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IMAGING VIGNETTE

ADVANCED

CLINICAL VIGNETTE

Stone Heart

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ABSTRACT

We describe a rare case of infiltrative cardiomyopathy characterized by multiple low-signal myocardial lesions consistent with nodular calcifications. A retrospectively detailed clinical history and the use of multimodality imaging enabled us to identify the final diagnosis. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2021;3:1509-1511)

CLINICAL PRESENTATION

A 72-year-old woman with a previous history of suspected tuberculosis, type 2 diabetes, glaucoma, and normal renal function presented with increasing breathlessness. The electrocardiogram showed left bundle branch block and first-degree atrioventricular block (**Figure 1**). Echocardiography showed mild left ventricular (LV) systolic dysfunction, with concentric LV hypertrophy, grade 3/4 LV diastolic dysfunction (E/E′ 60; normal value <8), reduced right ventricular longitudinal function, and elevated pulmonary artery systolic pressures (48-53 mm Hg) (**Figure 1**). There was also suspicion of mitral annulus calcification.

DIFFERENTIAL DIAGNOSIS

Differential diagnoses included ischemic heart disease and infiltrative cardiomyopathy.

INVESTIGATIONS AND MANAGEMENT

Cardiovascular magnetic resonance (CMR) was requested for further characterization. Multiple diffuse nodular signal-void lesions were identified in anatomical and functional images (cines), with a degree of pericardial infiltration. Cines showed marked concentric LV hypertrophy (maximum, 21-mm basal inferoseptum), as well as extensive mitral annulus calcification with fixed posterior mitral leaflet and restricted movement of the anterior leaflet that caused mild mitral stenosis and regurgitation. LV deformational analysis with feature tracking showed severely impaired LV longitudinal, radial, and circumferential strain (longitudinal strain, −1.86% [normal range, −11.1% to −27.1%]; circumferential strain, −5.12% [normal range, −12.7% to −24.1%]; and radial strain, 4.28% [normal range, 23.5% to 56.1%]).

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ABBREVIATIONS AND ACRONYMS

CMR = cardiac magnetic resonance

CT = computed tomography

LV = left ventricular

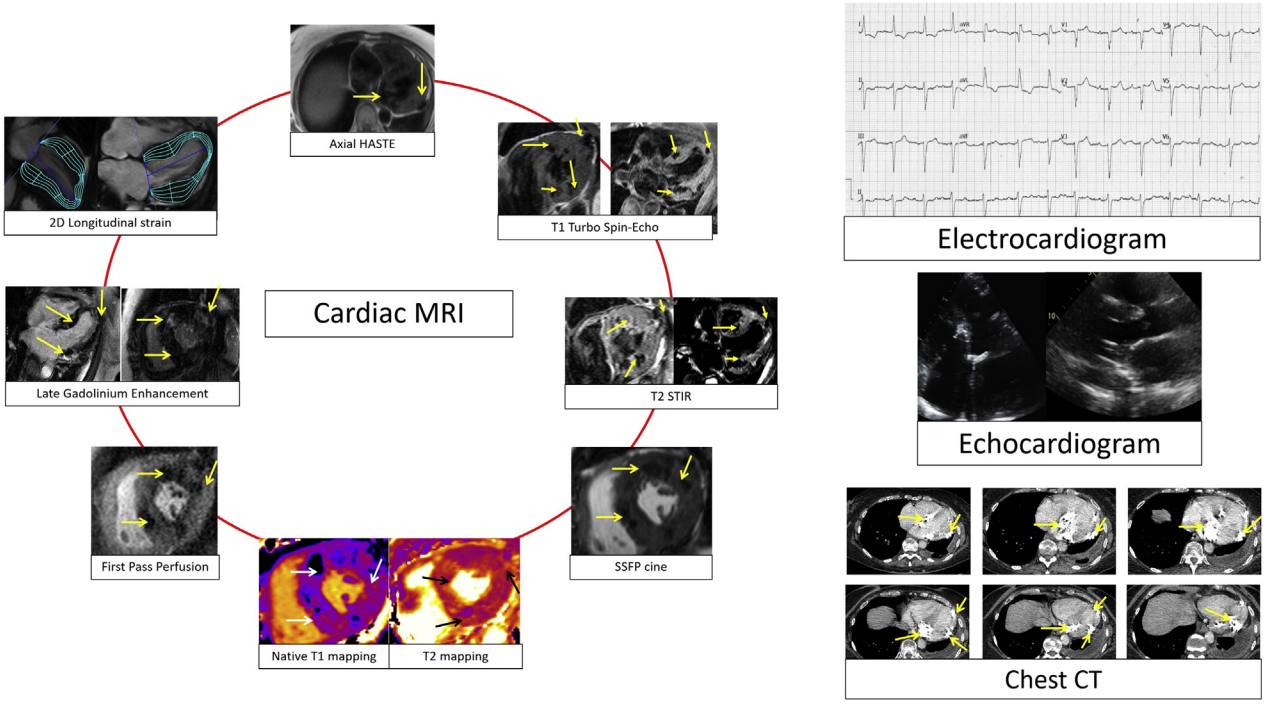
On tissue characterization, the lesions appeared to have low signal in all sequences: half-Fourier acquisition single-shot turbo spin echo imaging (HASTE), cine, T_1 turbo spin echo (TSE), fat-saturation sequences, T_2-weighted imaging, first-pass perfusion, and early and late gadolinium enhancement, demonstrating low or no vascularization, suggesting a calcific nature of these lesions. This was confirmed by low values on native T_1 and T_2 mapping (Figure 1). The findings were consistent with infiltrative restrictive cardiomyopathy with diffuse nodular calcifications. Chest radiography also showed myocardial and pericardial calcifications. The calcific nature of the lesions was further confirmed on subsequent chest computed tomography (CT) (Figure 1). Abdominal CT also showed bilateral renal calcifications.

Hyperparathyroidism and hypercalcemia were excluded. A more detailed retrospective clinical history investigation identified a diagnosis of lung tuberculosis 40 years earlier. The patient was referred to the respiratory team, but it was decided not to investigate because the current presentation was unlikely related to active tuberculosis. The then patient suddenly deteriorated neurologically, and she died before further assessment and treatment could be administered.

DISCUSSION

Infiltrative diffuse myocardial calcifications are rare findings; they can be associated with ventricular wall motion abnormalities, present as restrictive cardiomyopathy, and can be associated with sudden cardiac death (1).

FIGURE 1 Imaging of Myocardial Calcifications



(Left) Images of the left ventricle showing rounded lesions, which had low signal on all sequences (localizers: half-Fourier acquisition single-shot turbo spin echo imaging [HASTE], turbo spin echo T_1, native T_1 mapping, first-pass perfusion, and late gadolinium enhancement), thus confirming the calcific nature of the lesions **(arrows). (Top left)** Analysis of severely impaired longitudinal strain by a novel feature tracking technique. **(Top right)** Electrocardiogram of the patient showing left bundle branch block and first-degree atrioventricular block. **(Middle right)** Long-axis echocardiographic views showing severe mitral annulus calcification. **(Bottom right)** Chest computed tomography (CT) showing multiple hyperintense lesions involving the left ventricle and pericardium **(arrows),** thereby confirming the calcific nature of the lesions observed on cardiac magnetic resonance (CMR). 2D = 2-dimensional; STIR = short tau inversion recovery; SSFP = steady-state free precession.

The possible causes of myocardial calcifications are dystrophic or metastatic (2); some investigators have also described idiopathic causes (3). Dystrophic calcifications are the most common and result from direct tissue damage and cellular necrosis of different origins (eg, traumatic, ischemic, infectious). Metastatic calcifications are consequences of systemic disorders and abnormal calcium metabolism. The most common example is found in patients with renal failure, which was excluded in our patient.

Myocardial calcifications can be secondary to hyperparathyroidism and subsequent hypercalcemia, and accurate identification is crucial to guide treatment and reverse the course of the disease. A detailed clinical history and multimodality imaging are key in the assessment of myocardial calcifications. Cardiac CT is the most adequate technique to image myocardial calcifications; however, this case shows that CMR tissue characterization also identified the calcific nature of these lesions.

CONCLUSIONS

Infiltrative cardiomyopathy with diffuse nodular calcification is a rare entity. Cardiac calcification secondary to tuberculosis is also a rare finding. Multimodality imaging was crucial to determine the final diagnosis.

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REFERENCES

1. Henderson RR, Santiago LM, Lombardo M, Spring DA, Harrington AR. Metastatic myocardial calcification in chronic renal failure presenting as atrioventricular block. *N Engl J Med*. 1971;284(22):1252–1253.

2. Nance JW, Crane GM, Halushka MK, Fishman EK, Zimmerman SL. Myocardial calcifications: Pathophysiology, etiologies, differential diagnoses, and imaging findings. *J Cardiovasc Comput Tomogr*. 2015;9(1):58–67.

3. El Ghannudi S, Ohlmann P, Roy C, et al. Idiopathic myocardial calcification: insights from multimodality imaging. *Int J Cardiol*. 2016;221:1053–1055.

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