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Porcelain left atrium associated with pulmonic valve disease

Nicholas King *, Cory Jackson , and Sandip K. Zalawadiya

Department of Internal Medicine, Vanderbilt University Medical Center, 1211 Medical Center Drive, Nashville, TN 37232, USA

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Left atrial calcification is a rare radiographic finding. Here, we describe a case of left atrial calcification associated with congenital pulmonic valve disease requiring multiple interventions.

A 50-year-old female presented with exertional dyspnoea, hypoxia, and orthopnoea. History was significant for congenital pulmonic valve stenosis surgically repaired at age 4 that degenerated to severe insufficiency requiring surgical pulmonary valve replacement at age 31. The bioprosthetic valve subsequently developed severe stenosis that required balloon valvuloplasty 8 months prior to presentation. Surface echocardiogram on admission revealed severe pulmonic insufficiency and significant left atrial calcifications. The mitral valve had posterior annular calcification with normal leaflet mobility and function (*Video 1*). Computed tomography chest revealed a heavily calcified left atrium (*Figure 1*). The patient underwent percutaneous endovascular pulmonic valve replacement during which invasive haemodynamic pressure measurements revealed an elevated pulmonary capillary wedge pressure and normal left ventricle end-diastolic pressure consistent with a stiff left atrium. She was diuresed and had significant improvement in her symptoms.

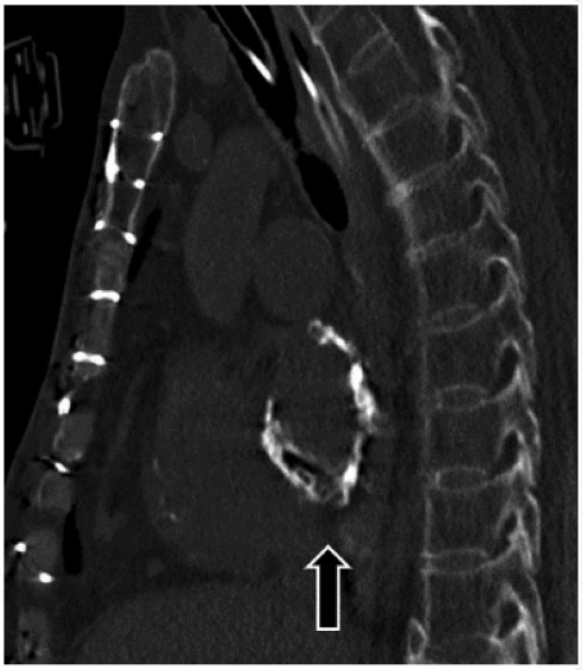
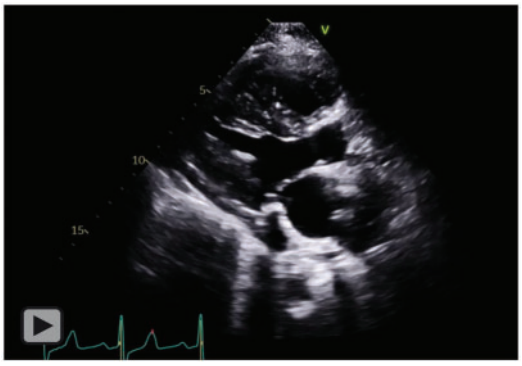


Figure 1 A sagittal image from a non-contrasted chest computed tomography. Note the diffuse calcifications involving the left atrium (black arrow).



Video 1 A parasternal long-axis view from a surface echocardiogram. Note the dense calcifications of the left atrial and normal mitral leaflet mobility.

* Corresponding author. Tel: +1 615 838 7285, Fax: +1 269 210 2719, Email: nicholas.king@vumc.org
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A 'porcelain' left atrium, better termed massive left atrial calcification (MLAC), was first described by Claude and Levaditi[1] in 1891, and since that time has continued to be sporadically published in the medical literature. Massive left atrial calcification is most commonly associated with rheumatic heart disease; other associations include end-stage renal disease, tuberculosis, radiation, and cardiac surgery.[2,3] Its cause is ultimately unknown, but the associated conditions suggest that chronic inflammation is a major driver of MLAC.[3]

The treatment for MLAC is surgical endoatrioectomy, however, this is only possible in a subset of MLAC patients who do not have involvement of the interatrial septum, the endocardium, or the mitral annulus.[3] Medical management consists of diuresis for venous congestion and anticoagulation if indicated for atrial fibrillation or thrombus.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

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