

Journal of Cardiovascular Disease Research

An Official Publication of SciBiolMed.Org (A publishing division of Phcog.Net)

"Our mission, Your Outstanding Research Work"

Volume 7, Issue 2, Apr-Jun, 2016

About Journal

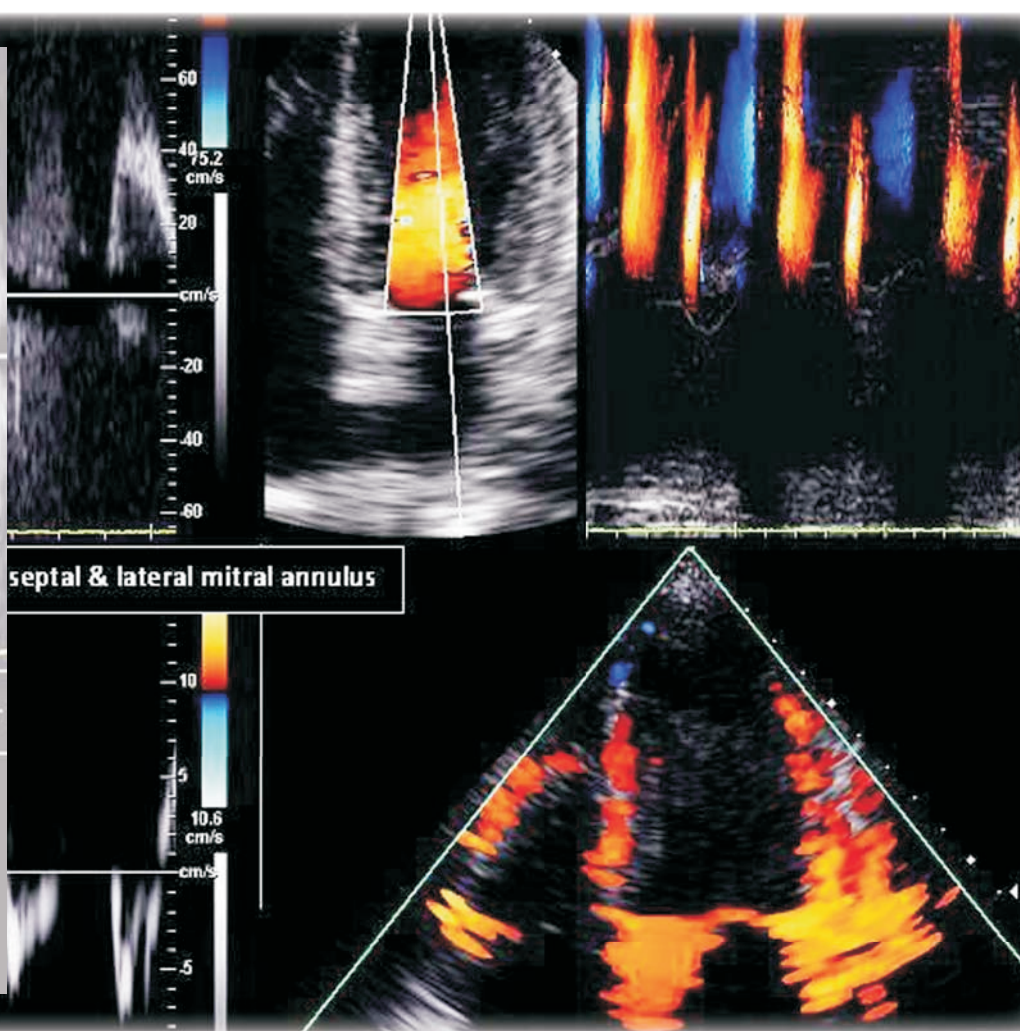
Journal of Cardiovascular Disease Research (J Cardiovasc. Dis. Res.)

[www.jcdronline.org]

[ISSN: Print -0975-3583, Online - 0976-2833]

An official publication of SciBiolMed.Org (www.scibiolmed.org);

it is a double-blind peer-reviewed, open access international circulating professional journal led by a group of research scientists, vascular disease experts and cardiologists coming from North America, Asia and Europe etc.



Indexed and Abstracted in : The journal is indexed with Caspur, Chemical Abstracts, CNKI (China National Knowledge Infrastructure), DOAJ, EBSCO Publishing's Electronic Databases, Expanded Academic ASAP, Genamics JournalSeek, Google Scholar, Health & Wellness Research Center, Health Reference Center Academic, Hinari, Index Copernicus, MANTIS, OpenJGate, PrimoCentral, ProQuest, Scimago Journal Ranking, SCOLOAR, SCOPUS, SIIC databases, Summon by Serial Solutions and Ulrich's International Periodical Directory



JCDR - Providing Cutting Edge Research information on Cardiovascular Diseases

Senior International Editor: Dr. Dayi Hu
Editor : Dr. Peng Zhou

Manuscript

Large unruptured Pseudoaneurysm of Mitral-Aortic Intervalvular Fibrosa-Managed Conservatively for Three Years: Case Report and Review of Literature

Jahangir Rashid Beig¹, Shazia Bashir², Hilal A. Rather¹, Mohammad Iqbal Dar¹

¹Department of Cardiology, SKIMS, Soura, Srinagar, Jammu and Kashmir, INDIA. Pin- 190020

²Department of Radiology, SKIMS, Soura, Srinagar, Jammu and Kashmir, INDIA. Pin-190020

ABSTRACT

Here in, we describe a rare case of an eleven years old boy who was diagnosed with infective endocarditis of a congenitally bicuspid aortic valve with severe aortic stenosis, severe aortic regurgitation and complicated by cerebral embolism. He was managed with intravenous antibiotics for 6 weeks. A pre-discharge transthoracic echocardiogram revealed a large, unruptured pseudoaneurysm of the mitral-aortic intervalvular fibrosa that was confirmed by transesophageal echocardiography and cardiac MRI. Patient's parents initially refused surgery and he was managed conservatively for 3 years. Serial echocardiograms performed during this period demonstrated progressive dilatation of the left ventricle and aorta, with a small increase in the size of pseudoaneurysm. Patient was subsequently referred for surgery (Aortic valve replacement with reconstruction of mitral-aortic intervalvular fibrosa). In conclusion, our case demonstrates that close clinical follow up and periodic imaging may be a reasonable alternative to early surgery in managing patients with uncomplicated pseudoaneurysm of mitral-aortic intervalvular fibrosa.

Key words: Pseudoaneurysm, Mitral-aortic intervalvular fibrosa, Bicuspid-aortic valve, Infective endocarditis.

Correspondence:

Dr. Jahangir Rashid Beig (MD,DM),

Senior Resident, Dept. of Cardiology,
SKIMS, Soura, Srinagar,
Jammu & Kashmir, INDIA. PIN: 190020

Phone: +91-0194-2401765

Mobile: +919622671622

Submission Date: 16-03-2016; **Review completed:** 29-04-2016;

Accepted Date: 08-05-2016.

Email: jahangir3582@gmail.com

DOI : 10.5530/jcdr.2016.2.10

INTRODUCTION

Pseudoaneurysm of mitral-aortic intervalvular fibrosa (P-MAIVF) is a rare but potentially lethal complication of aortic valve endocarditis, especially with prosthetic aortic valves.¹ Early surgery is generally recommended in these patients because of risk of possible fatal complications.^{2,3} There are very few cases reported in literature wherein patients with P-MAIVF have been managed conservatively for years after initial diagnosis.^{4,5} We report a case of large P-MAIVF in a patient who was treated for infective endocarditis of a native aortic valve, and was managed conservatively for 3 years with an uncomplicated course, and a small increase in the size of the pseudoaneurysm.

CASE REPORT

An eleven years old boy presented to us with a history of high grade fever for 28 days and sudden onset weakness of left side of the body for 4 days. He had received intravenous ceftriaxone for 1 week prior to admission from a private practitioner outside the hospital. On general examination he was conscious and oriented to time, place and person. His temperature was 104°F, pulse rate was 120 beats per minute, blood pressure was 110/60 mm Hg and respiratory rate was 18 per minute. He had pallor and grade 2 clubbing. Cardiovascular examination revealed a carotid thrill, hyperdynamic precordium, Grade IV ejection systolic murmur and an early diastolic murmur at the aortic area. There were no signs of heart failure. Neurological examination showed left sided hemiparesis with grade 2/5 power in both left upper and lower limbs, brisk deep tendon reflexes on left side, an extensor left plantar response and left upper motor neuron type facial palsy. Chest and abdominal examination was unremarkable. Haemogram revealed normocytic normochromic anemia with hemoglobin - 9.5 gm/dl, total leucocyte count - 13,500/ ml, differential counts - 75% neutrophils, 20% lymphocytes and 8% monocytes,

and platelet counts - 2,54,000/ml. ESR was 45 mm/hour. Blood urea, serum creatinine, liver function tests, arterial blood gases, serum electrolytes, urine examination, chest X-ray and USG abdomen were normal. Blood culture was sterile thrice, as was urine culture. ECG showed sinus tachycardia. CT head was suggestive of an ischemic infarct in the right MCA territory which was confirmed on MRI brain. A transthoracic echocardiogram (TTE) was performed and revealed a bicuspid aortic valve with severe aortic stenosis (mean gradient- 45 mmHg, aortic valve area- 0.6 cm²/m²), moderate aortic regurgitation and vegetations attached to the ventricular surface of non-coronary cusp. Patient was managed with intravenous ceftriaxone and vancomycin for 6 weeks and showed signs of clinical recovery with mild residual hemiparesis. A pre-discharge TTE was performed that revealed a pulsatile outpouching arising from the left ventricular outflow tract, just beneath the aortic annulus, in the region of mitral-aortic intervalvular fibrosa (Figure 1). Its diastolic dimensions were 1.8 × 2.4 cm. Color flow Doppler examination demonstrated that the lesion was freely communicating with the left ventricular outflow tract (LVOT) and showed typical systolic expansion and diastolic collapse (Video 1). Patient was further investigated with transesophageal echocardiography (TEE), and cardiac MRI which confirmed the diagnosis of bicuspid aortic valve disease with a large (diastolic dimensions-1.8 × 2.5 × 2.9 cm) P-MAIVF related anteriorly to the aortic root and posteriorly to left atrium (Figure 2). Ascending aorta was dilated and measured 3.6 × 3.5 cm. There were no signs of rupture of the pseudoaneurysm into left atrium, ascending aorta or pericardial cavity. The case was discussed in a forum consisting of senior cardiologists and cardiac surgeons and it was decided that the patient should undergo an early surgical treatment. However patient's parents refused surgery and he was managed conservatively for a period of 3 years with

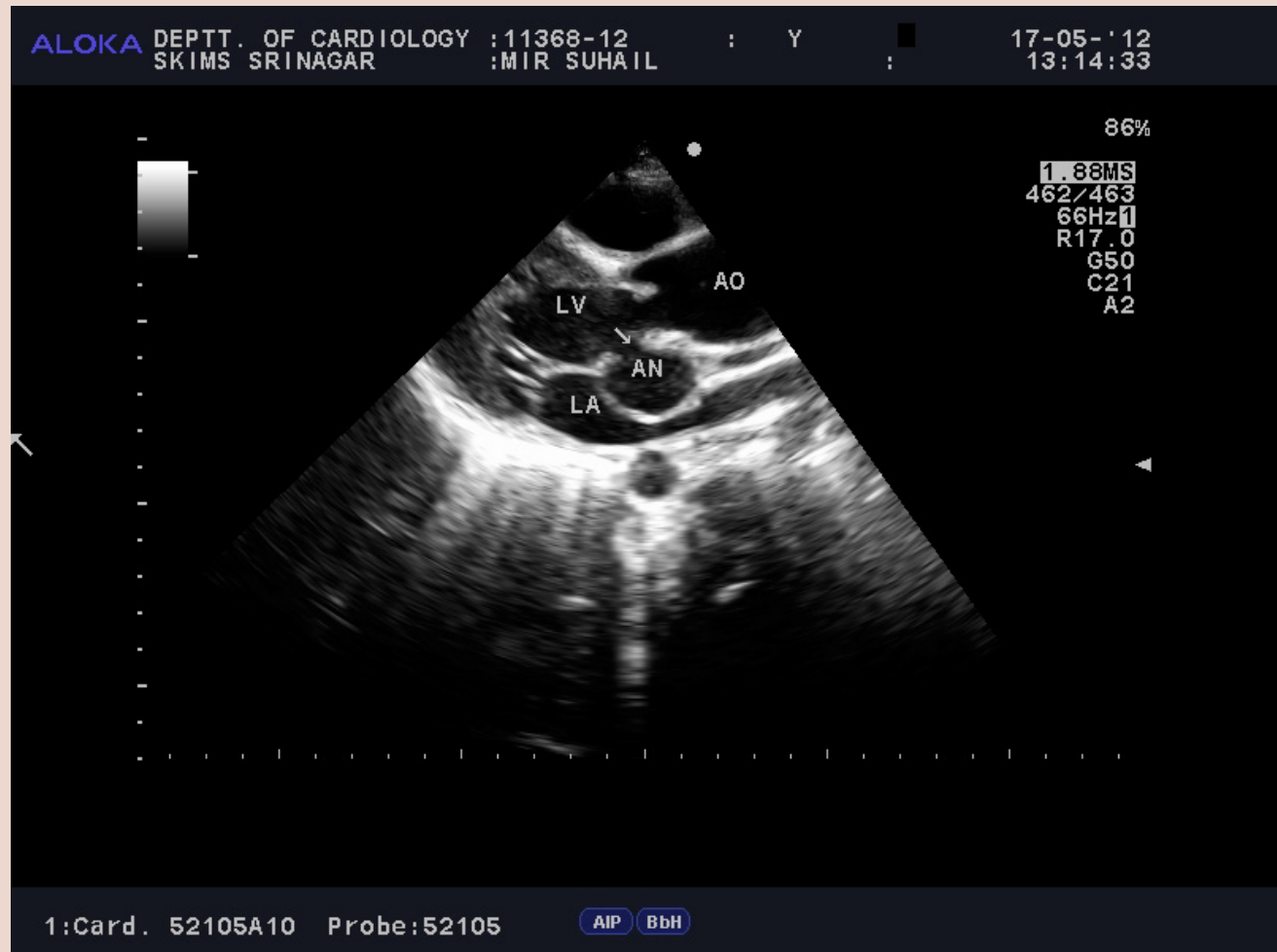


Figure 1: Transthoracic echocardiogram (parasternal short axis view) demonstrating pseudoaneurysm of mitral-aortic intervalvular fibrosa (AN) posterior to left ventricular (LV) outflow tract.

periodic echocardiographic assessment of aortic valve, left ventricular size and functions, pseudoaneurysm size and development of any complications. The need of surgical treatment was also reemphasized to the patient's parents at each visit. At the end of 3 years patient had minimal cardiac symptoms, aortic regurgitation had become severe, left ventricular dimensions had progressively increased and pseudoaneurysm had mildly but definitely increased in size (diastolic dimensions- 2.0×2.7 cm on TTE). Ascending aortic size had also increased (3.9×3.8 cm). This time, patient's parents accepted surgery and he was referred for aortic valve replacement and reconstruction of MAIVF.

DISCUSSION

Mitral-aortic intervalvular fibrosa (MAIVF) is a thin, translucent membranous structure joining the left half of noncoronary cusp and adjacent third of left coronary cusp of the aortic valve and the anterior mitral leaflet.⁶ Its anterior edge is contiguous with the left fibrous trigone and posterior edge is contiguous with the right fibrous trigone. It is bound posterolaterally by the left atrium, superiorly by the pericardium and inferiorly by the left ventricular outflow tract.⁷ It is of great clinical and surgical importance as it is responsible for maintaining the structural and

functional integrity of both mitral and aortic valves.⁶ Pseudoaneurysm of MAIVF (P-MAIVF) most commonly occurs as a complication of aortic valve endocarditis, particularly with prosthetic aortic valves.¹ It can also be associated with chest trauma, cardiac surgery, tuberculosis and Takayasu arteritis.⁸⁻¹¹ Some cases are believed to be of congenital origin.¹² Being a fragile and sparsely vascularized structure, it offers little resistance to infection and is thus prone to develop abscess or pseudoaneurysm.¹³ Clinically, patients with P-MAIVF may be asymptomatic, present with features of infective endocarditis or complications. Complications of P-MAIVF include rupture into left atrium leading to supra-annular mitral regurgitation and consequent acute heart failure, rupture into pericardial cavity causing cardiac tamponade, systolic compression of left coronary artery causing angina, rupture towards chest wall producing fistula, stroke due to thromboembolism and sudden death.¹³⁻¹⁷ Echocardiography is the primary imaging modality for the diagnosis of P-MAIVF. Transesophageal echocardiography (TEE) is superior to transthoracic echocardiography (TTE) for diagnosing P-MAIVF. Afridi *I et al* reported that diagnostic sensitivity of TTE to detect P-MAIVF is 43% which may increase to 90% when TEE is utilized.² On echocardiography, P-MAIVF appears as a pulsatile outpouching located between the aortic root and the anterior mitral leaflet.² Its most characteristic echocardiographic fea-

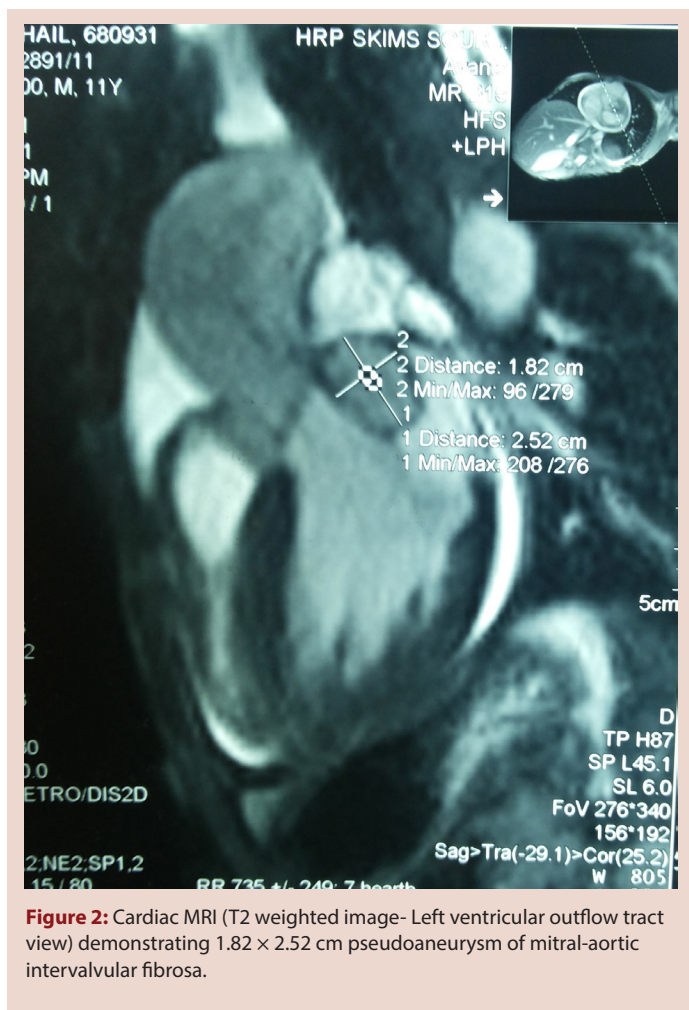
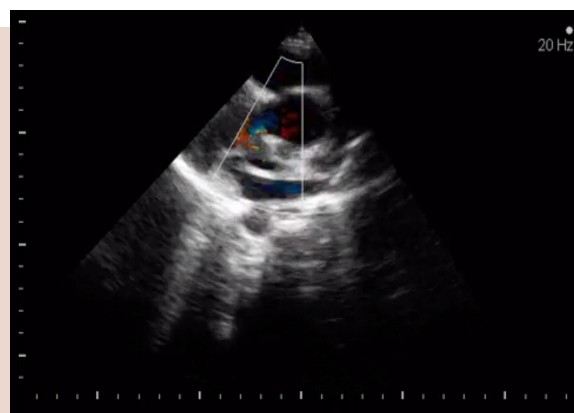


Figure 2: Cardiac MRI (T2 weighted image- Left ventricular outflow tract view) demonstrating 1.82 x 2.52 cm pseudoaneurysm of mitral-aortic intervalvular fibrosa.



Video 1: Color flow Doppler examination demonstrating free communication between the pseudoaneurysm and left ventricular outflow tract.

ture is marked pulsatility with systolic expansion and diastolic collapse which indicates its direct communication with the left ventricular cavity. Color flow Doppler demonstrates free flow of blood from left ventricular cavity into P-MAIVF and also detects rupture of P-MAIVF into the left atrium or aorta.² Differential diagnosis of P-MAIVF includes aortic ring abscess, aorto-left ventricular tunnel and aneurysm of sinus of Valsalva. It may be particularly difficult to differentiate small P-MAIVF from an aortic ring abscess on echocardiography. In general ring abscesses are small, nonpulsatile and show either no flow or continuous systolic and diastolic flow.² Real time 3D echocardiography has also been used for providing useful anatomical insight regarding location of P-MAIVF and its relationship with the adjacent structures.¹⁸ Recently, ECG gated multidetector CT and cardiac MRI have been utilized for detailed 3D anatomical reconstruction and dynamic evaluation of P-MAIVF with high spatial and temporal resolution.¹⁹ Natural history of P-MAIVF is unclear. Surgery is generally recommended, even in asymptomatic patients, because of risk of possible fatal complications.² Surgical treatment includes resection of P-MAIVF and reconstruction of MAIVF with or without aortic valve, mitral valve or homograft aortic root replacement.¹⁶ Coronary artery bypass grafting may be necessary when there is coronary involvement.¹³ Surgery of P-MAIVF is technically demanding and associated with high risk of mortality and morbidity. De Oliveira *et al* reported an operative mortality of 10%, late mortality of 24% and 10 year survival of 50% in 76 patients undergoing reconstruction of MAIVF with aortic and mitral valve replacement.²⁰ Additionally, successful percutaneous device closure of P-MAIVF has also been reported.⁷ Till date, there

are very few cases reported in literature wherein patients with P-MAIVF have been managed conservatively for years after initial diagnosis.^{4,5} In most of these cases, the pseudoaneurysms were small and not associated with any complications. In our case, the P-MAIVF was large, although unruptured and not causing any significant compression of surrounding vital structures. It had become only slightly larger after three years and was not causing any symptoms, although due to severe aortic regurgitation the left ventricle and aorta had progressively dilated. This increase in size could be attributed to the persistent blood flow in and out of the pseudoaneurysm over a period of time. Patient was ultimately referred for surgery (Aortic valve replacement and reconstruction of MAIVF). In summary, our case underscores the role of routine pre-discharge echocardiography in detecting the sub-valvular complications of aortic valve endocarditis, and also demonstrates that close clinical follow up and periodic imaging may be a reasonable alternative to early surgery in managing patients with uncomplicated P-MAIVF. Further research is needed to establish whether this nonsurgical strategy should be recommended for the management of such patients.

CONCLUSION

Pseudoaneurysm of mitral-aortic intervalvular fibrosa is a rare but dreaded complication of aortic valve endocarditis. Although early surgery is the treatment of choice, expectant management with close clinical follow up and serial imaging may be a reasonable alternative strategy of managing patients with uncomplicated P-MAIVF. Further studies are needed to establish whether this approach should be recommended for the management of such patients.

ACKNOWLEDGEMENT

We are sincerely thankful to Dr. Nisar A. Trambo (Prof & Head, Dept. of Cardiology, SKIMS, Srinagar) for his guidance in preparing this manuscript.

CONFLICT OF INTEREST

The authors declare that they have no conflict of interest.

SOURCE OF SUPPORT

Nil.

ABBREVIATIONS USED

CT: Computerized tomography; **ECG:** Electrocardiogram; **ESR:** Erythrocyte sedimentation rate; **MCA:** Middle cerebral artery; **MRI:** Magnetic resonance imaging; **USG:** Ultrasonogram; **3D:** 3-dimensional.

REFERENCES

- Yokoyama Y, Tamaki S, Kato N, Yokote J, Mutsuga M. Pseudoaneurysm from the mitral-aortic intervalvular fibrosa following endocarditis. *Jpn J Thorac Cardiovasc Surg.* 2003;51(8):374-7.
- Afridi I, Apostolidou MA, Saad RM, Zoghbi WA. Pseudoaneurysms of the mitral-aortic intervalvular fibrosa: dynamic characterization using transesophageal echocardiographic and Doppler techniques. *J Am Coll Cardiol.* 1995;25(1): 137-45.
- Sudhakar S, Sewani A, Agrawal M, Uretsky B. Pseudoaneurysm of the mitral-aortic intervalvular fibrosa (MAIVF): a comprehensive review. *J Am Soc Echocardiogr.* 2010;23(10):1009-18.
- Meyerowitz CB, Jacobs LE, Kotler MN, Ioli AW, Wertheimer JH. Four-year follow-up of a pseudoaneurysm of the mitral aortic fibrosa. *Am Heart J.* 1991; 122(2):589-92.
- Gin A, Hong H, Rosenblatt A, Black M, Ristow B, Popper R. Pseudoaneurysms of the mitral-aortic intervalvular fibrosa: Survival without reoperation. *Am Heart J.* 2011;161(1):130-e1.
- Cavalcanti JS, De Melo NC, de Vasconcelos RS. Morphological study of the human mitral-aortic intervalvular fibrosa. *Braz J Morphol Sci.* 2005;22:37-40.
- Jiménez Valero S, García E, González Pinto A, Delcán JL. Percutaneous closure of pseudoaneurysm of the mitral-aortic intervalvular fibrosa. *Rev Esp Cardiol.* 2005; 58(12):1473-5.
- Mathews RV, French WJ, Criley JM. Chest trauma and subvalvular left ventricular aneurysm. *Chest.* 1989;95(2):474-5.
- Yeo TC, Malouf JF, Oh JK, Seward JB. Clinical profile and outcome in 52 patients with cardiac pseudoaneurysm. *Ann Int Med.* 1998;128(4):299-305.
- Takawira FF, Joshi JA, Du Plessis DJ. Development of a subaortic aneurysm secondary to disseminated tuberculosis in a child. *Ann Thorac Surg.* 2010;90(2): 644-7.
- Tufekcioglu O, Ozlu MF, Cay S, Tuna F, Basar N, Gurel OM, *et al.* Pseudoaneurysm of the mitral-aortic intervalvular fibrosa in a patient with Takayasu's arteritis. *Can J Cardiol.* 2008;24(19):718.
- Chesler E, Joffe N, Schamroth L, Meyers A. Annular subvalvular left ventricular aneurysms in the South African Bantu. *Circulation.* 1965;32(1):43-51.
- Parashara DK, Jacobs LE, Kotler MN, Yazdanfar S, Spielman SR, Janzer SF *et al.* Angina caused by systolic compression of the left coronary artery as a result of pseudoaneurysm of the mitral-aortic intervalvular fibrosa. *Am Heart J.* 1995;129(2):417-21.
- Espinosa-Calani JS, Montijano A, Melero JM, Montiel A. Pseudoaneurysm in the mitral-aortic intervalvular fibrosa. A cause of mitral regurgitation. *Eur J Cardiothorac Surg.* 2000;17(6):757-9.
- Qizilbash AH, Schwartz CJ. False aneurysm of left ventricle due to perforation of mitral-aortic intervalvular fibrosa with rupture and cardiac tamponade. Rare complication of infective endocarditis. *Am J Cardiol.* 1973;32(1):110-3.
- Agirbasli M, Fadel BM. Pseudoaneurysm of the mitral-aortic intervalvular fibrosa: a long-term complication of infective endocarditis. *Echocardiography.* 1999;16(3):253-7.
- Koch R, Kapoor A, Spencer KT. Stroke in patient with an intervalvular fibrosapseudoaneurysm and aortic pseudoaneurysm. *J Am Soc Echocardiogr.* 2003;16(8):894-6.
- Chan KL, Liu X, Asch KJ, Beauchesne LM, Burwash IG. Comparison of real-time 3-dimensional echocardiography with conventional 2-dimensional echocardiography in the assessment of structural heart disease. *J Am Soc Echocardiogr.* 2004;17(9):976-80.
- Ghersin E, Litmanovich D, Agmon Y, Milo S. Pseudoaneurysm of the mitral-aortic intervalvular fibrosa following aortic valve replacement - diagnosis and dynamic evaluation with multidetector CT and transesophageal echocardiography. *Interact Cardiovasc Thorac Surg.* 2005;4(6):502-4.
- De Oliveira NC, David TE, Armstrong S, *et al.* Aortic and mitral valve replacement with reconstruction of the intervalvular fibrous body: an analysis of clinical outcomes. *J Thorac Cardiovasc Surg.* 2005;129(2):286-90.

Cite this article : Beig JR, Bashir S, Rather HA, Dar MI. Large unruptured Pseudoaneurysm of Mitral-Aortic Intervalvular Fibrosa-Managed Conservatively for Three Years: Case Report and Review of Literature. *J Cardiovasc Disease Res.* 2016;7(2):100-3.