Supramitral Membrane with Left Atrial Appendage Thrombus: A Rare Entity

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ABSTRACT
A 4-year-old child with supramitral membrane (SMM) causing severe mitral stenosis (MS) was taken for excision of the membrane. Intraoperative transesophageal echocardiography showed a large thrombus in the left atrial appendage (LAA) in addition to SMM. The case underscores the importance of this extremely rare association and prompt therapy to prevent catastrophic consequences.

Keywords: Congenital mitral stenosis, left atrial appendage thrombus, supramitral membrane

INTRODUCTION
Supramitral membrane (SMM) or supramitral ring, a rare cause of congenital mitral stenosis (MS), is characterized by an abnormal connective tissue on the atrial side of the mitral valve, which often obstructs mitral valve inflow. In 90% of the cases, it is associated with other congenital anomalies such as Shone’s complex, ventricular septal defect, subaortic stenosis, or coarctation of aorta.[1,2] Patients with MS, especially those with atrial fibrillation, are more prone to thrombus formation in the left atrium (LA) or left atrial appendage (LAA). The prevalence of LA thrombus in patients with MS is reported as 26.1% and 13.5%, respectively, in the presence and absence of atrial fibrillation.[3] While the presence of LA thrombus in adults with severe MS is quite common, the association of SMM with LAA thrombus in a child has not been described to the best of the author’s knowledge.

CASE REPORT
A 4-year-old female child, weighing 9.8 kg, presented with frequent respiratory tract infections and failure to gain weight. The baseline parameters were heart rate 130 beats/min, sinus tachycardia, blood pressure 94/56 mm Hg, and peripheral pulse oximetry saturation 97%. Two-dimensional transthoracic echocardiography (TTE) showed a thick SMM adherent to both anterior mitral leaflet (AML) and posterior mitral leaflet (PML), severe MS (pressure gradient 38/19 mmHg), enlarged LA (2.65 cm), no mitral regurgitation, severe tricuspid regurgitation, bilateral superior vena cava (SVC), left SVC to coronary sinus, and no coarctation of aorta. The child was taken up for excision of the SMM and inspection of the mitral valve. Intraoperative TEE (Philips pediatric biplane S7-3t probe, iE33 system) showed a thrombus in the LAA measuring 1.3 × 0.9 cm [Figure 1a and Video 1], a thick SMM covering the entire mitral annulus [Figure 1b], and severe MS (pressure gradient 28/16 mmHg) [Figure 1c]. The surgical procedure consisted of circumferential excision.
Singh: Supramitral membrane with LAA thrombus

Figure 1: Mid-esophageal (ME) transesophageal echocardiography images showing: A thrombus in LAA, enlarged LA, and left ventricle (LV) in 2-C view (a), SMM, AML, and PML in 2-C view (b), and continuous wave Doppler showing a transmitral gradient of 26/18 mmHg (peak/mean, (c))

of SMM and removal of LAA thrombus via left atriotomy. Postoperative anticoagulation was achieved with heparin and the patient was discharged on oral ecosprin 37.5 mg once a day. Postoperative follow-up at 1 and 6 months showed clinical improvement and normal echocardiogram.

DISCUSSION

SMM causing congenital MS, is a rare malformation, found in about 1.5% of autopsied hearts with congenital heart diseases.\[2\] It is anatomically characterized by the presence of a stenosing ring in the form of a shelf-like membrane above the mitral valve. It results from a failure of the endocardial cushions to divide completely. The mitral valve may be normal or deformed. This condition must to be distinguished from cor triatriatum, in which a membrane divides the LA into two chambers: a proximal chamber, that receives the pulmonary veins and a distal chamber that gives rise to the LAA. The membrane is characteristically present below the LAA and foramen ovale in SMM, while it is above the LAA in cor triatriatum.\[4\]

TEE is perhaps the best tool to diagnose SMM, and certainly better than TTE to diagnose LAA thrombus. TEE uses high-frequency probes and can visualize posteriorly located structures such as LA, LAA, and interatrial septum better than TTE.\[5\] A thrombus located in the LA or LAA is the most prevalent source of cardioembolic events and adverse neurologic outcomes. Prompt therapy for LAA thrombus includes its correct diagnosis, immediate surgical removal and/or therapeutic anticoagulation. Embolism from the heart can lead to transient ischemic attacks, stroke, or occlusion of peripheral arteries. In many echocardiography laboratories, evaluation for a source of embolism is the most common indication for TEE. The anatomic and hemodynamic changes contributing to thrombus formation in LA/LAA include enlarged volume of LA, stasis of blood, low cardiac output, edema, endothelial denudation, impaired extracellular matrix turnover, and abnormalities of coagulation cascade.\[3\] Surgical resection is the preferred method of treatment for SMM, with little risk of mitral regurgitation. Balloon mitral valvuloplasty does not offer favorable results and is not advisable in the presence of LAA thrombus. Previous studies and case series have reported excellent immediate and late outcomes after surgical resection of the SMM, with little risk of recurrence after surgery.\[6,7\]

In conclusion, the case underscores the importance of this extremely rare association in a child with sinus rhythm, and prompt therapy to prevent catastrophic outcomes.

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Conflicts of interest
There are no conflicts of interest.

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