Case Report

Congenital Sub-Mitral Aneurysm: Anesthetics and Surgical Management

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ABSTRACT

A sub-mitral left ventricular aneurysm is a rare condition. It is a congenital outpouching of the left ventricular wall, invariably occurring adjacent to the posterior mitral leaflet. Sub-mitral aneurysm (SMA) has usually been reported as a consequence of myocardial ischemia (MI), rheumatic heart disease, tuberculosis, and infective endocarditis. Nevertheless, there have been few case reports of congenital SMA in India. It usually presents with symptoms of heart failure. We report a rare case of congenital SMA in a 27-year-old young Indian and its successful management through a trans-aneurysmal approach.

Keywords: Aneurysmorrhaphy, congenital sub-mitral aneurysm, mitral valve replacement, transesophageal echocardiography

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INTRODUCTION

Sub-mitral aneurysm (SMA) is a rare condition characterized by the presence of an aneurysm near the posterior mitral leaflet. It is more commonly seen in African blacks but has also been reported in people of all races, including Indians.[1] The condition typically occurs due to a weakened fibrous annulus of the mitral valve, which can be congenital or predisposed by factors such as rheumatic heart disease, tuberculosis, infective endocarditis, ischemia, or trauma.[2-4] The true incidence of the disease is underestimated because early stages often do not present with clinical symptoms. Patients with SMA typically present at a young age, and the most common symptom is mitral regurgitation (MR), which can progress to heart failure. We report a rare case from central India of a congenital SMA, in which the patient underwent successful aneurysm repair along with mitral valve replacement.

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CASE PRESENTATION

A 27-year-old man, weighing 49 kg, presented with a recent onset of chest pain, breathlessness -New york heart association Intravenous (NYHA IV), and other symptoms suggestive of congestive heart failure. Initially, he was diagnosed with inferior wall myocardial ischemia (MI) and ischemic mitral regurgitation (MR) and referred to a higher center for further treatment. There was no history of specific triggering events such as rheumatic fever or infective endocarditis. Upon clinical examination, the patient had a heart rate of 108/min, low blood pressure (78/64 mm hg), feeble peripheral pulses, tachypnea, and room air saturation of 86%, which improved to 96% with supplemental oxygen via a face mask. Coarse crepitations were heard throughout the lung fields, and a grade 5/5 pansystolic murmur was heard at the

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mitral area, radiating to the back and axilla. Preoperative hematological (Hb 10.2 gm%) and biochemical results were within normal limits except for a slightly elevated serum creatinine level (1.2 mg). Electrocardiography showed ST-T- ST & T wave of ECG changes in the inferior leads. Chest radiography revealed cardiomegaly with bilateral pleural effusion and interstitial edema.

Transthoracic echocardiography revealed a large sub-mitral true aneurysm, with the posterior mitral leaflet being sail-like, prolapsing into the left atrium (LA), causing severe MR. The aneurysm's neck diameter measured around 33 mm and had a size of 30 × 36 mm, with mild pericardial effusion. Other findings included dilated LA (40 mm), a left ventricular internal diameter of 38 mm, and an ejection fraction of 40% [Figure 1B and Video 1]. Computed tomography (CT) angiography confirmed the presence of focal saccular dilation of the left ventricular cavity in the sub-mitral region, with no significant coronary artery disease [Figure 1A]. The patient was initially monitored and stabilized in the intensive care unit (ICU) with diuretics, beta-blockers, and oxygen through a face mask. Two days later, he underwent surgery.

The patient was induced with intravenous morphine (0.1 mg/kg) and a titrated dose of IV propofol. Muscle relaxation was achieved with an IV vecuronium bromide at 0.1 mg/kg body weight, and the trachea was intubated with a cuffed endotracheal tube. Anesthesia was maintained with oxygen (FiO2 40%), air, and sevoflurane (up to 2%) administered intermittently. Continuous monitoring of Electrocardiogram (ECG), direct arterial blood pressure, and central venous pressure (CVP) was performed.

The pre-bypass period was uneventful. Transesophageal echocardiography (TEE) confirmed the presence of SMA below the posterior mitral leaflet, communicating with the -left ventricle (LV) cavity with a wide neck of size of 2.5–3 cm. LA and LV were dilated [Figure 2a and b, e and f and Videos 2 and 3]. LA pressure was high with pulmonary vein Doppler, showing reversal of the systolic & Diastolic (S/D) ratio (S < D) with an A-wave duration of 207 ms, mild tricuspid regurgitation (TR), and pericardial effusion [Figure 2d]. Severe MR with two distinct jets contributed to MR, a larger one from LV to SMA and another smaller one from LV to LA [Figure 2c and Videos 4-6]. Therefore, minimal LV ejection entered the systemic circulation, leading to cardiogenic shock.

Intraoperatively, a globular swelling of size 5 × 4 cm was observed externally on the posterior wall of the LV [Figure 3a]. The mitral annulus showed changes in tissue architecture. A large SMA, with a cavity lined with

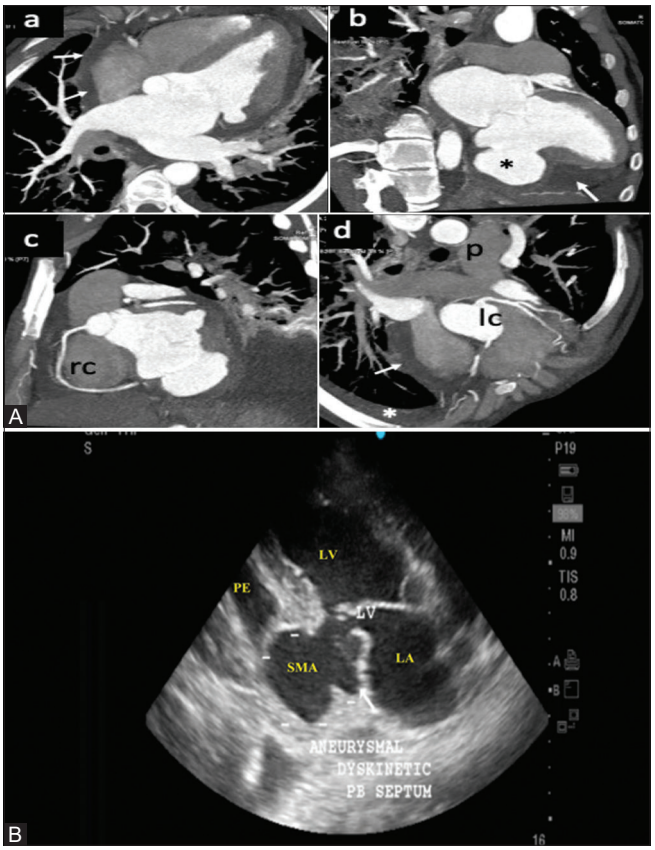


Figure 1: [A] Cardiac-gated CT angiography images: (a) four-chamber view, (b) two-chamber view, (c) oblique sagittal view, and (d) oblique axial view. The images show pericardial effusion (white arrows shown in a, b, and d). Four chambers (a) show pericardial effusion and a mildly dilated left atrium. The two-chamber view (b) shows a ventricular aneurysm (black asterisk) projecting posteroinferior behind the posterior mitral leaflet with a peripheral wall and thrombus. The normal coronaries (rc = right coronary and lc = left coronary) are demonstrated in images (c and d, respectively). Pericardial effusion (white arrow) and pleural effusion (white asterisk) are clearly demonstrated in the image (d). It also shows the prominent pulmonary arteries (on measuring, they were rpa = 22 mm and lpa = 27 mm) suggestive of pulmonary arterial hypertension. [B] Preoperative TTE, parasternal long-axis view, showing a subpericardial echo-free space below the posterior mitral leaflet, that is, a large sub-mitral aneurysm communicating with the left ventricle

organized mural clots, was surgically addressed through a trans-aneurysmal ventriculotomy [Figure 3b]. The neck of the aneurysm cavity, communicating with the LV, was closed using a Dacron patch, the aneurysm wall was closed with aneurysmorrhaphy using a polytetraflouroethylene (PTFE) patch, and the mitral valve was replaced with a 29 St. Jude bi-leaflet mechanical mitral valve [Figure 3c]. Hematocrit on bypass was maintained between 28% and 30%. The patient was gradually weaned from cardiopulmonary bypass (CPB) with an infusion of dobutamine (0.5 to 0.7 μ/kg/min) and noradrenaline (0.02 to 0.05 μ/kg/min). Immediately after CPB, his systolic blood pressure was around 90 mm Hg. The bypass time was 248 min, and the ischemia time was 217 min.

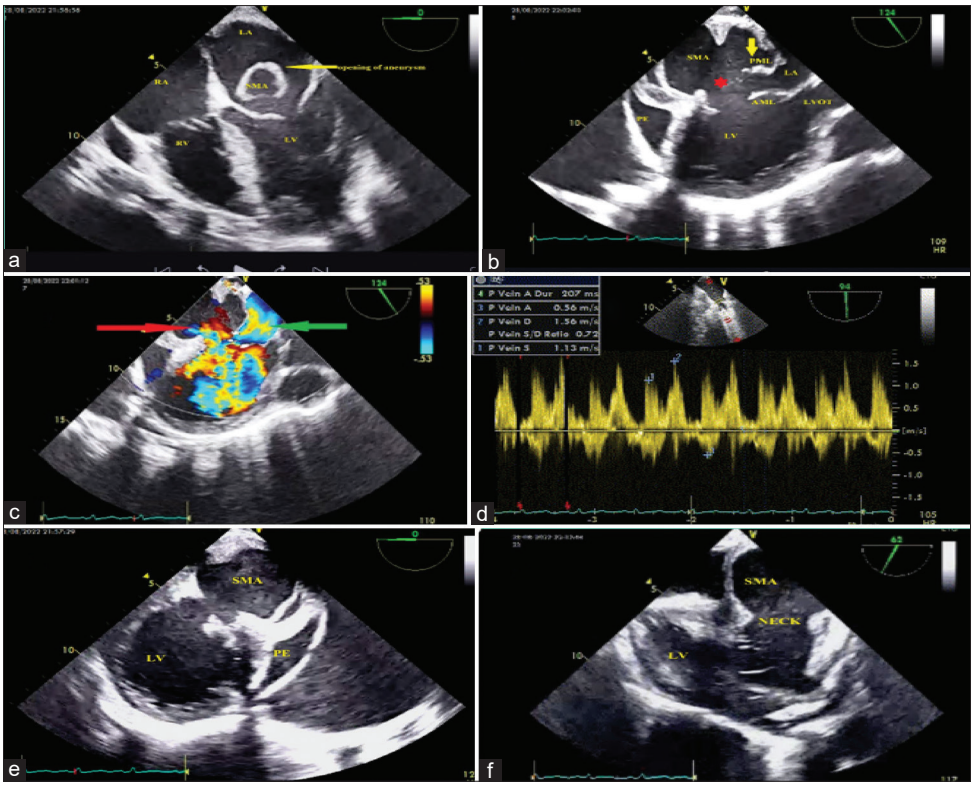


Figure 2: (a) Mid-esophageal four-chamber view, with an arrow pointing to the neck of sub-mitral aneurysm (b) Mid-esophageal long-axis view illustrating that the SMA is originating behind the posterior mitral leaflet (arrow) with a well-defined neck. A red asterisk showing neck of sub-mitral aneurysm (c) Mid-esophageal view with color Doppler showing the mitral regurgitation jet into SMA (red arrow) and LA (green arrow) (d) Pulmonary vein Doppler flow, showing reversal of flow, that is, D > S (e) Transgastric view, with deformed LV shape/figure of 8 appearance of LV due to SMA before surgery (f) Transgastric two-chamber view, showing SMA involving the inferolateral wall of LV

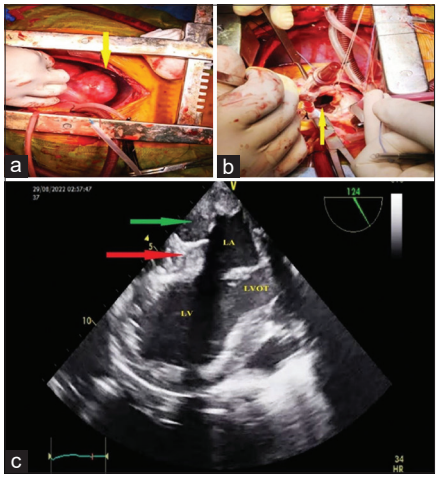
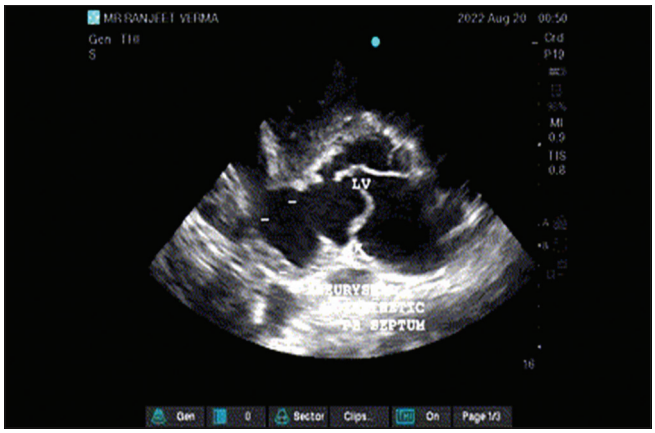


Figure 3: (a) Intraoperative image (yellow arrow) showing SMA swelling at the posterior–inferior part of LV (b) Intraoperative image (yellow arrow) showing the neck of SMA (c) Post-CPB, TEE, and mid-esophageal long-axis view showing a Dacron patch closing the mouth of an aneurysm (red arrow) and aneurysmal wall closure by the PTFE graft (green arrow)

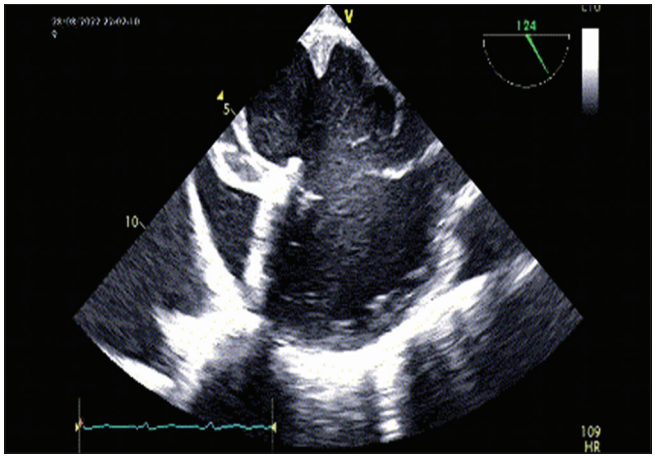
Post-CPB, TEE showed that the prosthetic mitral valve functioned well, with mild MR, and the mean gradient across the valve was <5 mm Hg. Moreover, a trivial TR with an right ventricle systolic pressure (RVSP) of 26 mm Hg was noted. A regional wall motion abnormality (RWMA) was observed in the inferior wall of the LV, and the calculated ejection fraction (EF) was 40% using the Simpson method [Videos 7-9].

He received one unit of packed red blood cells, two units of fresh frozen plasma (FFP), and two units of platelets in the postoperative period and was extubated after 16 hrs. Inotropic support was gradually tapered off; he was started on tablet acitrom 4 mg on alternate days and tablet ecosprin 150 mg daily, and the rest of his recovery was smooth. Follow-up evaluations, including CT angiography and echocardiography, showed near-normal findings. He was discharged after 14 days in stable condition with normal sinus rhythm, with advice to continue anticoagulant medication and follow a low-vitamin K diet.

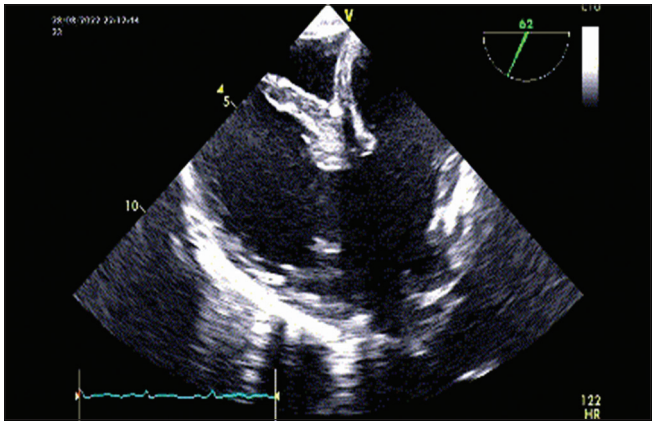
A week later, during a follow-up visit, the patient was doing well and had normal daily function. However, a month after surgery, he experienced black-colored stool at home, followed by headache, dizziness, and unconsciousness. He was hospitalized and received transfusions of two units of FFP due to coagulation abnormalities. Unfortunately, he died of cardiorespiratory arrest.



Video 1: Preoperative TTE, parasternal long-axis view, showing a subpericardial echo-free space below the posterior mitral leaflet, that is, a large sub-mitral aneurysm communicating with left ventricle

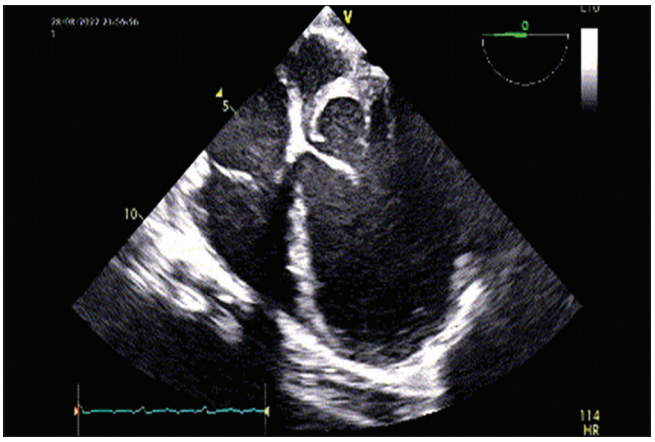


Video 3: Mid-esophageal long-axis view illustrates that the SMA originates behind the posterior mitral leaflet with a well-defined neck

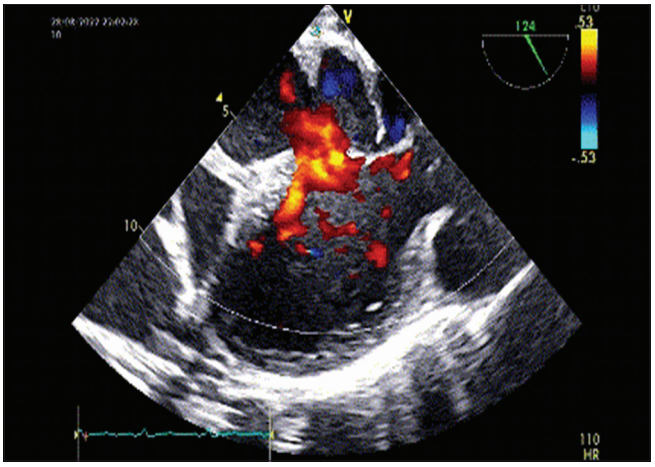


Video 5: Transgastric two-chamber view, showing SMA involving the inferolateral wall of LV

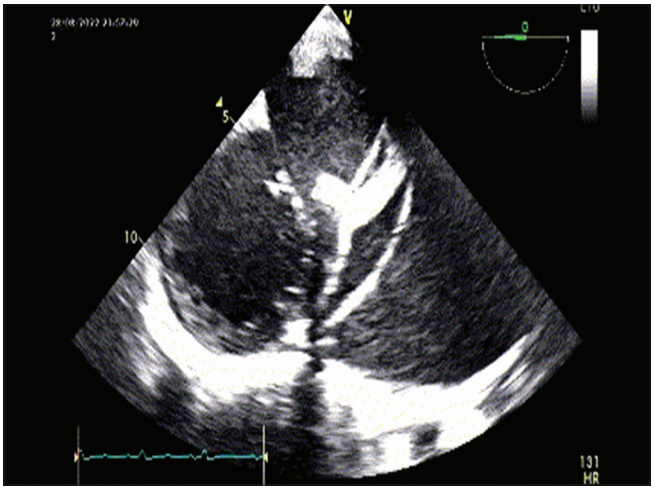
Histopathological examination of mitral tissues revealed areas of fibrosis and focal myxoid changes without active inflammation or calcification. The aneurysmal wall, lined by a flattened epithelium and subepithelial tissue, showed congested blood vessels and inflammatory cell infiltration.



Video 2: Mid-esophageal four-chamber view, showing the opening of sub-mitral aneurysm



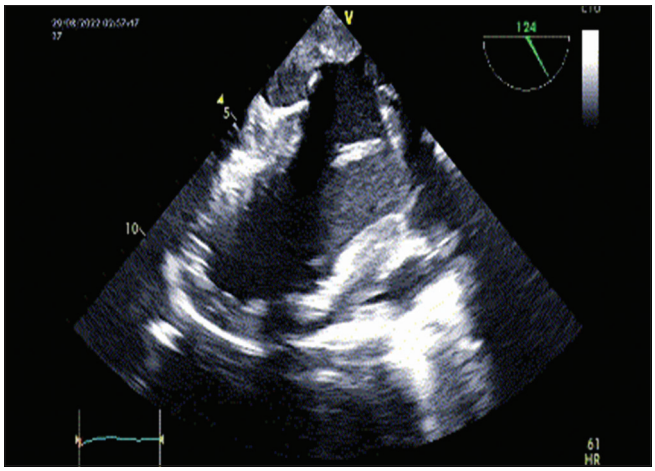
Video 4: Mid-esophageal view with color Doppler showing the mitral regurgitation jet into SMA and LA



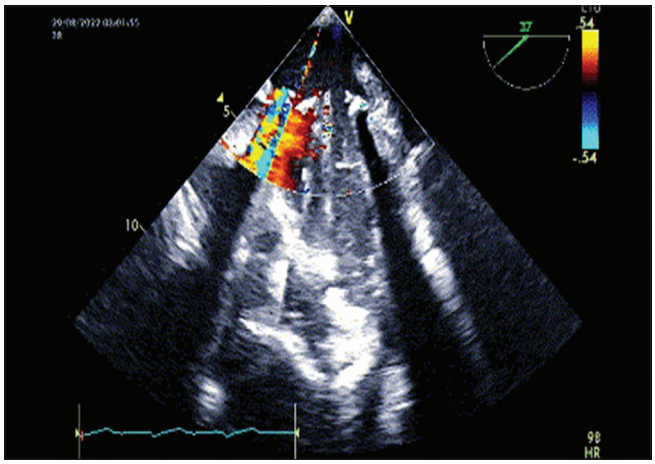
Video 6: Transgastric view, with deformed LV shape/figure of 8 appearance of LV, due to SMA before surgery

DISCUSSION

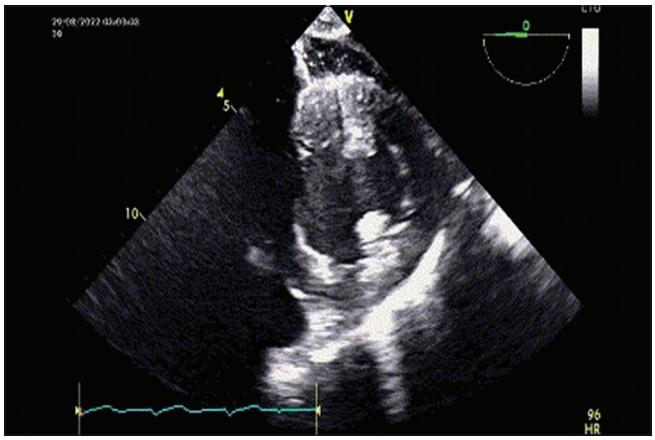
We report a case of a rare condition called congenital SMA from the central part of India. This patient did not have



Video 7: Mid-esophageal long-axis view showing a Dacron patch closing the mouth of an aneurysm and aneurysmal wall closure by the PTFE graft



Video 8: Mid-esophageal mitral commissure view, showing moving leaflets of replaced mitral valve



Video 9: Transgastric view, LV shape after surgery

any other underlying pathologies commonly associated with SMA, such as rheumatic carditis, tuberculosis, syphilis, infective endocarditis, Takayasu arteritis, or polyarteritis nodosa.[2,5-7] While genetic predisposition has been suggested as a possible cause, we were unable to evaluate the patient's genetic condition due to financial constraints. The patient denied any known cardiac diseases in their family. SMA is characterized by the presence of an aneurysm near the posterior mitral leaflet.[8] It is classified into three types based on the extent of involvement of the posterior mitral annulus: type 1 with a single neck, type 2 with multiple necks, and type 3 involving the entire posterior portion of the annulus. Our case belonged to type 1, involving the posterior basal sub-mitral region.

Clinical manifestations of SMA can vary from an incidental finding in an asymptomatic patient to more severe symptoms such as congestive heart failure, severe MR, thromboembolism, ventricular arrhythmias, MI, and even sudden death.[9,10] Most patients present at an early age. Due to the rarity of the disease, initially, our patient was misdiagnosed with inferior wall MI, leading to ischemic MR. However, considering the patient's young age, normal coronaries, the aneurysm's location below the posterior mitral leaflet (PML), and nonspecific histopathology findings exclude ischemia, and diagnosis of congenital SMA was made.

These are sick subsets of patients and require a multidisciplinary, integrated approach for better outcomes. Treatment involves initial stabilization with medical therapy, followed by early surgical repair. Although surgical outcomes are poor, open heart surgery is the treatment of choice.[2] Two common techniques are used: an extracardiac repair, where the aneurysm is accessed from the epicardial side, and intracardiac repair, which involves accessing the aneurysm via the LA through the interatrial groove.[11] In our patient's case, the big size, narrow neck, and densely adhered clot to the aneurysm necessitated the use of an extracardiac technique to remove the clot and repair the aneurysm.

The morphine-based anesthetic technique is safe and effective for maintaining hemodynamics in these patients. In MR, the goal is to achieve relative tachycardia, reduced afterload, adequate preload, and contractility to minimize the regurgitant flow and ensure hemodynamic stability.[12,13] Although the combination of morphine and vecuronium can potentially cause bradycardia, it was not observed in our patient, and the period before the bypass was uneventful.

Monitoring the patient's ECG (ST-segment and T-wave changes) throughout the perioperative period is mandatory, as MI is common due to compression of the circumflex coronary artery by the aneurysm or injury to its branches during surgery. The inevitable prolongation of ischemic

time can further compound this problem. It might have necessitated inotropes in our patient for weaning from the bypass.

TEE plays a crucial role in confirming the diagnosis of SMA, assessing the location and size of the aneurysm, evaluating its relationship with the left-sided chambers of the heart, determining the mechanism of MR, and identifying associated complications. TEE also helps in the assessment of mitral valve repair or replacement and assists in de-airing the cardiac chambers before aortic unclamping.

Previous case reports have shown varying outcomes for patients with congenital SMA, with or without surgery. In a case report by Rajesh *et al*., SMA in a 10-year-old girl underwent an extracardiac approach to aneurysm repair and has had a successful repair and good postoperative recovery.[14] In another case report by Jetley *et al*., the SMA was in a 14-year-old boy who underwent mitral valve repair and resection of an aneurysm that did not come off bypass and died.[15] Also, Rajendra *et al*. reported a huge SMA in a 24-year-old man who succumbed to the condition before corrective surgery could be attempted.[16] In our case, despite early surgical intervention and an uneventful immediate postoperative course, the patient developed hemorrhagic coagulation abnormalities one month later due to oral anticoagulant therapy, which ultimately led to his demise.

CONCLUSION

Congenital SMA, although rare, can be a differential diagnosis of MR with left ventricular dysfunction and heart failure in the young.

Managing patients with congenital SMA requires a thorough understanding of the disease's pathophysiology, initial stabilization of the patient's condition using medical therapy for heart failure, timely surgical intervention, optimization of the anesthetic technique, and careful consideration of associated problems such as MR, MI, left ventricular dysfunction, prolonged bypass, and ischemic times. A multidisciplinary approach is vital for the successful management of these complex cases.

Regular follow-up, counseling, and education are crucial for patients with prosthetic valves on oral anticoagulants to detect and manage potential hemorrhagic complications, as they can adversely affect both mortality and morbidity outcomes.

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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