.[1-4] This anomaly can be seen with associated with

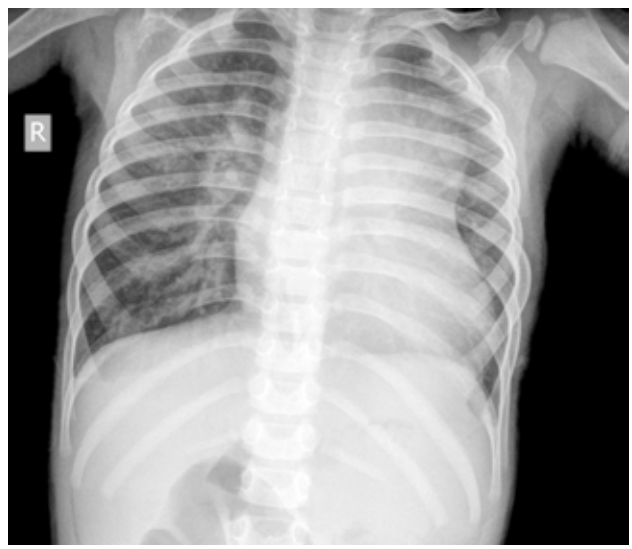

Figure 1. A chest X-ray image showing a dilated pulmonary artery with plethora.

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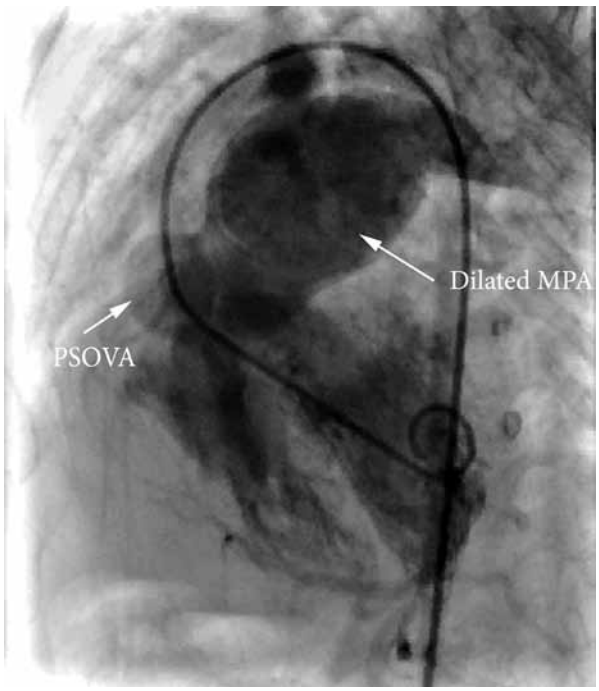

Figure 2. A left ventriculogram image in left anterior oblique cranial view showing VSD with a dilated MPA and an aneurysm of anterior sinus of Valsalva of pulmonary artery.
VSD: Ventricular septal defect; MPA: Main pulmonary artery.

congenital heart disease, pulmonary arterial hypertension, valvar pulmonary stenosis, connective tissue disorders and vasculitis. Other causes include infections (i.e., tuberculosis, syphilis), atherosclerosis, hypertension, hereditary hemorrhagic telangiectasia, cystic media necrosis, and traumas.[1] The pathological cause is intrinsic weaknesses of the arterial wall in combination with an increased hemodynamic stress are responsible for its formation.[2-4] The clinical manifestations are mostly non-specific and symptoms are usually due to associated lesions.[2] Cardiac catheterization and angiography are the gold standards for the diagnosis; however, non-invasive imaging methods including spiral computed tomography angiography and magnetic resonance imaging are also useful tools.[3,4] Surgical intervention is often recommended to symptomatic patients and to those with underlying diseases or complications, left-to-right shunts, pulmonary arterial hypertension, and large-sized aneurysms.[4] In the treatment of low-pressure pulmonary artery aneurysms, intervention is required when the right ventricular size and function alter due to pulmonary regurgitation or pulmonary stenosis. However, asymptomatic, small-sized aneurysms need close follow-up, as the risk of rupture is low, while these lesions requires an intervention if patient is undergoing open heart surgery for associated lesion.[4]

In conclusion, pulmonaty sinus of Valsalva aneurysm is an unusual entity mostly associated with other congenital anomaly. The weakness of arterial wall with increased hemodynamic stress leads to this anomaly. Diagnosis can be achieved by conventional echocardiography and angiography. The computed

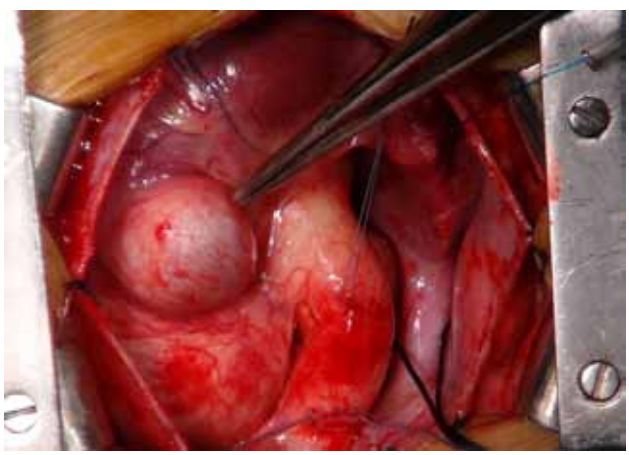

Figure 3. An intraoperative view showing a dilated main pulmonary artery with an aneurysm of sinus of Valsalva.

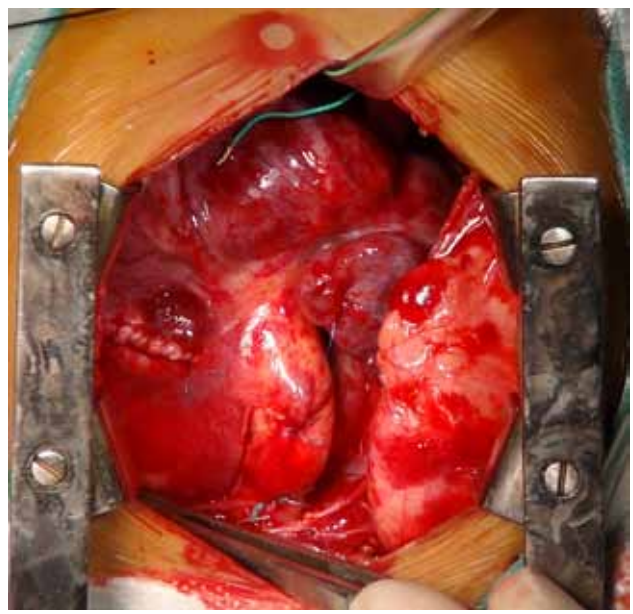

Figure 4. An intraoperative view showing repaired aneurysm of sinus of Valsalva.

tomography angiography and magnetic resonance imaging helps in better delineation of anatomy. Small and asymptomatic aneurysm needs observation while associated congenital heart disease and large aneurysm needs surgical intervention.

Declaration of conflicting interests

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Interesting Image Open Access

A muscular bridge with an absent left main trunk: A rare coronary artery anomaly

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The appearance, prevalence, and the clinical importance of the coronary artery anomalies should be well-appreciated by the cardiologists and the cardiovascular surgeons who are engaged with the coronary artery disease.[1] Coronary artery anomalies are seen approximately 6% of the general population.[2]

An 84-year-old male was admitted to the emergency department with a complaint of angina pectoris. His vital signs were normal, including the blood pressure (130/65 mmHg) and the heart rate (59 bpm). Although his troponin-I levels were within normal limits, coronary angiography was performed to exclude possible underestimation of a coronary artery disease. Although no prominent lesion in the coronary vasculature was detected, a rare anatomic variation was observed. The left coronary arteries were originating from the aorta with a separate ostium having no common left main trunk (Figure 1a, b). The course of the left coronary arteries was normal. However, there was a muscular bridge on the mid-portion of left anterior descending artery (Figure 1c, d). The origin and course of the right coronary artery were normal.

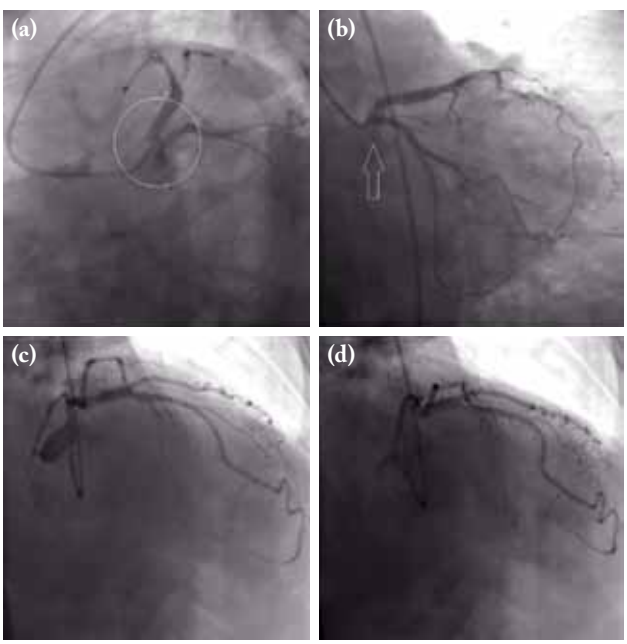

Figure 1. **(a)** A left anterior oblique view showing the absence of the left main trunk. **(b)** Arrow indicates the separate origins of the left anterior descending and the circumflex artery. **(c)** Left anterior descending artery in diastole. **(d)** Left anterior descending artery in systole (Arrow indicates the muscular bridge).

We present this case with an uncommon anatomical variation to highlight the variability of the normally functioning vascular structures. The coronary artery anomalies in which the origin is from wrong coronary sinus may have a risk of sudden death in the younger population.[3] Therefore, this significantly and clinically important condition should be kept in mind, in cases of a coronary artery anomaly.

Declaration of conflicting interests

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Cardiovascular Surgery and Interventions

Interesting Image Open Access

An Iceberg in the heart: A calcified amorphous tumor of mitral valve

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Calcified amorphous tumor (CAT) is a rare and benign pathology which is composed of calcified nodules on a background of a degenerative and inflammatory amorphous fibrous material.[1-3] Although the pathogenesis of cardiac CAT is unknown, abnormal calcium-phosphorous metabolism, particularly in renal failure, is suspected. Differential diagnosis includes benign/malignant cardiac tumors such as a myxoma, teratoma, or rhabdomyoma, thrombosis, and vegetations.

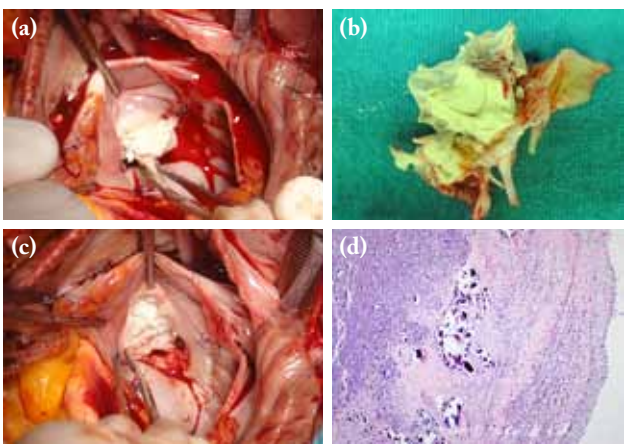

Figure 1. **(a)** An intraoperative view showing extensive calcification of the anterior leaflet of the mitral valve with decreased mobility of the leaflet, leading to severe insufficiency. **(b)** An intraoperative view after resection of the valve showing dense calcification in the periannular area of the anterior mitral leaflet. **(c)** A macroscopic view of the resected mitral leaflets. **(d)** A histological view of the specimen showing the presence of the calcified nodules in an amorphous background of fibrin and dense inflammation with macrophages and plasma cells (H-E x 200).

Herein, we report a 67-year-old case who underwent mitral valve surgery through a transseptal incision with a favorable outcome. During operation, we observed that the anterior leaflet of the mitral valve and its subvalvular apparatus were calcified and presented as a giant tumor (Figure 1). After resection of the calcified leaflet, the annulus of the mitral valve was reconstructed using a pericardial patch, and the valve was successfully replaced with a mechanical prosthesis.

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