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Supramitral Ring An Uncommon Form of Congenital Mitral Stenosis

Supramitral Ring Konjenital Mitral Stenozun Nadir bir Formu

Kalp ve Damar Cerahisi

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Özet

Supramitral ring (SMR), fibromüsküler bir membranla sol ventrikül doluşuna farklı derecede engel oluşturan konjenital mitral darlığının nadir bir alt tipidir. SMR, tüm konjenital kalp malformasyonlarının % 0,6'sını oluşturmaktadır. Bu yazıda SMR, ventriküler septal defekt ve patent duktus arteriosus tanısı konan 5 aylık bir erkek olgu sunuldu. Sorunsuz bir cerrahi müdahale sonrasında hasta ameliyattan 7 gün sonra taburcu edildi. Diğer doğumsal mitral stenoz formlarına göre ameliyat sonrası mükemmel prognoza sahip olduğu için erken tanı konması önemlidir. Bu olgunun sunulmasının amacı SMR konusunda farkındalık geliştirmektir.

Anahtar kelimeler: *supramitral ring*, *mitral stenoz*, *infant*

Abstract

The supramitral ring (SMR) is a rare subtype of congenital mitral stenosis that produces a variable degree of obstruction to left ventricular filling through a fibromuscular membrane. SMR is the cause of 0.6% of all congenital heart malformations. In pediatric age mitral stenosis is a challenging issue nevertheless supramitral ring is a treatable anomaly at most times. Compared to other forms of congenital mitral stenosis, early diagnosis of SMR brings excellent postoperative prognosis. The purpose of this report is to raise awareness of SMR. So, if the pediatric cardiac team's awareness is open then the patient's surgery may be arranged more early. In this paper, we discuss an infant with large left to right defects in association with supramitral ring.

Keywords: *supramitral ring*, *mitral stenosis*, *infant*

Introduction

Supramitral ring (SMR) is a rare anomaly that is among the congenital causes of mitral stenosis [1]. Although, SMR can occur as an isolated disorder, it may appear with many other congenital cardiac anomalies [2]. Besides, it is known that SMR can have different types in itself [2,3]. In this article, the management of the patient who was investigated because of respiratory distress and heart failure and diagnosed with patent ductus arteriosus (PDA), ventricular septal defect (VSD) and SMR was discussed.

Case Report

A five-month-old boy presented to our clinic with the complaints of difficulty in feeding, rapid breathing, sweating and low weight gain. Electrocardiography revealed sinus rhythm and sinus tachycardia (150 beats/min). Telecardiography showed that the plethora in the lung parenchyma and cardiothoracic ratio were enlarged (0,63).

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Transthoracic echocardiography showed an 8 mm diameter non-restrictive malaligment subaortic VSD, patent foramen ovale (PFO) and 4 mm large PDA. Additionally, a fibrous membrane, circulating all the circumference of the valve located 3 mm above the mitral valve, was observed (Figure 1a). Transmitral mean gradient was 20 mmHg. Pulmonary artery pressure was 45 mmHg. Also, the structure of the mitral valve was normal. The membrane that formed the SMR was not adherent to the mitral valve leaflets. The left atrium and left ventricle Z score was +3. Written consent was obtained from the family for surgery, and then the patient was prepared for surgery.

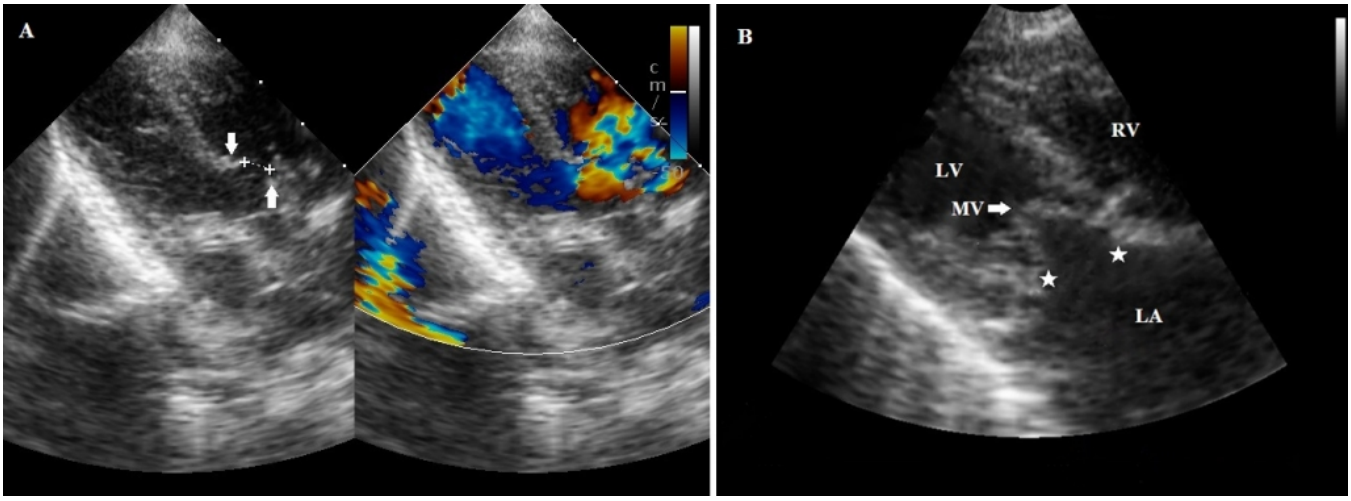

Figure 1

Echocardiographic images. A. Preoperative Transthoracic Echocardiography revealed an additional membranous structure attached to the left atrial wall circumferentially, just above the mitral valve annulus (White arrow). B. Postoperative Transthoracic Echocardiography showed normal Mitral inflow anatomy after resection of the ring.

Mediastinum was approached through a full median sternotomy. Following pericardial incision, PDA looped. Cardiopulmonary bypass was established by ascending aortic and bicaval cannulation. Just after initiation of bypass PDA was ligated. The aorta was cross-clamped and cardiac asystole was achieved using antegrade cardioplegia. Ventricular septal defect closure was accomplished with polytetrafluoroethylene (PTFE) patch using the interrupted suture technique via the right atrium. In the evaluation of the mitral valve by transseptal route, circular fibrotic ring structure was seen 2 mm away from the mitral valve (Figure 2a). The fibrotic ring was completely resected by sharp dissection (Figure 2b).

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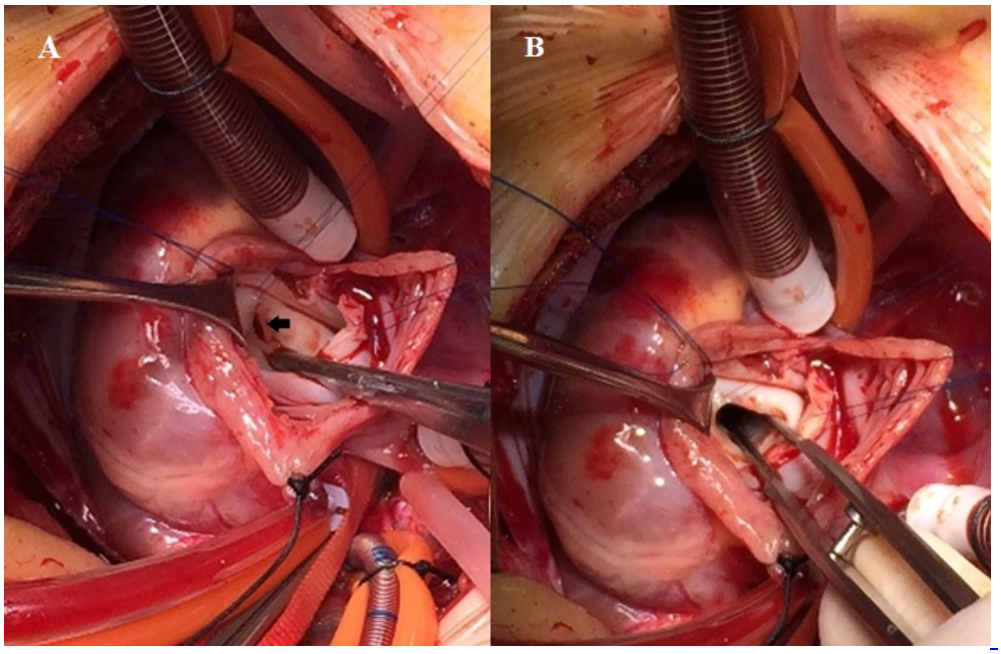

Figure 2
Operative view. A. Intraoperative view of stenotic orifice and circumferential ring (black arrow). B. Intraoperative view after resection of the supramitral ring.

Atrial transseptal incision was repaired primarily. Cardiopulmonary bypass was ended without any problem and without inotropic support. In intensive care follow-up, the patient was extubated at the 6th postoperative hour. The patient had an uncomplicated recovery period after surgery and discharged on the 7th postoperative day without any problem. Echocardiography controls were performed at the 1st and 3rd months after discharge. Pulmonary artery pressure was 30 and 20 mmHg respectively, and the transmitral mean gradient was 6 and 4 mmHg respectively (Figure 1b).

Case Discussion

In literature series, there are two types of mitral ring pathologies, supramitral and intramitral ring. Mitral valve pathology and surgical approach differ in these patients [4]. The supramitral ring is mostly associated with a normal mitral valvular apparatus structure and various congenital heart diseases. Intramitral ring often has an abnormal subvalvular apparatus [4,5]. Supramitral ring pathology has been described within the Shone complex. This anomaly complex described by Shone et al., in 1963 consists of 4 obstructive components: supramitral ring in the left atrium, parachute mitral valve, subaortic stenosis and aortic coarctation. In order to call an anomaly a Shone complex, all 4 components must be found. Shone-like syndrome is usually mentioned if there are two or three components of left-sided obstructive heart defects. In our patient, the gradient in the left ventricular outflow tract was normal (peak gradient: 4 mmHg), and other components were not detected in the preoperative examination and intraoperative observation. In our case, there was a non-restrictive subaortic VSD accompanying SMR and a normal mitral valvular apparatus structure was present. Therefore, our definition of this patient was an isolated supramitral ring. SMR is the rarest cause of congenital mitral stenosis [4]. However, surgical results are better than other types of congenital mitral stenosis. It usually coexists with a wide range (VSD, ASD, Shone Complex) with different cardiac anomalies [3,5]. Therefore, every patient diagnosed with SMR should be examined in detail whether there is an additional cardiac malformation. In a clinical situation detected during infancy and which cannot be explained by VSD, such rare anomalies should be considered. The most notable anomaly in the differential diagnosis of supramitral ring is the cor triatriatum sinister [2,4]. Cor triatriatum is more common than

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SMR. It has been shown that it is caused by a disruption during the development of the left pulmonary veins in the 5th week of embryonic life [2]. Supramitral ring is thought to be caused by the insufficiency of separation of endocardial cushions [2]. Cor triatriatum defined as a fibromuscular structure is a pathology that divides the left atrium in half, leaving a left atrial appendage in the distal region through a membrane. SMR is defined as a congenital stenotic mitral valvular anomaly with a ring of tissue above the mitral valve [4]. Pathophysiological reflection of supramitral ring anomaly causes pulmonary venous hypertension clinic similar to pulmonary venous stenosis, mitral valve stenosis and cor triatriatum sinister [2]. The clinical outcomes worsen with the coexistence of left-to-right shunted cardiac anomalies such as VSD, ASD, PDA, which cause pulmonary hypertension and excessive pulmonary flow [2,6]. As a matter of fact, in addition to SMR, non-restrictive VSD coexistence was present in our case and both pulmonary venous and pulmonary arterial hypertension were observed.

In the surgical method, it can be planned via transseptal route or left atriotomy for access to the mitral valve. If the transseptal method is preferred, both septal defects can interfere and sufficient field of view can be obtained for the SMR. For this reason, it is seen in the literature that the transseptal route is often preferred [2,4]. Complete resection of all components of the SMR with sharp dissection is important. During the resection of the ring, the presence of the circumflex artery extending along the anterolateral margin must be kept in mind and care must be taken to avoid injury. The most important factor affecting long-term follow-up in patients with mitral ring excision is the presence of intramitral type. In these patients, the repair of the mitral valve becomes difficult and the need for reoperation increases in the future [4]. In contrast, the mitral valve structure is usually normal in the mitral ring located just above the mitral valve and accompanying anomalies are often septal defects. For this reason, the results have been reported to be better in these patient groups [2,4].

Among the causes of congenital mitral stenosis, the presence of SMR, with or without concomitant congenital heart disease, can quickly worsen patient's clinic. Therefore, keeping in mind of SMR will ensure early surgical timing in a patient with follow-up due to another congenital heart disease. Especially, late diagnosis of the SMR may cause an increase in mortality and morbidity due to persistence of pulmonary hypertension after surgery, difficulty in leaving the mechanical ventilator and prolonged intensive care unit. Providing the correct diagnosis with an effective preoperative evaluation is very important for the preoperative and postoperative course of the patient.

Declaration of interest: The authors report no declaration of interest. The authors alone are responsible for the content and writing of the paper.

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