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Submitral aneurysm in children: A rare entity with varied presentation!



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ABSTRACT

We present echocardiographic images in two children with a diagnosis of submitral aneurysm. Both had absolutely different presentations. The diagnosis was established on echocardiography and no advanced imaging techniques were used.

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1. Clinical summary

1.1. Case 1

An 8-year-old male child presented to our institution with a history of progressive worsening of dyspnea on exertion. Clinical evaluation revealed severe mitral regurgitation with congestive heart failure. Chest radiogram showed cardiomegaly with left atrial enlargement. On transthoracic echocardiography, the patient was diagnosed to have a large submitral aneurysm arising from postero-lateral aspect of left ventricle adjacent to posterior mitral leaflet (PML) (Fig. 1). Color Doppler revealed to and fro flow into aneurysm and severe mitral regurgitation due to impaired coaptation

and avulsed PML (Fig. 1). The patient was planned for surgical correction but the child succumbed due to ventricular tachycardia.

1.2. Case 2

A three year six months old female child was admitted to our institution with short history of fever, breathlessness, and features of heart failure. Her chest radiogram showed cardiomegaly with cardio-thoracic ratio of 0.7. Transthoracic echocardiogram revealed a massive Pericardial effusion with cardiac tamponade. On further evaluation, we noticed a large submitral aneurysm arising from posterior wall of LV immediately below PML (Fig. 2). On color Doppler evaluation, she was found to have a moderate mitral regurgitation (Fig. 3).

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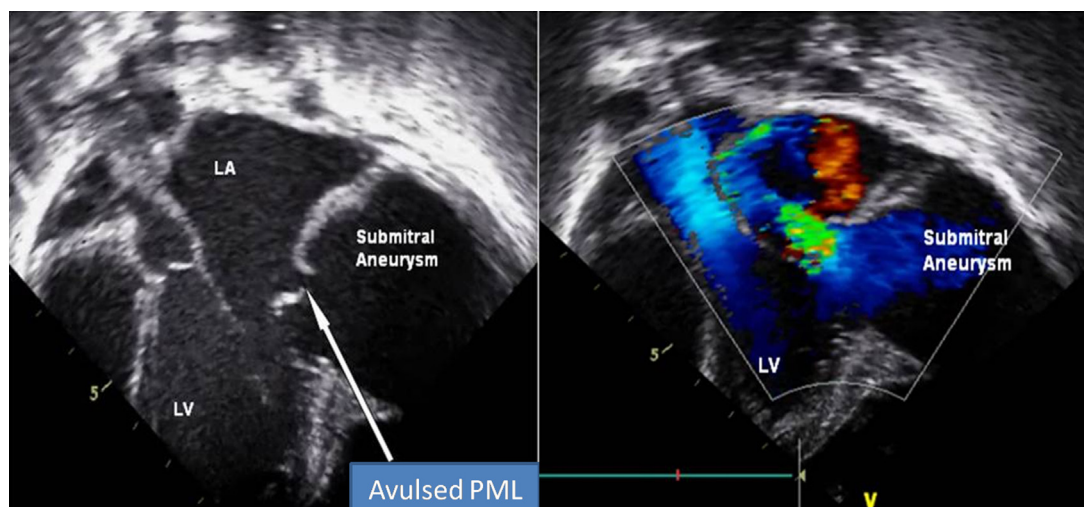


Fig. 1 – 2D echocardiography and color Doppler images in apical views showing a large submitral aneurysm adjacent to posterior mitral leaflet with severe MR.

Her blood reports showed raised erythrocyte sedimentation rate and C-reactive protein. However, procalcitonin was negative and cultures were sterile. Emergency pericardiocentesis was done and the patient was stabilized. Pericardial fluid was straw color, with high protein content (5.4 g%)

without any inflammatory cells. Zeihl nelson stain and gram stain were negative. Adenosine deaminase and lactate dehydrogenase levels were also normal. She was having a nonreactive Mantoux test. Her repeat echocardiogram showed submitral aneurysm without any pericardial effusion. She was discharged in stable hemodynamic condition. However, after two months, she was readmitted with similar complaints with persistently positive inflammatory markers. A pericardial window was created this time and she was stabilized. We are speculating inflammatory process like Takayasu Arteritis as an etiological basis for submitral aneurysm in this case. She was referred to a Pediatric rheumatologist for further evaluation and assessment. However, she was lost to follow-up.

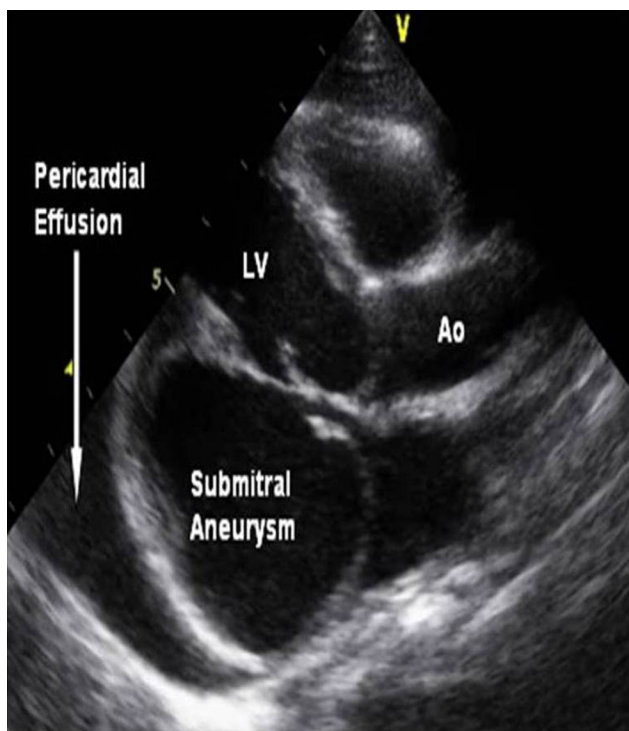


Fig. 2 – 2D echocardiography image in parasternal long axis views showing a thin-walled submitral aneurysm along with pericardial effusion.

2. Discussion

Submitral aneurysm though a known condition, is uncommon in children. After its first description in Africans,¹ cases have been reported from different races all over the world. Its etiology is not known exactly, however it is considered to be due to congenital weakness in posterior portion of mitral fibrous annulus and presence of submitral membranous curtain.² Rare case reports have shown its association with tuberculous pericarditis³ and Takayasu Arteritis.⁴ A case similar to our second case wherein submitral aneurysm was associated with pericardial effusion and positive inflammatory markers was reported by Baruah et al.⁵

Patients with submitral aneurysm can remain asymptomatic or may have symptoms due to significant Mitral regurgitation, thromboembolism, arrhythmia, or ischemic manifestation (secondary to compression of left circumflex artery). Echocardiography is the primary modality of diagnosis of this condition in children. Echocardiographic image of our two patients shows its varied presentation and

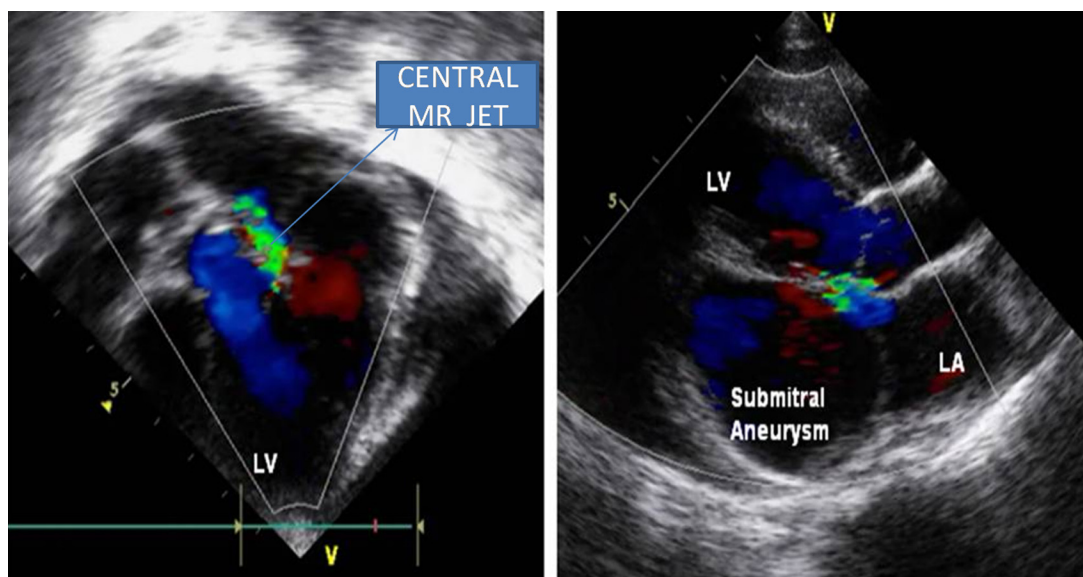


Fig. 3 – Color Doppler image in apical and parasternal long axis views showing submitral aneurysm and a central moderate mitral regurgitation.

possibility of different etiological basis of submitral aneurysm. Du Toit et al.⁶ had classified submitral aneurysm into: type I (single localized neck), type II, multiple necks (separate distinct openings), and type III (involvement of the entire mitral annulus). Both of our cases represented type III aneurysm.

Conflicts of interest

The authors have none to declare.

Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at [doi:10.1016/j.ihj.2016.02.010](https://doi.org/10.1016/j.ihj.2016.02.010).

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