A 70-year-old woman with a diagnosis of chronic lymphocytic leukemia in 2004 was recommenced on chemotherapy with chlorambucil and prednisolone in February 2014 owing to chronic lymphocytic leukemia progression. She stopped responding to the treatment after 3 months and was considered for a newly trialled chemotherapeutic agent. Before this she was referred for a routine 12-lead ECG (Figure 1) and a chest radiograph (Figure 2). These findings prompted further evaluation with transthoracic echocardiography, chest computed tomography (CT), and thereafter cardiac magnetic resonance imaging.

The CT scan demonstrated the presence of extensive intramyocardial calcification within the septum, lateral, and anterior walls, and patchy calcification within the inferior wall and mitral annulus (Figure 3A–3C). There was no involvement of the right ventricle or the pericardium. Although the cardiac magnetic resonance scan showed preserved biventricular function (Movie 1 in the Data Supplement), there were multiple areas of hypointense signal intensity within the myocardium, which corresponded to the areas of myocardial calcification visualized on the CT scan (Figure 3D and 3E). After the administration of gadolinium, these areas showed diffuse late enhancement on cardiac MR (Figure 4). The trans-thoracic echocardiogram confirmed the findings from the CT and cardiac magnetic resonance scans with corresponding areas of myocardial calcification and no evidence of constriction or restrictive physiology (Figure 3F–3I). A diagnosis of myocardial lymphoma was made and the new trial drug commenced. The myocardial calcification findings remained static after treatment; however, the patient noticed a significant symptomatic improvement in her systemic symptoms.

Intramyocardial calcification can occur by 1 of 2 basic mechanisms. In the presence of normal underlying myocardium, metastatic calcification can occur in patients with chronic kidney disease and hyperparathyroidism where there are associated abnormalities in calcium and phosphate metabolism. Dystrophic calcification however occurs as a response to necrotized or abnormal myocardium. In these cases, tiny basophilic granules known as calcospheres accumulate at the periphery of abnormal myocytes and then subsequently grow and coalesce in rings of apatite crystals. This form of myocardial calcification is commonly seen in a localized form as a response to myocardial infarction. More diffuse forms of intramyocardial calcification have been reported in patients with (1) sepsis and aggressive myopericarditis (ie, secondary to H1N1 virus), (2) tuberculosis, (3) rare cardiac tumors (ie, rhabdomyomas and endothelialomas), (4) endomyocardial fibrosis, and in patients with myocardial abscesses. Where advanced calcification exists the commonest clinical manifestation is that of heart failure secondary to restrictive physiology. Treatment options are largely confined to early identification of the underlying condition that may precipitate calcification and to medical management of heart failure when extensive calcification has ensued.

The current case first demonstrates a previously unreported cause of diffuse dystrophic intramyocardial calcification. Second, it shows the relative merits of echocardiography, CT, and cardiac magnetic resonance in providing complimentary diagnostic information in the evaluation of calcific myocardial lesions. Finally, our case highlights the need to consider cardiac involvement in patients who present with a recrudescence of their symptoms from chronic conditions that are known to precipitate intramyocardial calcification.

Disclosures

None.

References


Key Words: lymphoma | tomography, computed, scanners
Figure 1. Twelve-lead ECG demonstrating sinus rhythm and left axis deviation.

Figure 2. Chest radiograph demonstrating areas of calcification within the cardiac silhouette (white arrows).
Figure 3. A to C, Contrast enhanced chest computed tomographic scan showing diffuse dystrophic intramyocardial calcification (white arrows) in the 4-chamber (A), 2-chamber (B), and short-axis views (C). D to F, Corresponding appearances of the intramyocardial calcification on white blood cardiac magnetic resonance imaging sequences. G to I, Corresponding areas of calcification on transthoracic echocardiogram in the subcostal view (G), apical 2-chamber view (F), and parasternal short axis view (I). LA indicates left atrium; LV, left ventricle; RA, right atrium; and RV, right ventricle.

Figure 4. Cardiac magnetic resonance scan showing the delayed gadolinium–enhanced images at the areas of intramyocardial calcification in the 4-chamber (A), 2-chamber (B), and short-axis views (C). LA indicates left atrium; LV, left ventricle; RA, right atrium; and RV, right ventricle.
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Supplemental Material

Video Legends

Cardiac MR cine images of the 4-chamber view (Video 1), 2-chamber (Video 2) and short axis (Video 3) views demonstrating hypointense areas of intramyocardial calcification, normal biventricular function and normal septal wall motion.