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CASE REPORT
Cardiac imaging

Case report: Metastatic cardiac calcifications in a patient with a history of rickets

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Background Metastatic cardiac calcifications are often seen in patients with renal failure and dialysis associated with vascular calcification and calcifications in other organs. There is little to no evidence of metastatic cardiac calcification in patients with a history of rickets.

Case summary A 40-year-old patient with a history of rickets treated in infancy and no personal history of cardiovascular or renal disease came for a periodical examination. Transthoracic echocardiogram showed an important calcification in the mitral annulus and the mitro-aortic junction. Computed tomography (CT) showed marked calcification affecting the heart fibrous skeleton. Coronary arteries were not affected.

Discussion Distribution of heart calcification seen both with echocardiography and CT is important to report so they can be identified as dystrophic or metastatic. These two have different aetiologies so the description may help identify the possible cause.

Keywords Case report • Metastatic cardiac calcification • Mitro-aortic junction • Rickets • Echocardiography • Computed tomography

Learning points

- Metastatic cardiac calcification associated with a history of rickets is very rare.
- The description of the distribution and morphology of the calcifications with the cardiovascular images can help identify the aetiology of the cardiac calcification.

Introduction

Metastatic cardiac calcifications are often associated with secondary hyperparathyroidism related to renal failure and dialysis.[1] Rarely, they may be found in patients with primary hyperparathyroidism, or with calcium and vitamin D dietary deficiency.[2] This report describes a case of cardiac calcifications in a patient with a history of rickets in childhood.

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Timeline

Time	Event
Year 1989–90	Diagnosis of rickets. Treatment with calcium and vitamin D.
Day 0 (1st August 2019)	The patient is admitted at the medical institute for a periodical clinical examination. Electrocardiogram shows no abnormal findings. Transthoracic echocardiogram shows significant cardiac calcification.
Day 1	Computed tomography shows vast calcification in the heart. No involvement in coronary arteries revealed.
November 2019	The patient remained asymptomatic in follow-up with no new positive findings.

Case presentation

A 40-year-old male presented for a routine outpatient checkup. His medical history included rickets during childhood treated with calcium and vitamin D. He had no prior personal or family history of cardiovascular disease. The patient was asymptomatic. Physical examination showed blood pressure (120/70 mmHg) and heart rate (65 b.p.m.) within a normal range, no heart murmurs, and a normal respiratory examination. The electrocardiogram (EKG) presented a QRS axis between 90° and 120° and a PR segment of 120 ms; the rest of the tracing was within normal limits (*Figure 1*). Laboratory findings were within a normal range, including calcium and phosphate plasma levels. Data regarding vitamin D and parathyroid hormone (PTH) plasma levels were not available.

An echocardiogram, done as part of a routine checkup, revealed marked calcification of the heart's fibrous skeleton, including the mitral annulus, the mitro-aortic junction and the aortic valve (*Figure 2* and *Video 1*). Both mitral leaflets were preserved, maintaining normal functioning, with only a minimal regurgitation towards the posterior wall of the left atrium (*Video 2*). The aortic valve had a normal opening and closing movements, with no regurgitation. Right valves were preserved. Anterograde velocities were normal. Both right and left ventricles had preserved systodiastolic functions. The left atrium was moderately dilated, while the right atrium was in a normal range. The pericardium did not show any abnormalities, and there were no signs of constriction physiology.

Computed tomography (CT) exhibited vast calcifications with irregular borders and a glomerular distribution along with the fibrous skeleton. The main areas involved were the proximal interventricular septum, the mitro-aortic junction, the mitral annulus, and the atrioventricular groove. The aortic valve also displayed calcification

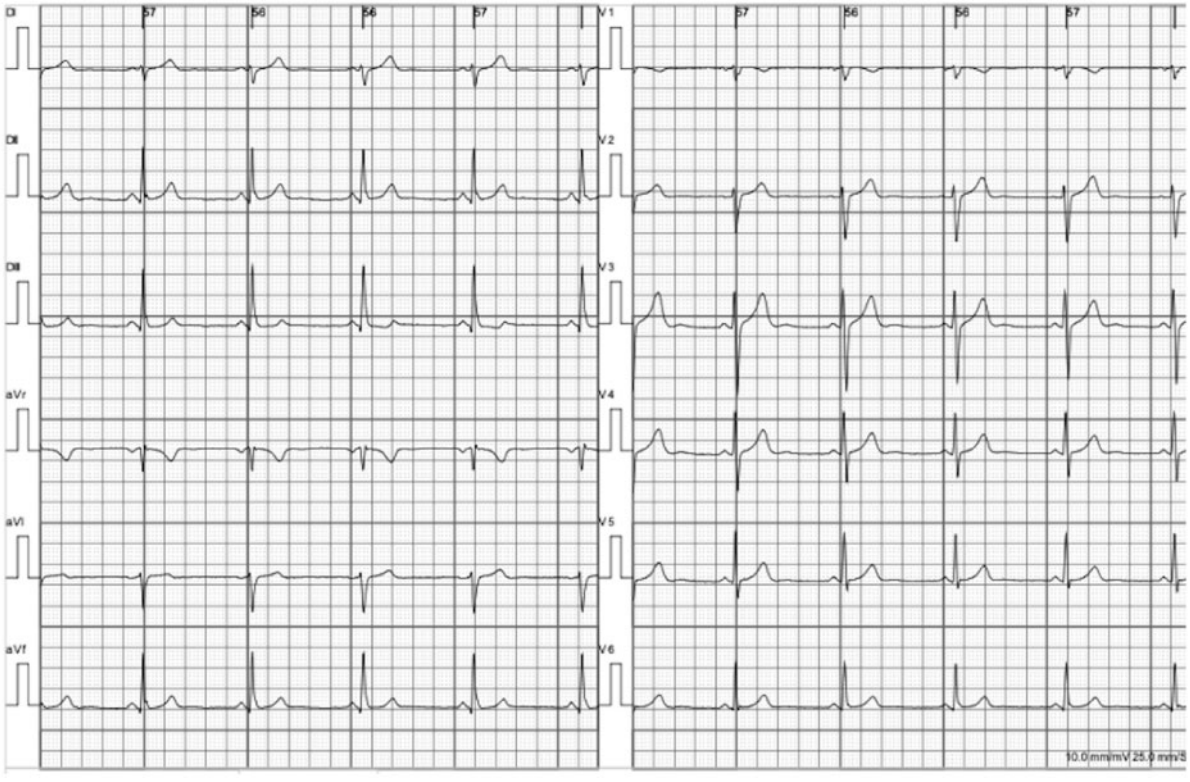


Figure 1 12-Lead electrocardiogram (EKG).

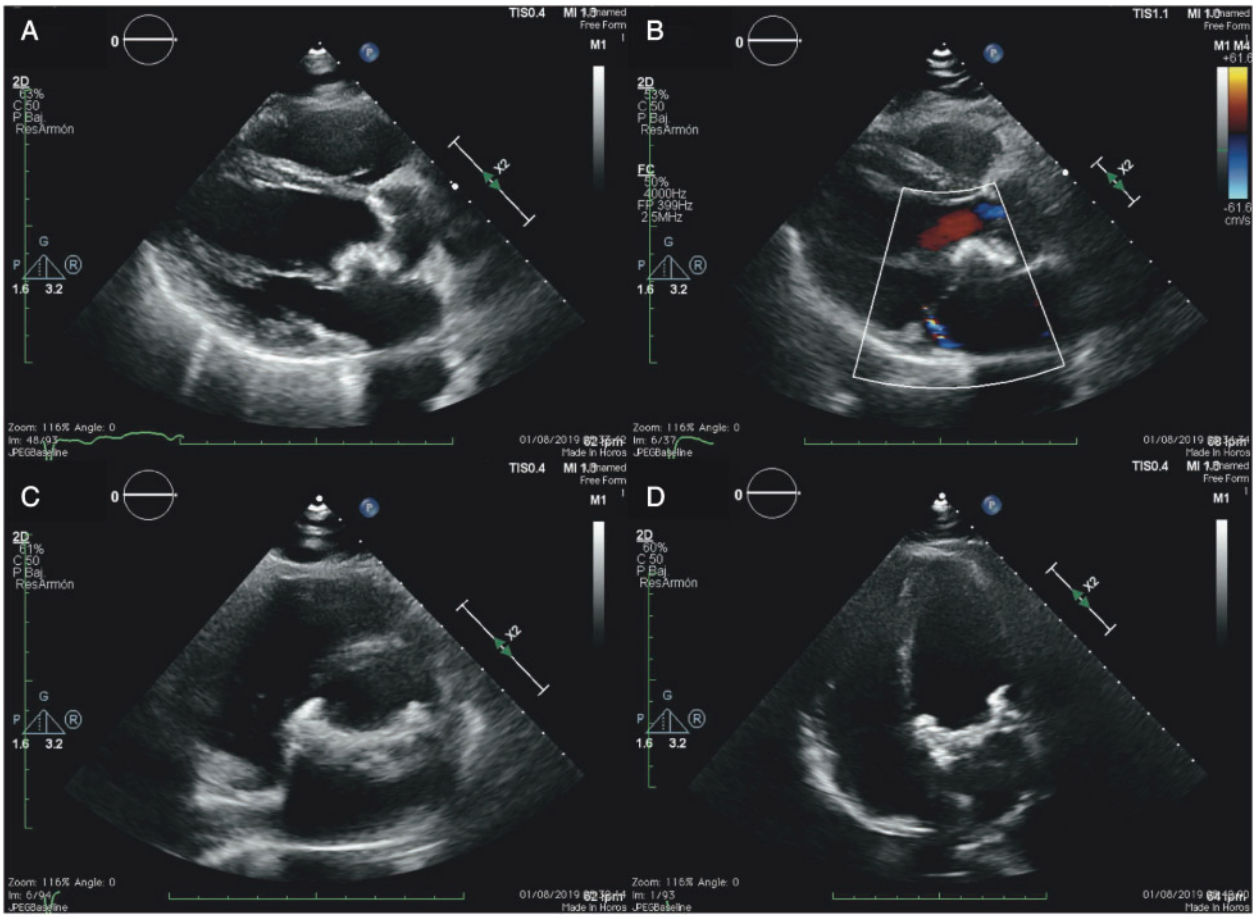
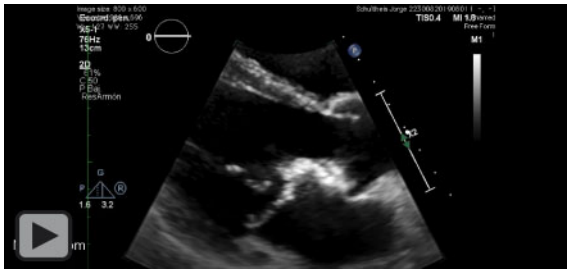
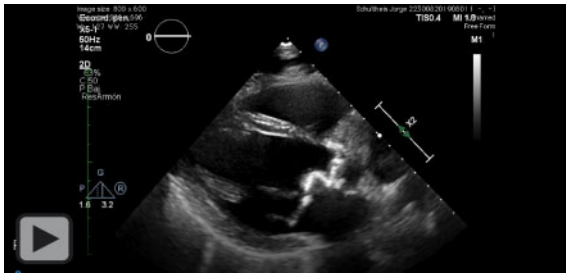


Figure 2 Two-dimensional transthoracic echocardiogram. (*A*) Long parasternal view showing the calcification of the mitro-aortic junction, involving the aortic valve and aortic root. (*B*) Minimal mitral regurgitation with the jet directed towards the posterior atrial wall. (*C*) Parasternal short-axis view exhibiting the calcification of the heart's fibrous skeleton along the fibrous trigonous. (*D*) Apical four chamber off-axis view showing calcifications within the atrioventricular groove and the mitral annulus.



Video 1 Parasternal long axis of the patient showing the marked calcification involving the mitral annulus, the mitro-aortic junction and the aortic valve. Notice the sparring of both leaflets of the mitral valve.



Video 2 Parasternal long axis of the patient with zoom focusing on the mitral valve. Notice that the calcification affected the mitral annulus but both leaflets were unaffected, as were the subvalvular apparatus. The valvular opening and closing motions were preserved.

nodules in the aortic commissures: between the non-coronary and the left coronary cusps, and between the non-coronary and the right coronary cusps (*Figures 3 and 4*).

A CT coronary angiography revealed no significant lesions, presenting with a calcium coronary score of 0 Agatston units (AU). No extracardiac metastatic calcifications were observed.

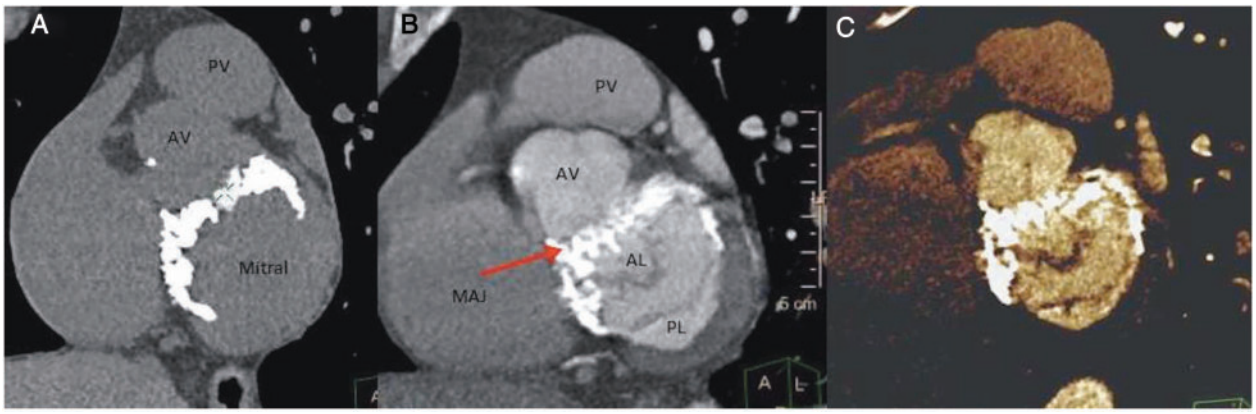


Figure 3 Multiplanar reconstruction of cardiac computed tomography scan. Short-axis view of the mitral annulus showing the mass calcification of the heart's fibrous skeleton, mitral annulus and the atrioventricular groove. (*A*) Non-contrast CT. (*B*) Gated contrast computed tomography. (*C*) Volume-rendered. AL, anterior mitral leaflet; AV, aortic valve; MAJ, mitro-aortic junction; PL, posterior mitral leaflet; PV, pulmonary valve.

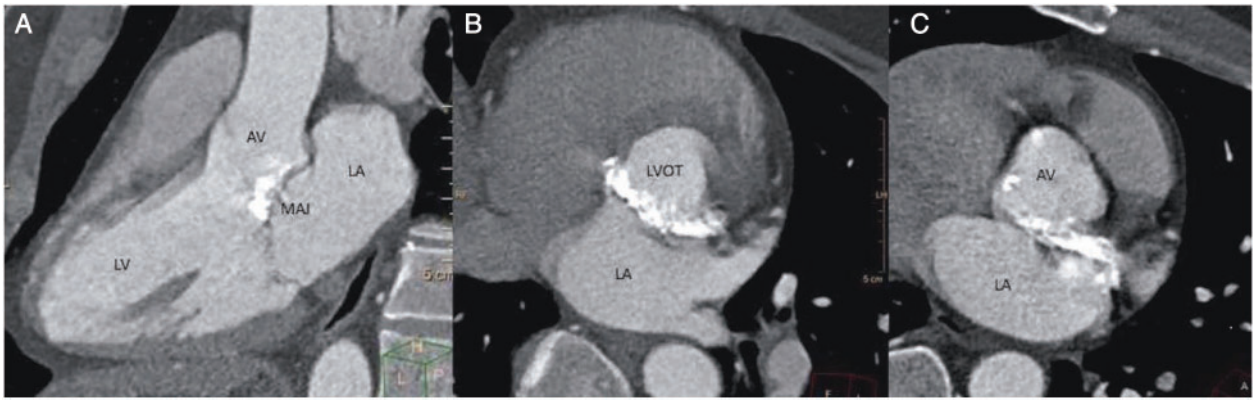


Figure 4 Contrast cardiac computed tomography showing calcification of aortic-mitral continuity. (A) Three chamber view. (*B*) Short-axis cross-section view at the left ventricular outflow tract. (*C*) Short-axis cross-section view of the aortic valve. AV, aortic valve; LA, left atrium; LVOT, left ventricular outflow tract; LV, left ventricle: MAJ, mitro-aortic junction.

It was decided that no intervention would be required. The patient remained asymptomatic and was referred to the Cardiology Department for outpatient follow-up. At the most recent follow-up, 4 months after admission, the patient remained asymptomatic.

Discussion

Cardiac calcifications have different aetiologies, but almost always suggest an underlying disease.[1] They present with two basic distributions: dystrophic and metastatic. Since each distribution is associated with different aetiologies, they are important to report.[3]

Dystrophic calcifications occur in the presence of local tissue damage, frequently of ischaemic cause. They tend to be distributed in regions affected by necrosis and therefore have a linear morphology. Metastatic calcifications are usually a result of systemic diseases affecting the phospho-calcium metabolism. As a consequence, their distribution tends to be irregular, with glomerular-like deposits.

Alterations in vitamin D metabolism and phospho-calcium homeostasis are found in many non-skeletal tissue diseases, including cardiovascular diseases.[5] For example, vitamin D deficiency, related to rickets in childhood and osteomalacia in adulthood, is associated with hypertension, diabetes mellitus, dyslipidaemia, and vascular calcifications.[4,6–8] Hypercalcaemia often presents with cardiac calcifications on the mitral and tricuspid annuli or, less frequently, on the aortic or pulmonary valves.[9] Cardiac calcifications can also occur in patients with hypocalcaemia. Schuster *et al.*[10] reported the case of a 10-year-old boy with pseudohypoparathyroidism, hypocalcaemia, and

elevated PTH levels, with calcification of the interventricular septum. The authors attributed this finding to the high levels of PTH.

Our patient had a history of rickets in childhood, a disease associated with alterations in the phospho-calcium metabolism and high PTH levels, both related to metastatic cardiac calcifications. We believe this childhood disease could be related to the finding of significant cardiac calcifications in this patient as an adult.

Due to lack of clinical evidence, it is difficult to establish an appropriate follow-up for this patient. However, since studies have shown the progression of mitral annulus calcifications on CT scans performed only 2 years apart,[11,12] we suggest yearly follow-up including physical examination, EKG, echocardiography, and CT scans. To the best of our knowledge, this is the first case report of a patient with metastatic cardiac calcifications presenting with rickets in childhood as the only plausible underlying cause.

Lead author biography

Matias Lopez Avecilla was born in Buenos Aires, Argentina in 1987. He received his medical degree from the Faculty of biomedical Sciences in Austral University, Buenos Aires, Argentina in the year 2012. In 2017, he finished his residency in clinical cardiology at the Sanatorio de la Trinidad Mitre in Buenos Aires. He then stayed as chief of residents for two years. He is now specializing in cardiological images, with his mainly interest being clinical echocardiography.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidelines.

Conflict of interest: none declared.

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