



Severe mitral annular calcification in rheumatic heart disease: A rare presentation

Rajesh Vijayvergiya, Kim Vaiphei, Sandeep S Rana

Rajesh Vijayvergiya, Department of Cardiology, Post Graduate Institute of Medical Education and Research, Chandigarh 160 012, India

Kim Vaiphei, Department of Histopathology, Post Graduate Institute of Medical Education and Research, Chandigarh 160 012, India

Sandeep S Rana, Department of Cardio-thoracic Surgery, Post Graduate Institute of Medical Education and Research, Chandigarh 160 012, India

Author contributions: All the authors were actively involved in management of the index case.

Correspondence to: Dr. Rajesh Vijayvergiya, MD, DM, FS-CAI, FISES, Associate Professor, Department of Cardiology, Post Graduate Institute of Medical Education and Research, Chandigarh 160 012, India. rajeshvijay999@hotmail.com

Telephone: +91-172-2756512 Fax: +91-172-2744401

Received: November 22, 2011 Revised: December 10, 2011

Accepted: December 17, 2011

Published online: March 26, 2012

Matej Podbregar, Professor, Center for Intensive Care, University Clinical Centre, 1000 Ljubljana, Slovenia

Vijayvergiya R, Vaiphei K, Rana SS. Severe mitral annular calcification in rheumatic heart disease: A rare presentation. *World J Cardiol* 2012; 4(3): 87-89 Available from: URL: <http://www.wjgnet.com/1949-8462/full/v4/i3/87.htm> DOI: <http://dx.doi.org/10.4330/wjc.v4.i3.87>

INTRODUCTION

Mitral annular calcification (MAC) is a common feature in patients with chronic rheumatic heart disease (RHD), chronic kidney disease, and advanced age^[1]. A severely calcified mitral annulus is frequently seen in patients with chronic kidney disease and degenerative valve disease, but it is rare in RHD. We hereby report a case of severe MAC in a chronic RHD patient and discuss the management issues related to it.

CASE REPORT

A 45-year-old female with chronic RHD who was under medical treatment for 2 years, presented with atrial fibrillation and gross congestive heart failure. Her routine serum biochemistry tests, including urea and creatinine, were normal. Two-dimensional echocardiography showed a thickened, calcified, retracted posterior mitral leaflet with severe mitral regurgitation. The posterior mitral leaflet and adjacent mitral annulus were calcified (Figure 1). She improved with diuretics and other supportive treatment. Fluoroscopy in right anterior oblique 30° revealed a "crown"-like severe calcification of the mitral annulus (Figure 2). Her angiography showed normal epicardial coronaries. A left ventriculogram revealed an ejection fraction of 0.50, and grade III mitral regurgitation. The pulmonary artery systolic pressure was 58 mmHg. During the hospital stay awaiting cardiac surgery,

Abstract

Severe mitral annular calcification (MAC) is frequently seen in patients with advanced age and chronic kidney disease, but it is rare in rheumatic heart disease (RHD). We hereby report a case of 45-year-old female with chronic RHD, who had severe MAC and mitral regurgitation. Fluoroscopy revealed a "crown"-like severe calcification of the mitral annulus. Autopsy of the heart revealed a calcified posterior mitral annulus, fused commissures, and calcified nodules at the atrial aspect of the mitral valve.

© 2012 Baishideng. All rights reserved.

Key words: Mitral annular calcification; Rheumatic heart disease; Mitral regurgitation; Autopsy; Aschoff nodule

Peer reviewers: Stephen Wildhirt, MD, PhD, Associate Clinical Professor of Cardiothoracic Surgery, Department of Cardiothoracic- and Vascular Surgery, Johannes Gutenberg University Mainz, Langenbeckstrasse 1, 55131 Mainz, Germany;

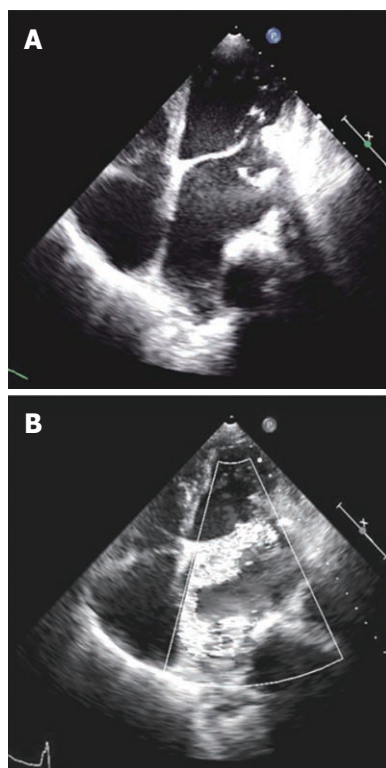


Figure 1 Echocardiography in apical 4 chamber view. A: Calcified, thickened and retracted posterior mitral leaflet with adjacent mitral annular ring calcification; B: Color Doppler showing severe mitral regurgitation.

she had a sudden cardiac arrest and died. The autopsy revealed severe calcification of the posterior mitral valve ring, irregular calcified nodules on the posterior mitral leaflet, and commissural fusion of the posterior leaflet (Figure 3). Histopathology of the left atrial inner wall revealed Aschoff Nodule with fibrosis suggesting a rheumatic etiology^[2].

DISCUSSION

MAC in patients with RHD usually involves commissures and leaflet tissue, with only late extension to the annulus. Severe MAC, as present in the index case, is rare in RHD, though it is often reported in patients with degenerative valve disease^[3] and chronic kidney disease^[4]. There is a risk of systemic non-thrombotic embolism of calcified material in such cases during the natural course of the disease and also at the time of percutaneous or surgical intervention^[5]. MAC is usually associated with mitral stenosis because of restricted posterior mitral leaflet and annulus movement^[6]. However, the index case had a thickened, retracted, poorly aligned posterior leaflet resulting into severe mitral regurgitation. Sudden cardiac death in the index case can be explained by a low ejection fraction of 0.50, left ventricular hypertrophy secondary to mitral regurgitation, and diuretics induced electrolyte imbalance^[7]. Surgical treatment in such a case is technically difficult. There is need for adequate debridement and annular reconstruction prior to mitral

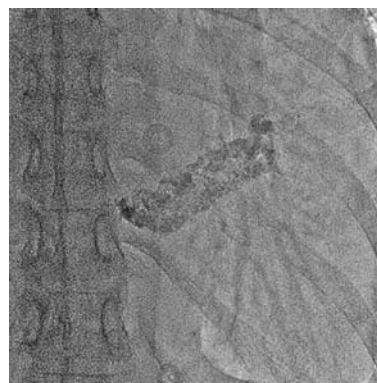


Figure 2 Fluoroscopy image in right anterior oblique 30° view showing severe mitral annular calcification.



Figure 3 Gross photograph of the inflow tract of the left heart showing a grossly dilated left atrium and ventricle. Both mitral leaflets show thickened and fused chordae. There is massive calcification of the posterior mitral annulus. Some of the calcified foci are visible as irregular nodules along the atrial surface of posterior leaflet.

valve repair or replacement^[8,9]. Mitral valve repair may not be technically feasible in such cases because of severe calcification and the difficulty of suturing at the calcified site, mandating a prosthetic valve replacement. Intra-atrial valve placement instead of usual positioning at the mitral annulus has also been tried in such cases when there is technical difficulty in reconstruction or suturing at the calcified annulus^[10,11]. The reoperation rate and technical complications are also higher in such cases^[8,10,12]. Unfortunately, the index case died prior to surgical intervention, and autopsy confirmed severe MAC and also RHD.

REFERENCES

1. Schott CR, Kotler MN, Parry WR, Segal BL. Mitral annular calcification. Clinical and echocardiographic correlations. *Arch Intern Med* 1977; **137**: 1143-1150
2. Chopra P, Wanniang J, Sampath Kumar A. Immunohistochemical and histochemical profile of Aschoff bodies in rheumatic carditis in excised left atrial appendages: an immunoperoxidase study in fresh and paraffin-embedded tissue. *Int J Cardiol* 1992; **34**: 199-207
3. Fernandes RM, Branco LM, Galrinho A, Timóteo AT, Tavares A, Feliciano J, Oliveira R, Fiarresga A, Mamede A,

- Banazol N, Roquette J, Ferreira RC. Caseous calcification of the mitral annulus. A review of six cases. *Rev Port Cardiol* 2007; **26**: 1059-1070
- 4 **Kato M**, Nakatani S, Okazaki H, Tagusari O, Kitakaze M. Unusual appearance of mitral annular calcification mimicking intracardiac tumor prompting early surgery. *Cardiology* 2006; **106**: 164-166
- 5 **Salem DN**, O'Gara PT, Madias C, Pauker SG. Valvular and structural heart disease: American College of Chest Physicians Evidence-Based Clinical Practice Guidelines (8th Edition). *Chest* 2008; **133**: 593S-629S
- 6 **Akram MR**, Chan T, McAuliffe S, Chenzbraun A. Non-rheumatic annular mitral stenosis: prevalence and characteristics. *Eur J Echocardiogr* 2009; **10**: 103-105
- 7 **Schneider J**, Bezabih K. Causes of sudden death in Addis Ababa, Ethiopia. *Ethiop Med J* 2001; **39**: 323-340
- 8 **Grossi EA**, Galloway AC, Steinberg BM, LeBoutillier M, Delianides J, Baumann FG, Spencer FC, Colvin SB. Severe calcification does not affect long-term outcome of mitral valve repair. *Ann Thorac Surg* 1994; **58**: 685-687; discussion 688
- 9 **Nomura A**, Fukuda I, Daitoku K, Fukui K. Enucleation of calcium core and in-situ valve replacement for massive posterior mitral annular calcification. *Interact Cardiovasc Thorac Surg* 2011; **12**: 652-654
- 10 **Atoui R**, Lash V, Mohammadi S, Cecere R. Intra-atrial implantation of a mitral valve prosthesis in a heavily calcified mitral annulus. *Eur J Cardiothorac Surg* 2009; **36**: 776-778
- 11 **Santana O**, Lamelas J. Intra-atrial placement of a mitral prosthesis in a patient with severe mitral annulus calcification: a case report. *Heart Surg Forum* 2010; **13**: E25-E27
- 12 **Sakamoto Y**, Hashimoto K, Okuyama H, Ishii S, Taguchi S, Kagawa H, Morita K. Repair of mitral periprosthetic leak secondary to severe annular calcification. *J Heart Valve Dis* 2007; **16**: 132-135

S- Editor Cheng JX L- Editor Cant MR E- Editor Li JY