

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

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

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

Accepted: December 17, 2011

Published online: March 26, 2012

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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>

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.

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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;

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,

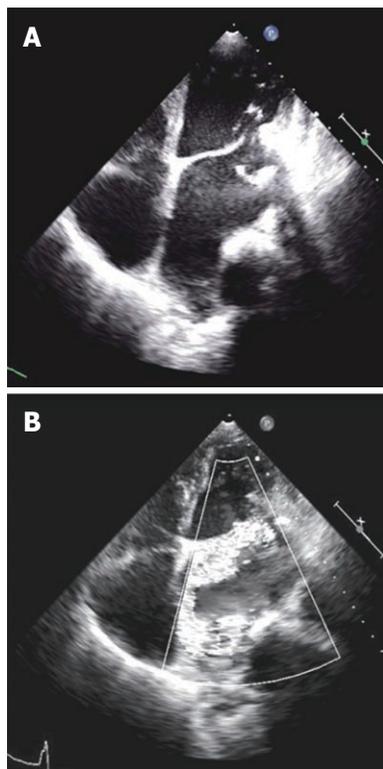


Figure 1 Echocardiography in apical 4 chamber view. A: Calcified, thickened and retracted posterior mitral leaflet with adjacent mitral annular ring calcification; B: Color Doppler showing severe mitral regurgitation.

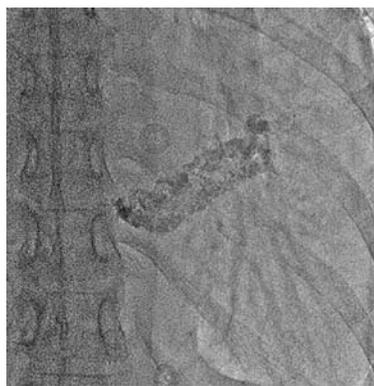


Figure 2 Fluoroscopy image in right anterior oblique 30° view showing severe mitral annular calcification.



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.

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

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.

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