Introduction

Systolic anterior motion (SAM) of the mitral valve is commonly observed in patients with hypertrophic obstructive cardiomyopathy (HOCM) and in few patients after mitral valve repair or aortic valve replacement. It may cause significant hemodynamic instability due to left ventricular outflow tract (LVOT) obstruction and resulting mitral regurgitation. Subaortic septal bulge is considered as one of the risk factors for the development of SAM as it narrows the LVOT. We hereby report a case of tetralogy of fallot with subaortic septal bulge who developed SAM of the anterior mitral leaflet, intraoperatively, after resection of a subaortic membrane.

Case

A 9-year-old male child weighing 16 kg presented to our hospital with chief complaint of failure to thrive and poor weight gain. His preoperative transthoracic echocardiogram (TTE) showed a 12 mm perimembranous ventricular septal defect (VSD), 40% aortic override, and right ventricular infundibular stenosis with a peak systolic gradient of 72 mmHg. In addition, LVOT was partially obstructed by a subaortic membrane with resulting peak LVOT gradient of 50 mmHg. Preoperative cardiac catheterization study revealed normal coronaries and insignificant multiple aortopulmonary collateral arteries. The patient was planned for complete intracardiac repair. In the operating room, after establishing standard monitoring, a 18 G intravenous cannula was inserted on the dorsum of the left hand and a 20 G arterial cannula was inserted in the right radial artery. Anesthesia was induced with intravenous fentanyl, ketamine, and titrated doses of propofol. Injection vecuronium was used to facilitate endotracheal intubation. Maintenance of anesthesia was done with inhalation of isoflurane (1-2%) and oxygen-air (50:50) mixture. A 5.5 French triple lumen central venous catheter was used to monitor central venous pressure. Intraoperative transesophageal echocardiography (TEE) done using Philips iE33 machine and S7-3t TEE probe (Philips, USA) confirmed the finding of TTE [Figures 1, 2 and Video 1]. After achieving adequate anticoagulation with heparin, cardiopulmonary bypass (CPB) was instituted using standard aortobical caval cannulation, roller pump, and a membrane oxygenator. The VSD was closed using polytetrafluoroethylene patch. The subaortic membrane and right ventricular outflow tract (RVOT) were resected and augmentation of RVOT was achieved using the transannular patch. After achieving adequate rewarming, CPB was terminated using milrinone 0.3 mcg/kg/min and...
noradrenaline 0.05 mcg/kg/min. The patient became hemodynamically unstable after few minutes of termination of CPB. The dose of noradrenaline was increased to 0.1 mcg/kg/min and adrenaline 0.05 mcg/kg/min was added. There was transient improvement on hemodynamics; however, the lung compliance decreased and systemic oxygen saturation gradually decreased to 78%. TEE examination revealed LVOT obstruction. Color flow doppler showed turbulent flow originating from the point of contact of the anterior mitral leaflet with interventricular septum [Figure 3 and Video 2]. Application of continuous flow doppler across LVOT showed a dagger-shaped envelope with peak systolic gradient of 78 mmHg at a heart rate of 76/min, suggesting dynamic LVOT obstruction [Figure 2]. Mitral valve assessment showed the presence of severe mitral regurgitation confirming SAM of the anterior mitral leaflet [Figure 3 and Video 2]. The maximum interventricular septal thickness at subaortic and midventricular septum level was 17 mm and 10.6 mm, respectively [Figure 4]. The minimum distance from the coaptation point to the septum (C-Sept distance) was 18 mm. Initially, the patient was managed with stopping all inotropes, administration of adequate fluid, and increasing afterload with titrated doses of vasopressin up to a maximum of 2 units/h; however, despite these measures, patient’s hemodynamics and lung compliance did not improve. Hence, the decision for surgical septal myectomy was taken. About 8 mm septal tissue as calculated by the difference between maximum interventricular septal thickness and midventricular thickness on TEE was excised under CPB. TEE examination after septectomy showed mild mitral regurgitation and LVOT gradient decreased to 16 mmHg [Figures 2, 5 and Video 3]. The patient remained hemodynamically stable after termination of CPB. He was shifted to intensive care unit (ICU) for elective mechanical ventilation. The lung compliance improved slowly and the patient was weaned from mechanical ventilation after 48 h. He was shifted to the ward after 4 days of uneventful stay in the ICU.

**Discussion**

The SAM of the mitral valve is a paradoxical motion of mitral valve leaflets towards the LVOT during the systole.[3] Initially, it was considered specific for HOCM; however, it has been associated with any condition that alters the dynamic, complex anatomy of the left ventricle (LV).[4] Besides HOCM, it has been reported to occur in various other conditions such as accessory papillary muscle, cleft anterior mitral leaflet, postaortic valve replacement for aortic stenosis, mitral valve repair, Takotsubo cardiomyopathy, during dobutamine stress echocardiography and, postmyocardial infarction.[1,4] Various described factors that predispose for the
development of SAM in adults includes excessive anterior or posterior mitral leaflet tissue; aortomitral angle <120°; elongation and buckling of chordae; anterior and medial displacement of the papillary muscles; bulging of subaortic septum; absolute height of the posterior leaflet >1.5 cm; anterior to posterior leaflet height ratio <1.4; minimum distance from the coaptation point to the septum (C-Sept distance) <2.5 cm; and basal septal hypertrophy (BSH). In the index case, only predisposing factor observed on TEE was BSH and decreased C-Sept distance.

BSH is diagnosed when the diastolic basal ventricular septum thickness is >14 mm and the ratio of diastolic basal to mid-septal thickness is >1.3. It may mimic as asymmetric septal hypertrophy seen in HOCM. Waller et al. showed that the basal portion of the ventricular septum bends towards LV and bulges into the LVOT as a result of the rightward shift of dilated ascending aorta. Others suggested that BSH is a nonpathological result of aging and has been related to aortic stenosis and hypertension. Diaz et al. reported that BSH is not associated with cardiovascular risk. However, in a case series of 21 adult patients with BSH, 6 patients developed clinical and echocardiographic features of LVOT obstruction during follow-up. Said et al. suggested that bulging of the subaortic septum may be a risk factor for the development of SAM after mitral valve repair and transaortic septal myectomy should be considered at the time of valvuloplasty if the ratio of basal septal thickness to midventricular septal thickness is >1.3.

In our case, there was no history of HOCM in the family. However, there was bulging of the subaortic septum. Bulging subaortic basal septal thickness measured 17 mm, whereas mid-septal thickness measured only 10.6 mm. The subaortic membrane induced increased LV afterload may explain the underlying mechanism for the development of hypertrophied subaortic septum. Due to the overriding and rightward shift of dilated ascending aorta in TOF, the thickened subaortic septum may have bend towards LV and bulged into LVOT resulting in the narrowing of LVOT in our case. Despite narrowing of LVOT, SAM did not manifest before intracardiac repair as the subaortic membrane maintained LV afterload. Resection of subaortic membrane may have caused acute reduction in afterload resulting in the development of SAM, due to the venturi effect. Severe LVOT dynamic obstruction and severe mitral regurgitation due to SAM resulted in hemodynamic instability.

The spectrum of presentation of SAM may vary from clinically silent to hemodynamically significant disease, which may occur due to LVOT obstruction and/or resulting mitral regurgitation. In the majority of cases, SAM resolves with medical management; however, in refractory cases, surgical management may be needed. In patients with...
subaortic septal hypertrophy bulging towards LVOT, septal myectomy increases the distance between the anterior mitral leaflet and the septum, resulting in the reduction in LVOTO. In our case, perioperative medical management failed to improve hemodynamics. Hence, septal myectomy was done after re-instituting CPB. The patient was successfully weaned off CPB with stable hemodynamics.

To conclude, patients of TOF with subaortic membrane may develop SAM of mitral valve after intracardiac repair and resection of the subaortic membrane. Preoperative echocardiographic examination for the assessment of risk factors for the development of postoperative SAM may help in deciding the appropriate surgical plan.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

**Conflicts of interest**

There are no conflicts of interest.

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