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## Coconut Atrium in Long-Standing Rheumatic Valvular Heart Disease

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Study Design A  
Data Collection B  
Statistical Analysis C  
Data Interpretation D  
Manuscript Preparation E  
Literature Search F  
Funds Collection G

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**Conflict of interest:** None declared

**Patient:** Male, 76  
**Final Diagnosis:** Rheumatic valvular heart disease  
**Symptoms:** Breathlessness and leg edema  
**Medication:** —  
**Clinical Procedure:** Medical treatment for heart failure  
**Specialty:** Cardiology

**Objective:** Rare disease  
**Background:** Complete calcification of the left atrium (LA) is called “coconut atrium”, which decreases the compliance of LA, leading to the elevation of LA pressure that is transmitted to the right-side of the heart. The pathogenesis of LA calcification in patients with rheumatic heart disease is unknown; however, possible mechanisms include chronic strain force in the atrial wall and inflammation. We report here a patient with long-standing rheumatic valvular heart disease with coconut atrium.

**Case Report:** A 76-year-old man presented with breathlessness and leg edema due to right-sided heart failure. He was diagnosed with rheumatic fever at 8 years of age. Mitral commissurotomy and the mitral and aortic valve replacement were previously performed to treat mitral and aortic valvular stenosis. The profile view of the chest X-ray indicated a diffuse calcified outline of the LA wall. A transthoracic echocardiogram revealed pulmonary hypertension and dilatation of both atria. Moreover, computed tomography showed nearly circumferential calcification of the LA wall. Despite intense medical treatment, he succumbed to heart failure. An autopsy demonstrated that the LA was markedly dilated, its wall was calcified, and its appearance was similar to the surface of an atherosclerotic aorta. Microscopic examination revealed intensive calcification in the endocardium. Minimal accumulation of inflammatory cells was noted. Although slight fibrosis was observed, the cardiac musculature was preserved.

**Conclusions:** To the best of our knowledge, this is the first report that identifies the histological changes of LA calcification associated with long-standing rheumatic valvular heart disease.

**MeSH Keywords:** Heart Atria • Heart Failure • Rheumatic Heart Disease

**Full-text PDF:** <http://www.amjcaserep.com/abstract/index/idArt/892449>



## Background

Ectopic calcification in the heart is often observed in patients with various cardiac diseases or other clinical conditions. For example, mitral annular calcification is a common feature of patients with chronic kidney disease or very elderly patients [1]. However, massive calcification of the left atrium (LA) is a rare clinical condition that involves the left atrial appendage, left atrial free wall, mitral valve apparatus, or all 3 in more severe cases [2]. Complete calcification of the LA has been described as “coconut atrium” [3]. There are several reports of patients with LA calcification as an uncommon complication of long-standing rheumatic valvular disease; however, there are no reports of histological findings. Here, we present the histological analysis of marked LA calcification in a patient with long-standing rheumatic heart disease.

## Case Report

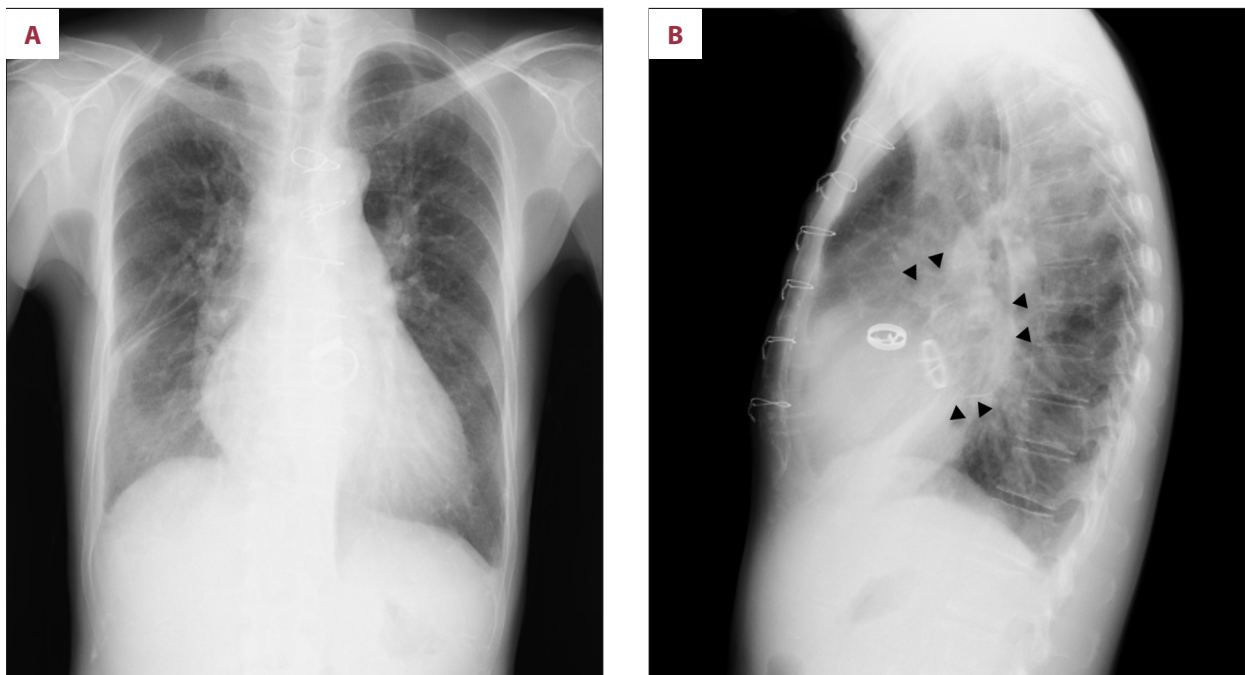
A 76-year-old man was admitted due to complaints of breathlessness and bilateral leg edema. His medical history is as follows: rheumatic fever at the age of 8 years; aortic and mitral stenosis at the age of 30 years, requiring open mitral commissurotomy; and mitral and aortic valve replacement for chronic rheumatic mitral and aortic valvular disease was performed at the age of 60 years.

On admission, his blood pressure was 128/86 mmHg and pulse rate was 70 beats per minute. The jugular vein was dilated,

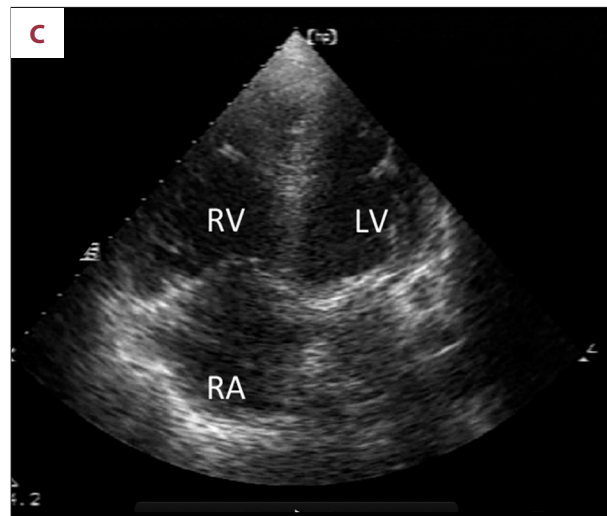
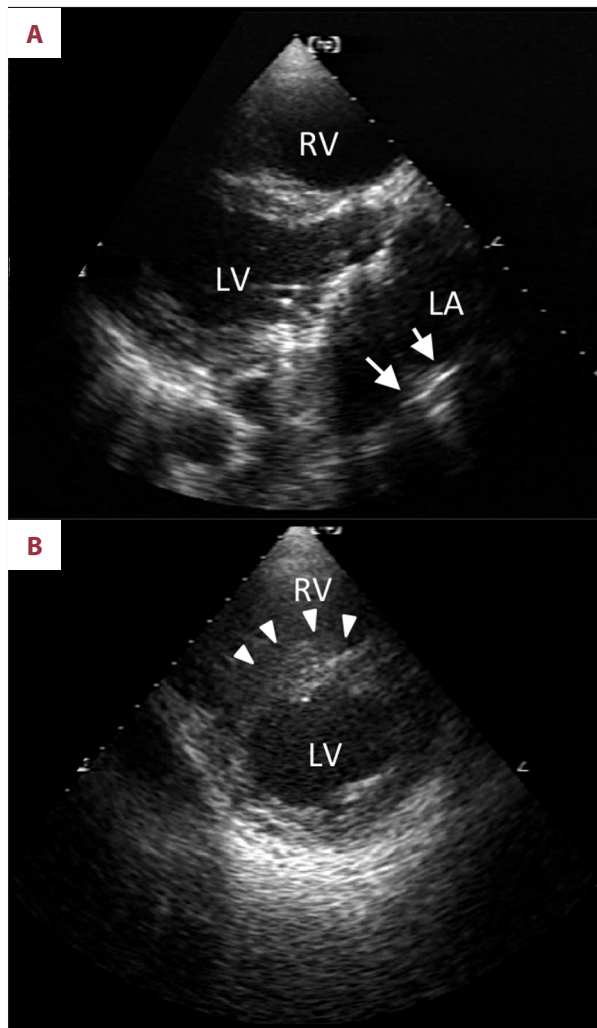
**Table 1.** Laboratory data on admission.

WBC 9400/mm <sup>3</sup>	TP 7.9 g/dl
RBC 270×10 <sup>4</sup> /mm <sup>3</sup>	Alb 4.3 g/dl
Ht 30.5 %	T-bil 0.73 mg/dl
Hb 9.6 g/dl	AST 31 IU/l
MCV 113 fl	ALT 13 IU/l
MCH 35.6 pg	Cr 2.2 mg/dl
Platelet 14.8×10 <sup>4</sup> /mm <sup>3</sup>	BUN 46.3 mg/dl
	Na 135 mEq/l
	Cl 87 mEq/l
	K 5.1 mEq/l
	Ca 9.8 mEq/l
	P 3.8 mEq/l
	BNP 2327 pg/ml

and pitting edema was present on both legs. A systolic murmur was audible. An electrocardiogram revealed atrial fibrillation (heart rate 72 beats/min), complete right bundle branch block, and right axis deviation. The frontal view of a chest X-ray showed cardiomegaly with mild lung congestion (Figure 1A). The profile view revealed a diffuse calcified outline of the LA wall (Figure 1B). Laboratory data indicated anemia and renal dysfunction (Table 1). The serum level of BNP was markedly elevated (2327 pg/ml).



**Figure 1.** Frontal (A) and profile views (B) of a chest radiograph. Triangles represent calcification.



**Figure 2.** Trans-thoracic echocardiogram on admission. Parasternal long-axis (A), short-axis views (B), and 4-chamber view (C) at end-diastolic phase showing the dilation of the RV and both atria. LV was also mildly dilated. The inversion of the interventricular septum curvature indicated elevated RV pressure, indicated by arrowheads (B). Although the acoustic shadow of a prosthetic valve did not allow complete evaluation, the calcification of the LA wall was observed, indicated by arrows (A).

Transthoracic echocardiography showed dilation of the right ventricle (RV) and inversion of the interventricular septum curvature, indicating elevated RV pressure (Figure 2A, 2B). The estimated systolic pulmonary pressure was 62.7 mmHg and the end-diastolic dimension of the RV was 38 mm. The left ventricle (LV) and both atria were also dilated; the size of the LV end-diastolic dimension was 57 mm, and the dimension of the LA was 56 mm (Figure 2). The wall motion of the LV was mildly reduced and the LV ejection fraction was 52%. Although the acoustic shadow of a prosthetic valve did not allow complete evaluation, calcification of the LA wall was observed (Figure 2A).

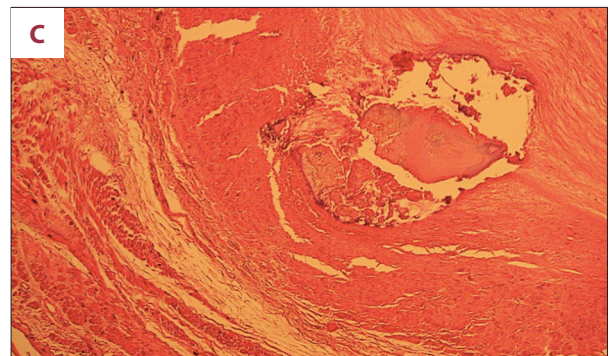
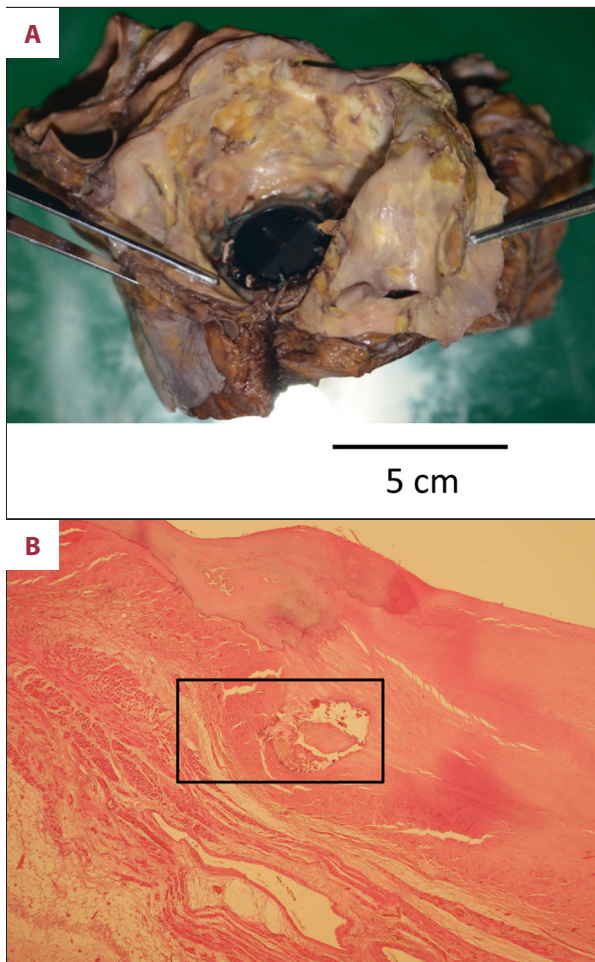
Chest computed tomography showed massive and near circumferential calcification of the LA wall (Figure 3). According to these clinical findings, we attributed his symptoms to right-sided heart failure.

Although the patient underwent intensive treatment with diuretics, dobutamine, and adaptive servo-ventilation therapy,



**Figure 3.** Computed tomography shows calcification of the left atrium wall.

he died due to heart failure. Autopsy showed marked dilation of the LA and calcification of the wall. The endocardial surface of the LA appeared similar to that of an atherosclerotic aorta (Figure 4A). Microscopic analysis revealed that intensive calcification under the endocardial layer (Figure 4B, 4C). There were few minimal inflammatory cells and some fibrotic changes were observed. Lipid deposition in the atrial wall was not detected. There was no specific finding in the respiratory system or gastro-intestinal tract.



**Figure 4.** Gross appearance of the left atrium (LA) (A). Microscopic view of the LA (B, C). A low-power view (B) of the LA wall revealed intensive calcification under the endocardial layer. Panel C is a high-power view of the area, indicated by the rectangle in B.

are no published studies that define the mechanisms of calcification in the myocardium.

Chronic mitral stenosis causes pressure overload eventually stretches the walls of the LA. Such long-term hemodynamic forces in the LA induce dysfunction of the endocardium. During atherosclerosis, endothelial dysfunction plays an important role in vascular calcification. For example, Yao et al. demonstrated that the endothelium is a source of osteoprogenitor cells in vascular calcification [5]. We speculate that dysfunction of endocardium induced by chronic pressure on the LA wall played a significant role in the atrial calcification in the present case. Further inflammation during the healing process of rheumatic carditis may be involved in the pathogenesis of rheumatic heart disease [6]. Although we observed the fibrotic changes here, there were few inflammatory cells in the LA wall, suggesting that active inflammation occurred during a very early phase of the rheumatic fever. These factors induce structural and electrical remodeling of the atrium, which lead to the development of atrial fibrillation. Indeed, atrial fibrillation persisted for a long time in our patient. The advanced electroanatomical remodeling of the LA exacerbates the hemodynamics of heart failure.

## Discussion

LA calcification was originally described in 1898. Calcification of the LA is an uncommon complication of long-standing rheumatic valvular heart disease, and complete calcification of the LA has been described as “coconut atrium” [3]. These characteristics are similar to those of the present case. Calcification of the LA decreased its compliance, which led to the transmission of elevated LA pressure to the right side of the heart.

The pathogenesis of LA calcification with rheumatic valvular heart disease is unknown; however, possible mechanisms include chronic strain force on the atrial wall and inflammation. Recent research on the mechanisms of ectopic calcification such as arterial calcification indicates that well-regulated processes, similar to those that drive osteogenesis, mediate vascular calcification. It is hypothesized that a subpopulation of vascular cells retains the potential to differentiate into osteoblasts and may play a pivotal role in vascular calcification by forming mineralized nodules and expression of osteoblast-specific proteins such as osteocalcin, bone morphogenetic protein-2, and alkaline phosphatase [4]. However, there currently

## Conclusions

We report here an autopsied patient with coconut atrium, which is a rare complication of long-standing rheumatic valvular heart disease. Our observation of intensive calcification under the endocardial layer of the coconut atrium is, to the best of our knowledge, the first demonstration of the histological changes that occur during LA calcification. Moreover, our analysis highlights the substantial variability in ectopic calcification of cardiovascular diseases.

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