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MINI-FOCUS ISSUE: ELECTROPHYSIOLOGY

BEGINNER

CASE REPORT: CLINICAL CASE

Ventricular Tachycardia

A Rare Case of Myocardial Silicosis

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ABSTRACT

Chronic exposure to silica is a recognized health hazard. Manifestations of pulmonary and extrapulmonary silicosis are well described. Secondary pulmonary arterial hypertension and pericardial involvement are described, but myocardial involvement has not been reported. In this case of newly diagnosed pulmonary silicosis, ventricular tachycardia results are shown from pathological involvement of ventricular myocardium. (**Level of Difficulty: Beginner.**)
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PRESENTATION

A 45-year-old man presented with a 15-day history of persistent palpitations associated with mild cough. Clinical examination was unremarkable except for regular tachycardia at 130 beats/min and scattered crepitations over the chest. Electrocardiography (ECG) at presentation showed regular wide QRS complex tachycardia with right bundle branch block pattern in precordial lead V_1, right-superior QRS axis, atrioventricular dissociation, and a cycle length of 460 ms (Figure 1A).

LEARNING OBJECTIVES

- To perform a thorough work-up of patients presenting with unusual ventricular tachycardia and with long-term occupational exposure to silica to assess for possible myocardial involvement even in the absence of abnormalities on echocardiography and coronary angiography.
- To use myocardial tissue characterization with cardiac magnetic resonance imaging to identify the substrate for arrhythmias and, along with endomyocardial biopsy, help establish the diagnosis.

MEDICAL HISTORY

The man had worked in a stone-cutting factory for 20 years. There was no history of cardiac disease, near-syncope, or chest pain. Family history was negative for syncope, sudden death, and cardiac arrhythmia.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of regular wide QRS complex tachycardia included ventricular tachycardia (VT); supraventricular tachycardia with aberrant conduction or pre-existing bundle branch block; or pre-excited supraventricular tachycardia. Presence of atrioventricular dissociation, capture beat, atypical QRS morphology for right bundle branch block with Rs

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pattern, and tall R-wave in V_1 and aVR with a right-superior QRS axis supported a diagnosis of VT originating close to the inferoseptal wall near the left ventricular apex.

INVESTIGATIONS

Routine blood test results, including electrolyte and troponin levels, were within normal ranges. Echocardiography demonstrated normal biventricular function without hypertrophy or wall motion abnormality. Idiopathic left ventricular tachycardia (ILVT) was the presumed diagnosis, and intravenous verapamil was administered without effect. VT was resistant to lidocaine, amiodarone, electrical cardioversion, and overdrive pacing. Intravenous phenytoin (2 doses of 100 mg every 5 min), however, resulted in successful conversion to sinus rhythm.

ECG during sinus rhythm showed T-wave inversions across the precordium and inferior leads (Figure 1B). Chest radiographs showed a diffuse fibronodular pattern in the lung parenchyma bilaterally (Figure 1C). Coronary angiography revealed normal coronary arteries. Contrast-enhanced cardiac magnetic resonance (CMR) imaging showed normal cavity size and ventricular function with left ventricular basal and mid-cavity wall striae of late gadolinium enhancement (LGE) at the antero septal and inferoseptal junctional regions (Figure 1D). Contrast-enhanced computed tomographic chest scan demonstrated mediastinal lymphadenopathy with eggshell calcification, bilateral apical fibrocalcific masses, and diffuse small nodules throughout the lung (Figure 1E). Serum angiotensin-converting enzyme levels, rheumatoid factor, antinuclear antibodies, Mantoux test results, and QuantiFERON-TB Gold (QIAGEN, Germantown, Maryland) test results were within normal range. Transbronchial lung biopsy showed focal fibrosis with anthracotic pigment deposition and chronic inflammatory cell infiltration (Figure 2). Endomyocardial ventricular septum biopsy showed interstitial and perivascular fibrosis (Figure 3). No acid-fast bacilli were detected in the sputum,

ABBREVIATIONS AND ACRONYMS

CMR = cardiac magnetic resonance

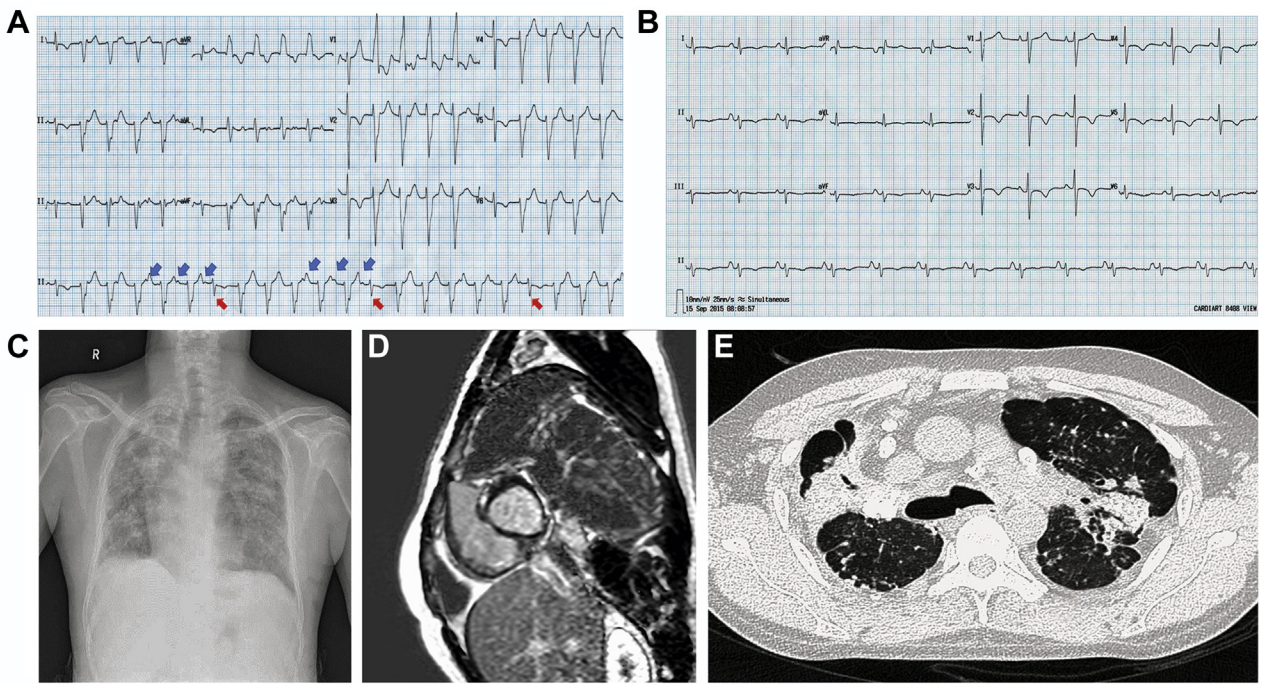
ECG = electrocardiogram

ILVT = idiopathic left ventricular tachycardia

LGE = late gadolinium enhancement

VT = ventricular tachycardia

FIGURE 1 Presenting Electrocardiogram and Chest Imaging



(A) Electrocardiogram shows ventricular tachycardia with right bundle branch block morphology. **Blue arrows** indicate P waves with AV dissociation; **red arrows** indicate captured beats with narrow QRS complexes. **(B)** Electrocardiogram in sinus rhythm shows left axis deviation with T-wave inversion in V_2 to V_6. **(C)** Chest radiographs demonstrate bilateral symmetrical opacities in the upper- and mid-zones with conglomerate masses of fibrosis with calcifications. **(D)** Cardiac magnetic resonance phase-sensitive inversion recovery imaging demonstrated basal and mid-cavity mid-wall striae on late gadolinium enhancement at the antero- and inferoseptal junctional regions. **(E)** Axial chest contrast-enhanced computed tomography demonstrates lesions in the upper and mid-zones, in the peripheral one-third of the lung, migrating toward the hilum with large, symmetric, bilateral, soft-tissue, mass-like opacities with irregular margins and calcifications, peripheral subpleural emphysematous bullae, and central fibrosis.

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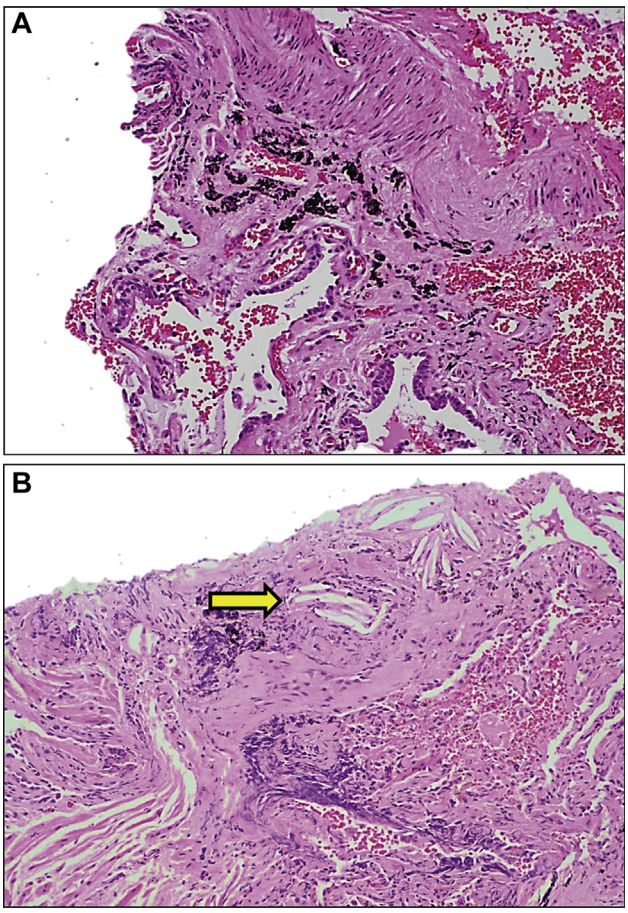
FIGURE 2 Lung Biopsy



(A, B) Photomicrographs of the lung biopsy sample (hematoxylin and eosin statin; ×20 original magnification) show moderate to marked fibrosis and hyalinization of the lung parenchyma. Cholesterol clefts **(B, arrow)**, few histiocytes, and dense, blackish anthracotic pigment depositions are noted in the fibrotic region without significant inflammatory cell infiltrate.

bronchoalveolar lavage, or pulmonary or myocardial biopsies. A diagnosis of pulmonary silicosis with myocardial involvement was made based on occupational and clinical history, ECG, and radiological and histological investigations, excluding ischemic heart disease, cardiomyopathy, tuberculosis, or sarcoidosis as the underlying pathology.

MANAGEMENT

The patient was advised to avoid silica exposure, undergo supportive management for pulmonary involvement, and take oral phenytoin (300 mg daily). The option of electrophysiological study and ablation of arrhythmia substrate was discussed. The patient chose medical therapy, with an ablation option if medicine failed.

DISCUSSION

In addition to pulmonary pathology, extrapulmonary involvement of the lymph nodes, bone marrow, kidneys, and brain has been described in cases of chronic silica exposure (1). The present patient had a 20-year history of exposure to silica dust with minor symptoms of pulmonary silicosis, presenting with palpitations and VT. ECG morphology suggested a VT exit in the inferoseptal left ventricle. No gross cardiac abnormality was detected on echocardiography, and the VT was initially considered to be ILVT. The clinical behavior of VT with unresponsiveness to verapamil, lidocaine, and amiodarone was unusual. Moreover, LGE at the antero- and inferoseptal junctional regions on CMR was suggestive of an underlying pathology not seen with ILVT. The presence of mediastinal lymph nodes with eggshell calcification and bilateral apical fibrocalcific masses and small nodules diffusely distributed throughout the lungs with anthracotic pigment deposition and chronic inflammatory cell infiltration on biopsy, with a history of silica exposure, was consistent with a diagnosis of pulmonary silicosis (1). A direct fibrogenic effect of silica particles in the lungs is described in silicosis, and an association with connective tissue disorders recognized (1), suggesting that extrapulmonary involvement may be mediated by an immunological response provoked by silicotic granulomatous nodular components. However, immunological markers, including antinuclear antibody and rheumatoid factor, were negative, and erythrocyte sedimentation rate was normal. Coexistent tuberculosis was ruled out by bacteriological, serological, and histological investigations. The distribution of scar and histology made the probability of sarcoidosis low. There was no evidence of myocarditis, and no apparent underlying disease was detected on echocardiography or coronary angiography. CMR findings with LGE in the anteroseptal and inferoseptal regions; myocardial biopsy with perivascular and interstitial scarring; and the VT morphology suggestive of exit close to the scar region, along with pulmonary and lymph node findings, suggested this to be a likely case of cardiac involvement with silicosis presenting with VT.

Pulmonary hypertension, constrictive pericarditis, and ischemic heart disease are reported cardiovascular complications of chronic silica exposure (2,3). In these authors' knowledge, there is no report of myocardial involvement in patients with silica exposure. In animal models, silica exposure is associated with focal myocardial fibrosis (4). Inhaled nanoparticles can be transported to heart and blood vessels through multiple mechanisms from the pulmonary

interstitium (5). The cardiac pathology is postulated to be caused by silica-induced mitochondrial dysfunction and apoptosis (6). Silicon dioxide can induce dysfunction in cytosolic calcium dynamics due to a reduction in sarcoplasmic reticulum Ca-ATPase activity. Abnormal diastolic calcium kinetics may play a role in triggering VT (7). The observation that the VT was resistant to amiodarone and overdrive pacing but responded to phenytoin suggested that the VT might have been of non-reentrant mechanism or micro-reentry around scarred myocardium. Phenytoin is known to alter the voltage-dependent sodium and calcium currents (8) and to terminate triggered activity-induced VT as seen with digoxin toxicity (9).

FOLLOW-UP

The patient has been asymptomatic from a cardiac standpoint for 3 years without any symptoms suggestive of VT or cardiac tachyarrhythmia on ambulatory ECG monitoring.

CONCLUSIONS

This paper reports the case of monomorphic VT due to patchy fibrosis within the ventricular myocardium in a patient with normal coronary arteries and echocardiogram and with pulmonary silicosis and lymph node involvement. In individuals with long-term silica exposure with new-onset cardiac episodes of arrhythmia, a high index of suspicion for myocardial involvement should be maintained and CMR imaging used early to define myocardial involvement that can be missed by standard echocardiography.

AUTHOR DISCLOSURES

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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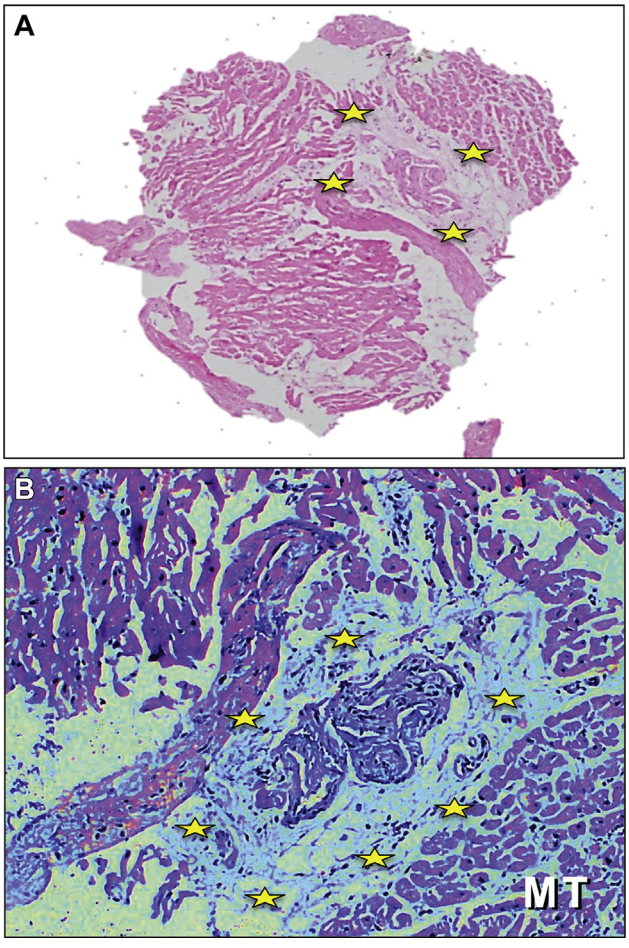
FIGURE 3 Endomyocardial Biopsy



(A) Endomyocardial biopsy showed increased perivascular fibrosis (hematoxylin and eosin stain; ×4 original magnification; **stars**). **(B)** Fibrosis **(stars)** is highlighted by Masson's trichrome (MT) stain.

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KEY WORDS LGE, silicosis, RBBB, ventricular tachycardia