Submitral Aneurysm: Exploring a Rare Pathology

Kellen Round[1], Jake Rosen[1], Colin Yost[1], and T. Sloane Guy[1]

[1]Thomas Jefferson University Sidney Kimmel Medical College

May 27, 2022

Title : Submitral Aneurysm: Exploring a Rare Pathology

Authors : Kellen Round BS[1], Jake L. Rosen BA[1], Colin C. Yost BA[1], T. Sloane Guy MD, MBA[2]

[1]Sidney Kimmel Medical College, Thomas Jefferson University, 1025 Walnut St #100, Philadelphia, PA 19107

[2]Division of Cardiac Surgery, Department of Surgery, Thomas Jefferson University, Curtis Bldg, Ste 620, 1015 Walnut St Philadelphia, PA 19107

Running Title: Submitral Aneurysm Commentary

Corresponding Author:

T. Sloane Guy, M.D., MBA

Professor of Surgery

Director, Minimally Invasive & Robotic Cardiac Surgery

1025 Walnut Street, Suite 607

Philadelphia, PA, 19107

215-955-6996

sloane.guy@jefferson.edu

Word Count: 1032

Funding: This project did not require any source of financial support.

Commentary :

In their case series on outcomes of submitral aneurysm (SMA), Aggarwal et al. provide one of the largest known characterizations and follow-up studies of this infrequently reported pathology[1]. We commend the authors for offering this unique case series and believe it will generate further dialogue on the topic. The series details the presentation, etiologies, and outcomes for ten patients presenting with SMA at a single center. Chiefly, this series demonstrates the need for urgent surgical repair to improve mitral regurgitation and overall prognosis.

Submitral aneurysms are an extremely rare pathology typically found in young patients[1-3]. While they were originally described in Western and Southern African populations, SMAs have been identified in patients across a variety of races and ethnicities[4]. In contrast to posterior left ventricular aneurysms, which often occur due to ischemic etiology, submitral aneurysms are believed to be largely congenital in nature[1,4–7]. The pathology likely stems from a weakening of the fibrous tissue of the posterior mitral annulus, which creates an outpouching between the annulus and the left ventricular endocardium that restricts the posterior

mitral leaflet[6]. Nayak et al. suggest the presence of a submitral curtain, a fibrous separation between the fibrous annulus and the left ventricular musculature, further predisposes patients to developing SMA[8]. As a consequence of dysfunctional posterior leaflet motion and coaptation, patients with SMA often present with severe mitral regurgitation. Additionally, patients with aneurysms that compress the left circumflex artery may present with symptoms resulting from coronary compression[9].

Acquired submitral aneurysms have also been described; prior case reports have outlined SMAs secondary to Takayasu's arteritis, rheumatic heart disease, and cardiac ischemia[6,10,11]. While transmural infarction typically causes apical aneurysm, ischemia-induced SMAs have been documented in prior literature and in three patients in the authors' series[1,7]. Infectious agents such as human immunodeficiency virus (HIV) and tuberculosis have also been proposed as etiologies of SMA, as inflammation is suspected to contribute to the pathogenesis and may exacerbate the congenital pathology of SMA. However, it is still uncertain whether these infectious etiologies are causative or a coincidental reflection of the diseases endemic to the demographic in which SMA is typically found[4,12].

SMA are classified based on the extent of posterior annular involvement: Type I SMA have a single localized neck, Type II SMA have multiple necks, and Type III SMA involve the entirety of the posterior fibrous annulus[4]. Clinically, the aneurysms are often asymptomatic and incidentally discovered on echocardiography[5]. However, symptomatic patients often present with severe mitral regurgitation and require surgery. Moreover, coronary compression and risk of aneurysmal rupture prompt surgical management in many cases[9,13]. This stands in clear contrast to the management of classical ventricular aneurysm, for which surgery is not immediately indicated[14]. As evidenced by previous cases, SMA can rapidly progress to cardiogenic shock without swift intervention[6]. Furthermore, failure to identify multiple aneurysmal necks during surgery has been suggested as an impediment to surgical intervention[4]. The importance of surgical intervention was emphasized in this series presented by Aggarwal et al., as two of the three patients who refused surgery died within three months[1].

The classical approach to SMA repair is extensively detailed in the literature dating back to 1963 by Drs. Schrire and Barnard[3]. Typically performed via median sternotomy, SMA repair also has been reported through left-sided thoracotomy[15]. During the operation, consideration must be given to the location of the left circumflex artery, as aneurysmal location may overlap. Coronary angiography can be applied to visualize both the location of the left circumflex and the direction of the aneurysmal growth[15].

Two techniques have largely been described as the means to repairing an SMA: the intracardiac and extracardiac approaches. The intracardiac approach described by Dr. Antunes includes left atriotomy anterior to the pulmonary veins and posterior to the vena cavae, similar to the approach of a classical open mitral valve repair[3]. In the series reported by Dr. Antunes, the posterior leaflet was retracted and its free edge was everted in order to reveal submitral dilatation. Subsequent incision 20-25 mm from the free edge of the posterior leaflet was then performed to enable closure from inside the aneurysm. Traction sutures can be placed at the aneurysmal neck to close the dilatation and reattach the posterior leaflet when necessary. Repair can also be facilitated through the use of a Dacron patch and excess tissue excision[16]. The extracardiac approach is performed by opening the epicardial side of the aneurysmal wall and exposing the aneurysm anterior to the atrio-ventricular groove by retracting and twisting the heart counterclockwise[4]. However, difficulties in exposing the mitral annulus have been noted with this approach.

With the poor prognosis demonstrated by this series (40% mortality at one-year follow-up), astute attention to management is imperative. While the study's sample is not of large power, it appears essential to address surgical repair in patients with SMA; surgical intervention in this series demonstrated 100% survival one year post-diagnosis. Moreover, the three patients who received mitral valve replacement experienced sustained improvement of mitral regurgitation and were doing well in one-year follow-up. As such, this data suggests a pattern of improvement following surgical intervention and reinforces a poor prognosis for SMA when surgery is refused. However, the lack of long-term follow-up data on complications and outcomes for patients with SMA exposes an existing gap in the literature.

While this series is one of the largest of its kind for follow-up explicitly involving SMA, we urge more in-depth investigation of the risk factors, comorbidities, and outcomes of SMA. Additionally, the approach to management and long-term follow-up of SMA is underreported, and the field would benefit from further investigation in this realm. Creative approaches to gain insight into anatomic variability have been reported, including the use of 3D printing to inform aneurysmal closure[17]. We anticipate less invasive approaches to SMA repair have been attempted, including the use of robotics, however no such characterizations exist in the literature. As technology progresses and allows for stronger visualization with less invasion, we expect to see an increase in the use of minimally invasive techniques to provide superior results and less associated postoperative adverse effects. With more extensive investigation into the long-term outcomes of various approaches to management of SMA, we believe the prognosis of this rare presentation can be improved.

Abbreviations : SMA: submitral aneurysm; HIV: human immunodeficiency virus

References

1. Aggarwal P, Shrivastava A, Kuber D, Nath RK, Sinha SK, Kumar T, Raj A, Pandit B. Outcome of Sub-mitral aneurysm. Journal of Cardiac Surgery. 2022.

2. Abrahams DG, Barton CJ, Cockshott WP, Edington GM, Weaver EJ. Annular subvalvular left ventricular aneurysms. Q J Med. 1962;31:345-360.

3. de Antunes M. Submitral left ventricular aneurysms. J Thorac Cardiovasc Surg. 1987; 94:241-5.

4. Du Toit H. Left ventricular sub-valvar mitral aneurysms. Interact Cardiovasc Thorac Surg. 2003;2(4):547-551. doi:10.1016/S1569-9293(03)00141-5

5. Corrado G. Submitral congenital left ventricular aneurysm: you recognize only what you know. Journal of Cardiovascular Echography. 2017;27(1):29. doi:10.4103/2211-4122.199057

6. Barnawi A, Al Otaibi A, Soliman A, Al Rifae M, Al Baradie A, Ghazal S. A Case of Submitral Aneurysm Presenting with Severe Mitral Regurgitation and Shock in an African Male Patient with Rheumatic Heart Disease. CASE (Phila). 2021;5(4):224-226. doi:10.1016/j.case.2021.04.001

7. Prasad K, Gupta H, Sihag BK, et al. Submitral aneurysm of varied aetiologies: a case series. Eur Heart J Case Rep. 2021;5(2):ytab066. doi:10.1093/ehjcr/ytab066

8. Nayak VM, Victor S. Sub-mitral membranous curtain: A potential anatomical basis for congenital Sub-mitral aneurysms. Indian J Thorac Cardiovasc Surg. 2006;22(4):205-211. doi:10.1007/s12055-006-0003-4

9. Thangasami S, Sahoo SS, Chandrasekaran A, Raval P, Shaniswara P. Large sub-mitral aneurysm compressing the left circumflex coronary artery presenting with atypical chest pain - Rare presentation. J Cardiol Cases. 2020;21(5):193-196. doi:10.1016/j.jccase.2020.02.008

10. Rose AG, Folb J, Sinclair-Smith CC, Schneider JW. Idiopathic annular submitral aneurysm associated with Takayasu's aortitis. A report of two cases. Arch Pathol Lab Med. 1995;119(9):831-835.

11. Baruteau A-E, Martins RP, Boulmier D, et al. Acquired left ventricular submitral aneurysms in the course of Takayasu arteritis in a child. Congenit Heart Dis. 2012;7(1):76-79. doi:10.1111/j.1747-0803.2011.00537.x

12. Chesler E, Joffe N, Schamroth L, Meyers A. Annular subvalvular left ventricular aneurysms in the south african bantu. Circulation. 1965;32:43-51. doi:10.1161/01.cir.32.1.43

13. Raghuram P, Sivakumar K, Sheriff EA. Acquired submitral aneurysms may be associated with mitral paravalvular leaks. Asian Cardiovasc Thorac Ann. 2021;29(8):822-825. doi:10.1177/02184923211006310

14. Ruzza A, Czer LSC, Arabia F, et al. Left ventricular reconstruction for postinfarction left ventricular aneurysm: review of surgical techniques. Tex Heart Inst J. 2017;44(5):326-335. doi:10.14503/THIJ-16-6068

15. Purushotham S, Manohar SRK, Sivasubramaniam S, Neelakandhan KS. Submitral left ventricular aneurysm: the location of the circumflex coronary artery in relation to the aneurysm influences the surgical approach and outcome. J Thorac Cardiovasc Surg. 2005;129(5):1175-1177. doi:10.1016/j.jtcvs.2004.08.044

16. Baruah DK, Kumar PVN, Reddy GSP, Babu VR. Submitral aneurysm of the left ventricle. Indian Heart J. 2012;64(1):77-79. doi:10.1016/S0019-4832(12)60015-7

17. Shetty I, Lachma RN, Manohar P, Rao PSM. 3D printing guided closure of submitral aneurysm-an interesting case. Indian J Thorac Cardiovasc Surg. 2020;36(5):506-508. doi:10.1007/s12055-020-00973-6