Surgical Correction of Isolated Discrete Sub-Aortic Membrane : A Case Report

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Abstract
The association between discrete sub-aortic membrane are frequently associated with other sub-aortic anomalies which is reported less often, and till today the surgical correction is considered challenging. An 18-year-old, young lady presented to us with the diagnosis of a discrete sub-aortic membrane producing severe Left Ventricular Outflow Tract Obstruction (LVOTO). We excised the membrane surgically and patient showed excellent symptom free recovery. Per-operative Trans-Esophageal Echocardiogram (TEE) and post-operative Transthoracic Echocardiogram (TTE) showed mild residual membrane and minimum LVOT pressure gradient.

Key words: Congenital anomaly; Left Ventricular Outflow Tract Obstruction (LVOTO); Sub-Aortic Membrane (SAM).

Introduction
The clinicopathological spectrum of stenosing aortic valves has classically been sub-divided into valvular, sub-valvular and supra-valvular stenosis. Amongst those, supra-valvular stenosis is the rarest, whereas valvular stenosis in the commonest.[1] Sub-aortic stenosis is responsible for left ventricular outflow tract obstruction in 8-20% of congenital cases.[1] This stenosis usually takesa discrete type (Membranous or fibromuscular type) or may take tunnel type.[2] A crescentic fibrous curtain, either attached to the ventricular septum or completely encircling the Left Ventricular Outflow Tract (LVOT) is present in the discrete type. It can be located anywhere in the LVOT, just immediately below the aortic valve to about ≥10 mm towards the left ventricle.The persistence of this discrete ridge or membrane typically causes features of LVOT obstructions after the first decade of life.[3]

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Submitted on : 01.07.2023
Accepted on : 18.11.2023

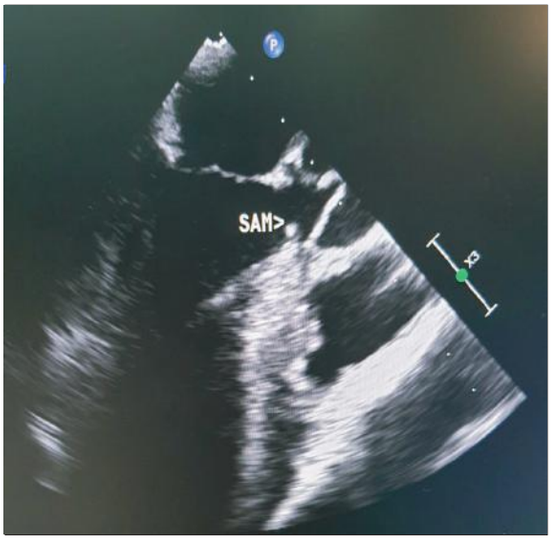

Figure 1 Preoperative echocardiography showing a discrete SAM

Case Presentation
An18-year-old girl presented at National Heart Foundation of Bangladesh, Dhaka on 10th July 2022 with the complaints of respiratory distress and palpitation for last few years and mild chest pain for last 5 months. On examination, she was having a systolic murmur which was best heard at her aortic area. Her X-ray chest and ECG were within normal limits. Patients' echocardiography revealed a discrete sub-aortic membrane adjacent to her aortic valve annulus, causing severe LVOT obstruction (PPG 105mm of hg). The aortic valve leaflets showed good systolic opening with mild AR. Echocardiography also showed severe concentric LV hypertrophy and biventricular functions.

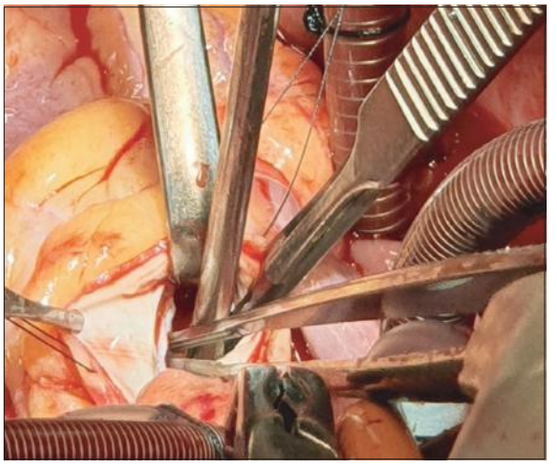

Figure 2 SAM visible through aortomy

After proper planning she was put on CPB with standard bi-caval cannulation though median sternotomy. Right atrium was opened and left ventricle was vented through inter-atrial septum. An oblique aortotomy was then done and aortic valve was identified. Through the aortic valve orifice, near the aortic valve annulus a circumferential sub-aortic membrane was identified. The membrane was then completely excised out taking great care not to injure the aortic valve leaflets. After satisfactory resection both aortic and mitral valves were checked with saline showing acceptable results. Both Aortotomy and RAtomy wounds were then closed and patient was weaned off from CPB to normal sinus rhythm. As satisfactory hemostasis was achieved chest was closed keeping temporary RV pacing wire and chest drain tubes in situ. She was then shifted to ICU in stable hemodynamic status. Later on, the same day she was extubated from mechanical ventilator on a stable hemodynamic condition.

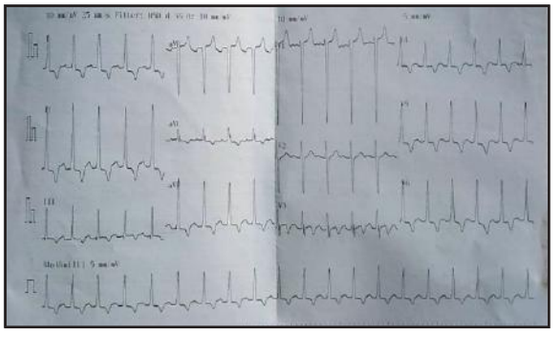

Figure 3 Post-operative ECG of the patient

The patient did well during the post operative follow up period. Per discharge echocardiography revealed a small residual SAM with LVOTO with trivial to mild valvular AR. Moderate concentric LV wall hypertrophy with good biventricular systolic function. The patient was discharged on 7th POD.

The authors have taken appropriate consent from the patient and patient attendants, that their images and other clinical information to be reported in the journal.

Discussion

Cheevers (1884), first described the discrete sub-aortic membrane and initially it was thought to be a congenital anomaly, due to failure of bulbus cordis degeneration.[4] But the theory of acquired origin, advocated by Somerville ultimately got substantial support.[5] When compared with congenital tunnel type sub-aortic obstruction, the discrete stenosis is thought to be developed because of hemodynamic and rheological abnormalities of the LVOT.[6] This membrane (SAM) can be isolated or may be accompanied with congenital ventricular septal defect.[7] Moreover, supplementary mitral valve anomalies can add to the obstructions produced by the sub-valvular ridge.[8] The natural history of SAM is usually variable, and in most of the cases the lesion advances to produce LVOTO before adolescence, nonetheless in few cases it may remain symptomless. The involvement of anterior mitral leaflet and the distance of aortic valve from the membrane, ultimately serve as a predictor of progression.[9] Our patient had isolated lesion and there were no mitral valvular regurgitation present in her.

Turbulent jet produced during left ventricular systole, or cusp tethering to the fibrous membrane or infective endocarditis may result into aortic regurgitation.[10,11] Furthermore, progressive left ventricular hypertrophy may produce dynamic obstruction mimicking hypertrophic cardiomyopathy.[12] Our patient had severe LV hypertrophy and mild aortic regurgitation. Considering all the above facts, an early intervention and precise resection like membranectomy, myectomy or aortic valve repair or replacement has been advocated.[13] Among the operative complications, 42% patients were found

having conduction abnormalities, while some of them requiring permanent pacemaker implantation.[14,15] Histological examination showed degree of disarray and vascular changes in one sixth and one fifth patients respectively.[16] Some study showed that, the occurrences of discrete membrane in the adults were characterized by growth retardation with occasional recurrences.

Minimally invasive surgery (Right anterior thoracotomy) showing promising result to approach the aorta for resection of SAM, but in our patient, the aortic annulus was only 14-15 mm, so we avoided minimal invasive approach and rather sticked with median sternotomy.[17] One year after surgery at follow-up patient is having excellent exercise capacity and very minimum gradient across the aortic valve.

Limitation

This is a case report only, a larger sample multicenter study would have brought a more representative result and would have given the readers a better understanding of the outcome.

Conclusion

Sub-aortic membrane should be considered as a cause of aortic stenosis in adults, as discrete sub-aortic membrane can occur as a complication after correction of congenital cardiac diseases. Although, SAM is a rare disease, proper examination and novel imaging techniques can successfully detect this condition. This day's surgical correction can be accomplished with minimum complication and very low mortality. As a result, early surgery should be considered with appropriate planning for best result.

Recommendation

These patients usually require a varied management guideline. So, we recommend early diagnosis and prompt corrective surgery for best result for the patients.

Acknowledgements

The authors would like to acknowledge all OT, ICU, and hospital staffs involved in the surgery for their relentless effort during surgery, data collection.

Contribution of authors

DIRC-Conception, citing references & final approval.

SDG-Drafting, citing references & final approval.

MNM-Drafting, citing references & final approval.

NP-Design, drafting, citing references & final approval.

APS-Design, drafting, citing references & final approval.

KSR-Drafting, design, drafting, citing references & final approval.

MMR-Design, drafting, citing references & final approval.

FA-Conception, critical revision & final approval.

Disclosure

All the authors declared no competing interests.

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