

Massive Left Atrial Calcification in Late Rheumatic Heart Disease



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INTRODUCTION

The process of active inflammation of the heart tissues caused by acute rheumatic fever might result in persistent chronic damage after the acute insult, leading to rheumatic heart disease (RHD). Despite the fact that the valvular endocardium is mostly commonly responsible for the clinical manifestations of RHD, it can involve all tissues of the heart. Within these tissues, the presence of massive left atrial (LA) calcification is extremely rare and has been proposed to represent the result of long-standing RHD, although the mechanisms implied are not fully elucidated.¹ We present a case of complete LA mural calcification of the left atrium (LA) 20 years after surgical mitral valve replacement (MVR) for RHD.

CASE PRESENTATION

We report the case of a 73-year-old woman with a medical history of acute rheumatic fever in childhood. She did not have precise information about any treatment received during the acute phase of the disease. When she was in her 40s, she started developing heart failure (HF) symptoms and was then diagnosed with atrial fibrillation and rheumatic mitral stenosis. She mentioned that a mitral valvuloplasty was performed afterward, but further details about this intervention were not available. About 10 years after that first procedure, she underwent MVR with a mechanical prosthesis. Importantly, she had always been adherent to her medications, which included bisoprolol for heart rate control and anticoagulation with coumadin. According to her hematologic controls, her international normalized ratio levels have always been within the therapeutic range. She has not been hospitalized since the MVR and denied any acute worsening of her functional capacity.

The patient started noticing progressive exertional dyspnea 2 weeks prior that limited her daily activities and was often associated with palpitations and dry cough that worsened when she lay flat. She denied fever, chills, or other extracardiac symptoms, including hoarseness or changes in her voice.

Vital signs were normal, and physical exam revealed the presence of tachypnea and fine lung crackles. The electrocardiogram showed atrial fibrillation with a ventricular response of 100 beats per minute, with

VIDEO HIGHLIGHTS

Video 1: Apical four-chamber view (**A**) and modified three-chamber view (**B**): severely enlarged LA with increased thickness and hyperechoic atrial walls and IAS with akinetic motion throughout the cardiac cycle. LV, Left ventricle.

View the video content online at www.cvcasejournal.com.

QRS axis on 90°. N-terminal pro-brain natriuretic peptide levels were 492 pg/mL (normal value < 300 pg/mL), supporting the HF diagnosis.

Patient was admitted and started on intravenous diuretics, with good clinical response. Chest radiography showed marked cardiomegaly, rectified left middle arch, and the presence of a well-defined circular hollow structure with calcified borders in the area corresponding to the LA location (Figure 1A, white arrows) and the presence of a prosthetic mechanical valve ring at the bottom of the LA. Lateral radiography image confirmed this finding, showing a large, rounded, calcified structure located posteriorly to the mitral prosthesis (Figure 1B, white arrows).

Considering these findings, potential etiologies related to hypercalcemic status were assessed. Calcium, vitamin D, and parathyroid hormone levels were within normal values. Creatinine levels were 1.3 mg/dL, and glomerular filtration rate calculated by the modification of diet in renal disease equation was 40.2 mL/minute/1.73 m². She was not taking any procalcific medications, and, except for mammograms every 2 years, she denied any chest radiation.

Transthoracic echocardiography showed that the left and right ventricles were normal in size and function and that both atria were severely enlarged. Mitral prosthesis was normal in structure and function, with freely movable disks and a mean diastolic gradient of 5 mm Hg. The LA wall and interatrial septum (IAS) were markedly thickened and hyperechoic (Figure 2A and B, white arrows), and complete absence of LA wall motion during the cardiac cycle was also noted (Video 1). Tricuspid regurgitation was moderate, with an estimated right ventricular systolic pressure of 52 mm Hg.

Chest computed tomographic (CT) scan confirmed severe biatrial enlargement and revealed the presence of massive LA calcification that also extended into the IAS, without involving the right atrium (Figure 3A-C).

The patient continued to do well and was discharged home after 5 days of hospitalization.

DISCUSSION

First described in 1912 by Oppenheimer,² massive calcification of the LA wall, also known as “coconut atrium,” is thought to be the end result of repeated and extensive rheumatic LA inflammation,

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Conflicts of Interest: None.

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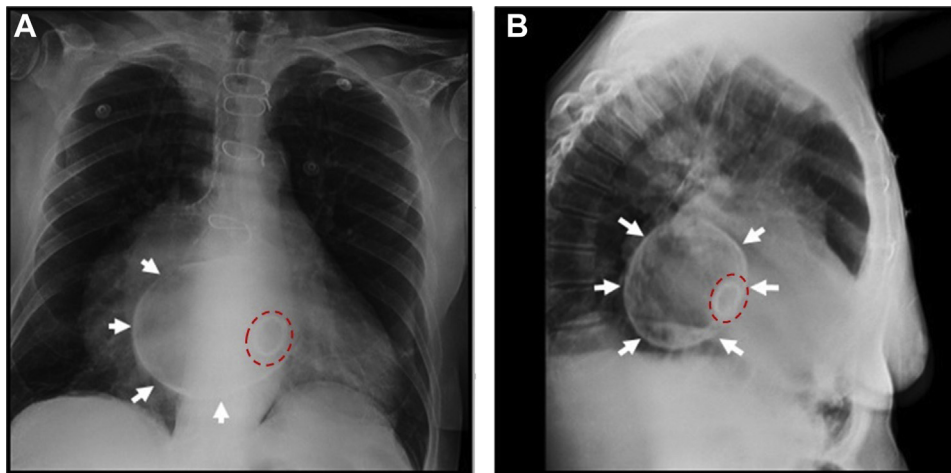


Figure 1 Chest radiography. **(A)** Anteroposterior projection shows calcified rim (*white arrows*) located within the LA topography and its spatial relationship with the MVR (*red dashed circle*). **(B)** Lateral projection corroborates these findings, showing the size, shape, and localization of the calcified borders of this radiolucent and circular structure (*white arrows*). The MVR can be identified within the *red dashed circle*.

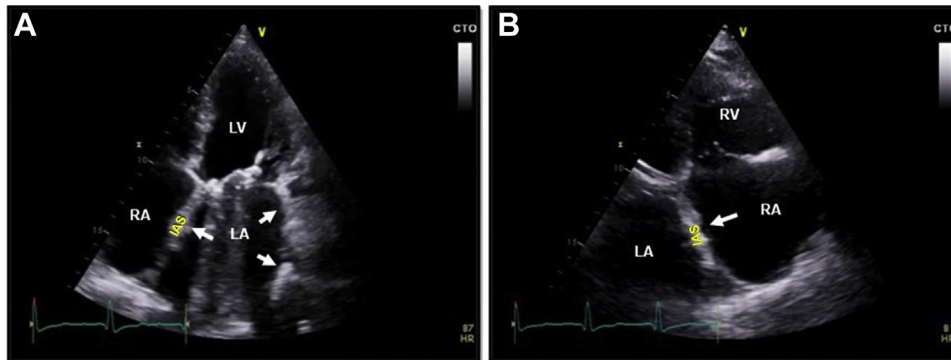


Figure 2 Transthoracic echocardiography images. **(A)** Apical four-chamber view shows severely enlarged LA with increased thickness and hyperechogenicity of its walls involving the IAS (*white arrows*). Acoustic shadow corresponding to the mitral mechanical prosthesis is also noted. **(B)** Right ventricular inflow view shows severe RA enlargement and corroborates the IAS involvement (*white arrow*). RA, Right atrium; RV, right ventricle.

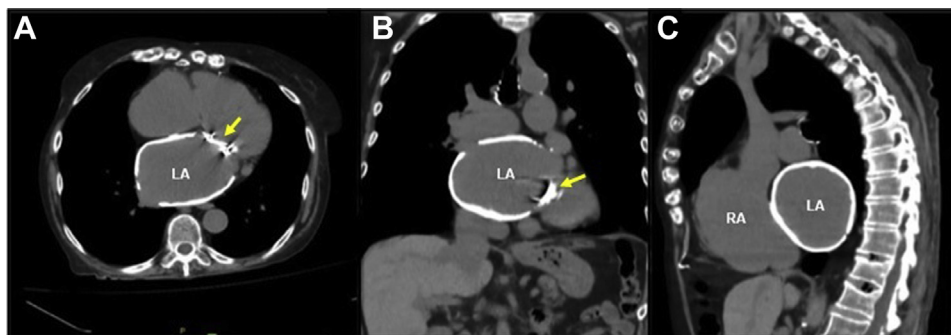


Figure 3 Chest CT. Notice the presence of severe biatrial enlargement and massive calcification of the LA walls and IAS, giving this chamber the appearance of an open coconut, in the **(A)** axial, **(B)** coronal, and **(C)** sagittal planes. The *yellow arrows* show the position of the MVR.

beginning with focal areas of endocardial ulceration and progressing to chronic fibrotic changes with subendocardial plaques of calcium. While the name “porcelain atrium” refers to the LA calcification that involves the LA appendage, free wall, and mitral valve apparatus, but spares the IAS,³ the term “coconut atrium” is reserved for the

involvement of the entire LA circumference.¹ Whether there are different pathophysiological mechanisms implied in the involvement or sparing of the IAS is still uncertain. Importantly, when this structure is affected, as in the presented case, it represents a major obstacle for surgical and interventional procedures.⁴

Interestingly, the degree of valvular calcification seems to have no relation to the presence LA calcification. Cases of massive LA calcification were reported prior to the valve surgery era,² but it has also been described more than 20 years after MVR.⁴ Long-lasting processes of inflammation, ulceration, and calcification of the LA wall and chronic strain forces in the setting of mitral disease have been suggested as possible mechanisms associated with this condition.⁵ This might discourage the hypothesis that hemodynamic factors are the main drivers of the massive LA dilation and calcification and could also explain why the right atrium is not affected. Additionally, this condition has been described as a complication of long-term hemodialysis⁶ and following thoracic radiotherapy.⁷

Women are more frequently affected (approximately 74% of cases),⁸ and although most patients remain asymptomatic for long periods of time, the progressive LA noncompliance and increased pressure status might lead to pulmonary hypertension, severe tricuspid insufficiency, and development of symptoms like progressive dyspnea and chronic dry cough.¹

Although radiography of the left lateral side of the chest provides a good assessment of the extent and localization of the LA calcification, multislice CT provides high-resolution three-dimensional imaging of the LA anatomy and its relationship to contiguous structures as well as better assessment of the localization and extent of the calcification, as seen in our patient.

Surgical removal of the internal cast of calcium has been described but has an operative mortality of up to 25%, which is related to the difficulty of entering the LA, potential embolization, and impaired hemostasis.⁸

CONCLUSION

This case shows that RHD is not limited to valvular disease, and extensive involvement of other structures might develop even

several years after the correction of the valvulopathy. Among these, extensive LA calcification should be especially suspected in patients with a history of RHD who present with new-onset HF that is not otherwise explained.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.case.2022.02.006>.

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