



Supravalvar Mitral Ring: a Case Report

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Abstract

Supravalvar mitral ring is a rare congenital heart defect of surgical importance. The condition is characterized by an abnormal ridge of the connective tissue on the atrial side of the mitral valve. It often substantially obstructs the mitral valve inflow. We herein introduce a case of a supravalvar mitral ring in a 17-year-old male, who was admitted to our hospital with cardiac syncope. He had undergone a cardiac operation for ventricular septal defect (VSD) closure and mitral valve repair 15 years before. Transthoracic echocardiography, transesophageal echocardiography, and finally cardiac catheterization revealed a neglected supravalvular mitral ring. The ring was resected in a second operation, and the patient was discharged from the hospital symptom free.

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Introduction

Supravalvar mitral ring is a circumferential ridge or membrane arising from the left atrial wall overlying the mitral valve and frequently attached to the mitral valve.¹ Variable in thickness and extent, the ring ranges from a thin membrane to a thick discrete fibrous ridge.² The membranous variety may be difficult to detect because the membrane often adheres to the anterior mitral valve leaflet while remaining just proximal to the posterior mitral leaflet. Adhesion to the valve may impair the opening of the leaflets, and this impairment may be the main mechanism of mitral valve inflow obstruction in some patients. In other patients, the ring may be large enough to protrude into the mitral valve inflow and cause obstruction. The supramitral ring may initially be incomplete and eccentric, allowing for an unobstructed flow through the

mitral valve. However, turbulence can cause a progressive increase in the supravalvar membrane or ridge thickness, worsening mitral inflow obstruction.³ The same mechanism is responsible for the acquired variety of supravalvar mitral stenosis, which occurs after mitral annuloplasty for the repair of mitral regurgitation.

We herein present a case of a supraventricular ring in a 17-year-old male, who presented with syncope.

Case Report

A 17-year-old male with cardiac syncope was admitted by our hospital. About 15 years ago, he had undergone a cardiac operation for ventricular septal defect (VSD) closure and mitral valve repair.

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In chest X-ray both lungs were clear, and the electrocardiogram result showed normal sinus rhythm, had undergone right bundle branch block, and right ventricular hypertrophy.

Transthoracic echocardiography, transesophageal echocardiography, and finally cardiac catheterization revealed ejection fraction of 50-55%, pulmonary hypertension with pulmonary artery pressure (PAP) of 80/40 mm Hg, pulmonary capillary wedge pressure of 23 mm Hg, gradient in the mitral valve plane of 40 mm Hg, no residual VSD, no patent ductus arteriosus (PDA), bicuspid aortic valve, no aortic insufficiency, no aortic stenosis, no sub-aortic web, normal coronary arteries, mild mitral regurgitation, moderate to severe mitral stenosis, supra mitral valve ring, normal left ventricle size, moderate right ventricle enlargement, and moderate systolic dysfunction.

There was a free floating high redundant obstructive membrane in the left atrium that was attached to the mid portion of the anterior mitral valve leaflet and also in the medial and base of the posterior mitral valve leaflet and large papillary muscles in the left ventricle (Figure 1).

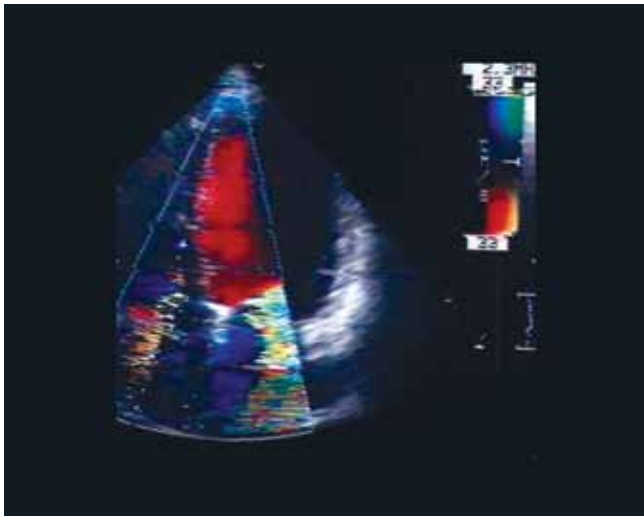


Figure 1. Transesophageal echocardiography (TEE): in the apical four-chamber view, an annular structure is attached like a membrane to the atrial wall and the anterior mitral leaflet, and in close proximity of the posterior leaflet (arrow)

It also extended just adjacent to the lower limb of the left atrium appendage mouth with a small orifice (6 mm) in the lateral side and also multiple small fenestrations (by 2 dimensional echocardiography), resulting in a severely restricted diastolic flow (mean peak gradient = 9.3 mm Hg).

This web was not found in MRI cine turbo-flash images acquired on a 1.5 T scanner (Magnetom Vision-Siemens) in the four-chamber view (3A) and the transverse plane, short-axis view (Figure 2).

The patient's ascending aorta, descending aorta, and aortic arch were tortuous with moderate narrowing of

the descending aorta distal to the subclavian artery with a systolic gradient of about 30 mm Hg, which was suggestive of insignificant coarctation of the aorta.

Doppler of the carotids and sonography of the abdomen and pelvis were both normal. The patient underwent surgery under endotracheal general anesthesia and in supine position. After heparinization, extracorporeal circulation was established between the venae cava and the ascending aorta. Following a median sternotomy, a cross clamp was placed on the aorta. Via antegrade intermittent hypothermic blood cardioplegia from the aortic root, cardiac arrest was established. Hypothermia was mild (30° C). A vent was placed through the right superior pulmonary vein. Standard left atriotomy was made. There was a long supravalvar muscular band, which was resected (Figure 3). Incision of the left atriotomy was closed. The saline test of the mitral valve was almost normal. The postoperative course was uneventful with successful anatomical correction. Postoperative echocardiographic data confirmed complete correction. The patient was underweight and acyanotic after the operation.

Discussion

Supravalvar mitral ring rarely occurs as an isolated defect; other congenital heart defects coexist in 90% of patients. The mitral valve itself is often abnormal and stenotic at the valvar or subvalvar level; fusion of leaflets, a small valve orifice, and abnormal papillary muscles are common abnormalities. Shone's complex is a combination of four congenital heart defects: supravalvar mitral ring, parachute mitral valve, subvalvar aortic stenosis, and aortic coarctation. Other common associated lesions in patients with supravalvar mitral ring are ventricular septal defect (VSD), PDA, atrioventricular (AV) canal defect, and tetralogy of Fallot.⁴

Uncommonly associated defects include atrial septal defect, cor triatriatum, left superior vena cava, unroofed coronary sinus, partial anomalous pulmonary venous drainage, pulmonary venous obstruction, double-orifice mitral valve, and Wolff-Parkinson-White syndrome.⁵ Lesions such as transposition of the great arteries, atrioventricular discordance, and double-outlet right ventricle are occasionally complicated by a supravalvar left atrioventricular valvular ring.

Obstruction to the mitral inflow results from the reduced area of the mitral valve orifice. When clinically significant, a diastolic pressure difference occurs between the left atrium and the left ventricle. Left atrial and pulmonary venous pressures increase, leading to exudation of fluid into the pulmonary interstitial, which increases lung stiffness. Breathlessness and tachypnea are secondary to the interstitial edema and diminished pulmonary compliance.⁶ In severe cases, frank pulmonary edema can occur.⁷ An associated atrial septal defect may decompress the left atrium, reducing

or masking the severity of the mitral valve obstruction. Associated lesions such as VSD or PDA, which increase the left ventricle output, exacerbate the manifestations of mitral inflow obstruction.⁸ Conversely, a supralvalvar mitral ring may be difficult to detect in conditions with a diminished pulmonary blood flow such as Tetralogy of Fallot.⁹ Persistently elevated pulmonary venous hypertension leads to pulmonary arterial hypertension, rise in pulmonary vascular resistance, and eventual failure of the right ventricle and tricuspid regurgitation. There are no data available on the incidence of supralvalvar mitral ring. It also deserves of note that predilection for race, age, and sex has not been found yet.

Treatment is the surgical resection of the web, but balloon dilatation is reported to have been successfully performed under fluoroscopic and transesophageal echocardiographic guidance by some authors.¹⁰



Figure 2. MRI cine turbo-flash images acquired on a 1.5 T scanner (Magnetom Vision-Siemens) in the four-chamber (3A) and transverse plane, short-axis view. These images do not show congenital heart disease.



Figure 3. The supra mitral valvular ring, which was resected from the atrial aspect of the mitral valve

Conclusion

Preoperative identification of a supralvalvar mitral ring is the target for obtaining good surgical results. For the

identification of this congenital heart disease, TEE is not only feasible but also easy.

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