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CASE REPORT

INTERMEDIATE

CLINICAL CASE

Unconventional Path to Healing

Diagnostic Value of CMR in a Patient With Incessant VT

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ABSTRACT

A 58-year-old male patient with incessant ventricular tachycardia was referred to cardiac magnetic resonance for scar assessment. He was found to have metastatic amelanotic melanoma of the heart. The cardiac magnetic resonance-based diagnosis of cardiac malignancy critically altered the subsequent clinical management. There was a marked response to immunotherapy as evidenced by follow-up imaging studies. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2019;1:638–42) © 2019 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

A 58-year-old male patient had a recent admission at an outside hospital for an out-of-hospital arrest and secondary prevention implantable cardioverter-defibrillator (ICD). His workup included a normal coronary angiogram. He was diagnosed with apical hypertrophic cardiomyopathy based on an echocardiogram. The patient was referred to our institution for the management of incessant ventricular tachycardia (VT) on amiodarone therapy with recurrent ICD shocks.

LEARNING OBJECTIVES

- Myocardial masses can result in serious complications including arrhythmias.
- An echocardiogram remains the first-line modality for the assessment of myocardial masses; the use of echo contrast is imperative in technically difficult studies.
- CMR has a great role in detecting and characterizing myocardial masses based on T1 and T2 tissue properties.

PAST MEDICAL HISTORY

The patient reported being in good health before this presentation.

DIFFERENTIAL DIAGNOSIS

Cardiomyopathy (hypertrophic, nonischemic, inflammatory, and ventricular dysplasia), infiltrative process resulting in scar-mediated VT, channelopathy, metabolic abnormalities, and myocardial mass were considered in the differential diagnosis at initial presentation.

INVESTIGATIONS

A cardiac magnetic resonance (CMR) scan was obtained for scar assessment in preparation for VT ablation. Images were acquired using a 1.5-T magnetic scanner (Avanto, Siemens, Erlangen, Germany). Steady-state free precession sequence cine images revealed 2 large intracardiac masses (**Figure 1A**) with normal biventricular function but focal hypokinesis

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Informed consent was obtained for this case.

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of the anterior and apical segments at the location of the masses in the left ventricle (Videos 1 and 2). Both masses were isointense to myocardium on T1-weighted imaging (Figure 1B), hyperintense to myocardium on T2-weighted imaging (Figure 1C), and heterogeneously hypoperfused in relation to the myocardium (Figure 2A). Late gadolinium enhancement imaging demonstrated central low signal intensities concerning for central necrosis (Figure 1D). Later that day, his ECHO (Figure 3) at our institution was also consistent with myocardial masses.

MANAGEMENT

The masses were biopsied via a transseptal approach under 3-dimensional intracardiac ultrasound guidance. The biopsies were interpreted as metastatic amelanotic melanoma supported by immunohistochemical studies (Figure 4). During his hospitalization, the patient later recalled having a remote history of wide excisions of a localized melanoma of the upper back. A whole-body positron emission tomography scan revealed multiple focal cardiac metastases with probable pericardial involvement (Figure 5A) along with retroperitoneal metastasis, suggesting peritoneal carcinomatosis. Oncology was consulted, and the patient was initiated on pembrolizumab. For his VT, his antiarrhythmic therapy was escalated to intravenous amiodarone and lidocaine and then transitioned to oral amiodarone and mexiletine. He remained arrhythmia free for a week before discharge. He was maintained on bimonthly pembrolizumab infusions as an outpatient and tolerated therapy fairly well.

FOLLOW-UP

The patient was followed with serial surveillance CMR with evidence of tumor regression within 2 months on therapy. Five months post-follow-up, the CMR scan indicated the stigmata of treated tumor with regions of late gadolinium enhancement and residual fibrotic changes (Figures 1E to 1G and 1I). A follow-up whole-body positron emission tomography scan was also negative at 5 months (Figure 5B).

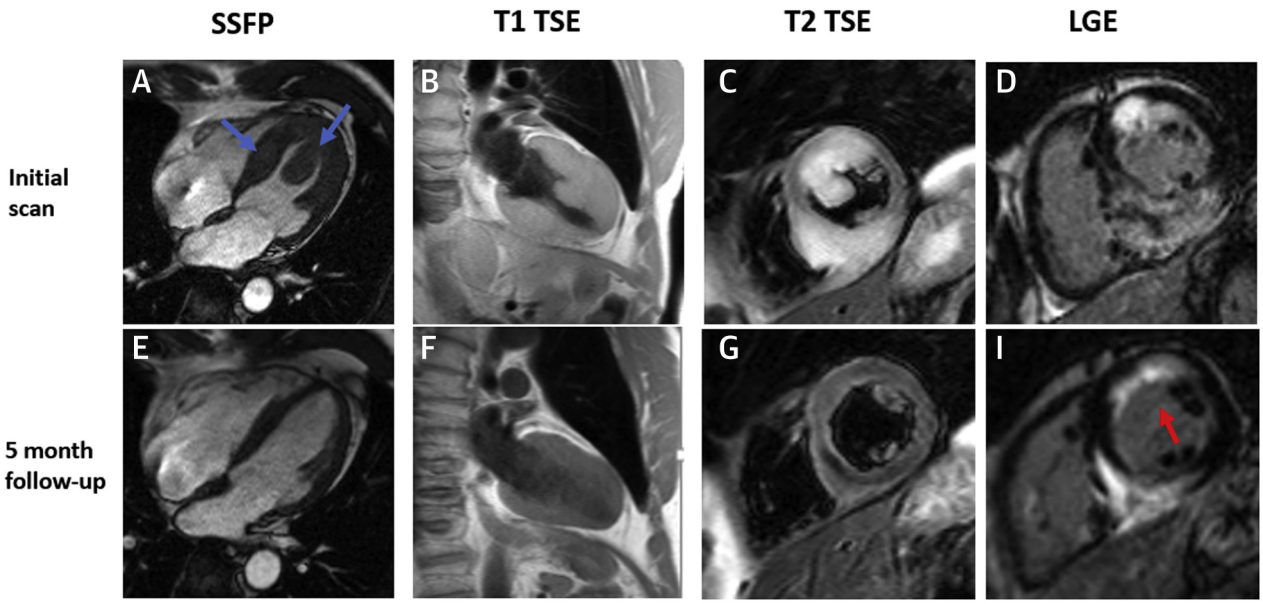
ABBREVIATIONS AND ACRONYMS

CMR = cardiac magnetic resonance

ICD = implantable cardioverter-defibrillator

VT = ventricular tachycardia

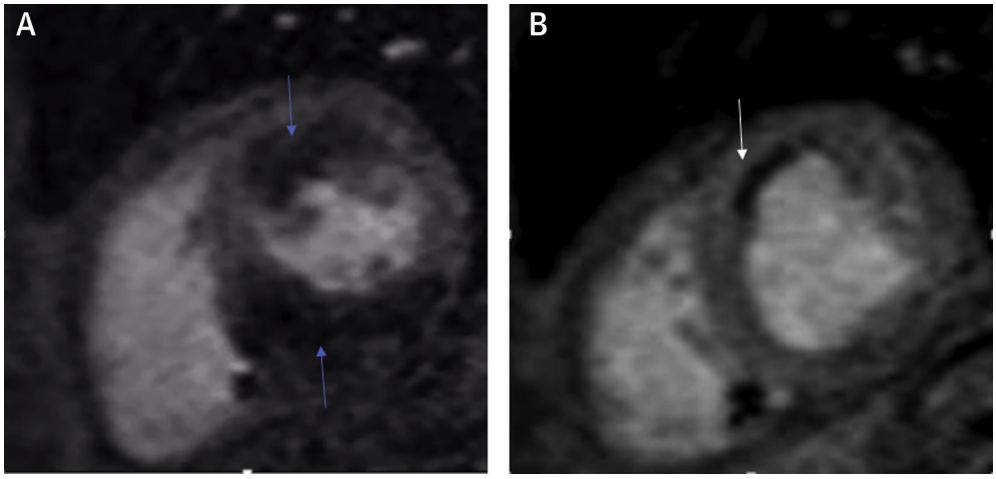
FIGURE 1 Tissue Characteristics of the Myocardial Masses on Cardiac Magnetic Resonance



(Top) The initial scan at the time of diagnosis of cardiac masses and **(bottom)** the 5-month follow-up scan on pembrolizumab. **(A)** The cardiac tumor at presentation with 2 masses **(blue arrows)** on steady-state free precession (SSFP) cine images. **(B)** The masses are isointense to myocardium on T1-weighted imaging and **(C)** hyperintense to myocardium on T2-weighted imaging. **(D)** Late gadolinium enhancement (LGE) imaging demonstrated central low signal intensities concerning for central necrosis. **(E to G)** Regression of intracardiac masses, and **(I)** on LGE imaging, there is prominent scarring consistent with replacement fibrosis **(red arrow)**. See Videos 1 and 2.

JACC: CASE REPORTS, VOL. 1, NO. 4, 2019
DECEMBER 2019:638–42

FIGURE 2 Cardiac Magnetic Resonance First Pass Perfusion Imaging



(A) Cardiac magnetic resonance perfusion imaging on the initial scan demonstrates the masses are hypoperfused and dark compared with the surrounding myocardium **(blue arrows)** compared with myocardium with regional contrast uptake concerning for malignancy. **(B)** The 5-month follow-up perfusion scan with normalization of perfusion in the inferior and inferoseptum. The anterior/anteroseptum is hypoperfused **(white arrow)** in a similar distribution to the late gadolinium enhancement pattern, which is consistent with a scar.

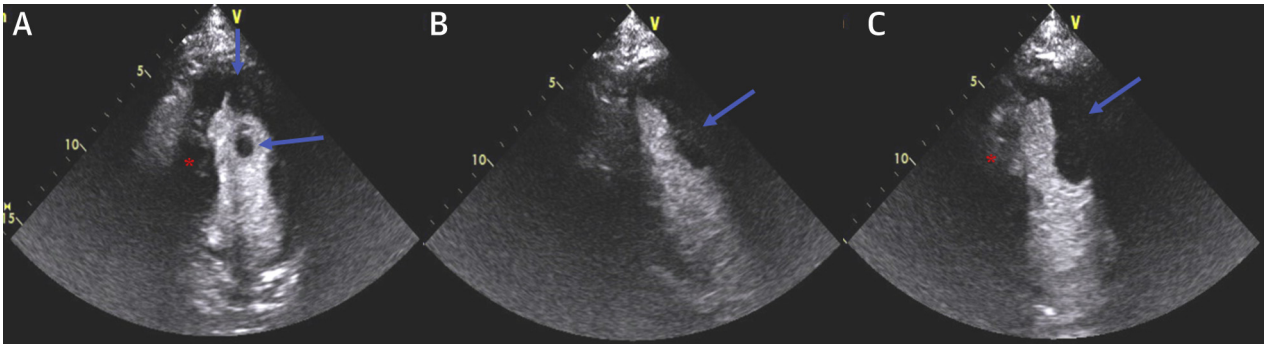
DISCUSSION

A comprehensive literature review identified only 3 published case reports of amelanotic melanoma metastasizing to the heart: 2 cases with evidence of right atrial metastasis and the third with metastasis involving the atrioventricular groove (1-3). To our knowledge, this is the first reported case of amelanotic melanoma with left ventricular involvement. A combination of immunotherapy and chemotherapy for right-sided metastatic melanoma has been reported with a mixed response to therapy (4-6). This is the first published case for the use of pembrolizumab as monotherapy in metastatic amelanotic melanoma to the heart with a dramatic response to treatment.

CONCLUSIONS

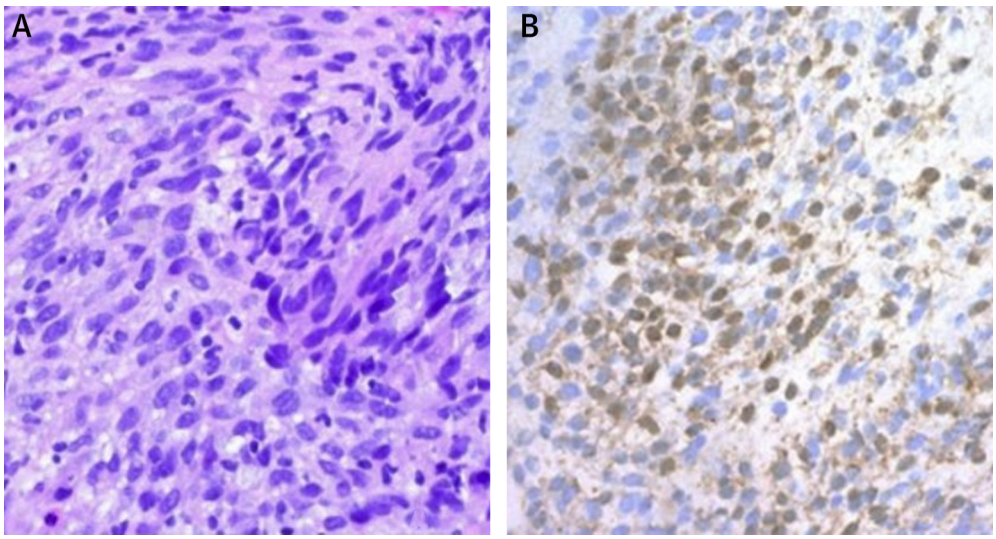
The CMR-based diagnosis of cardiac malignancy critically altered the subsequent clinical management.

FIGURE 3 Myocardial Masses on Contrast Echocardiogram



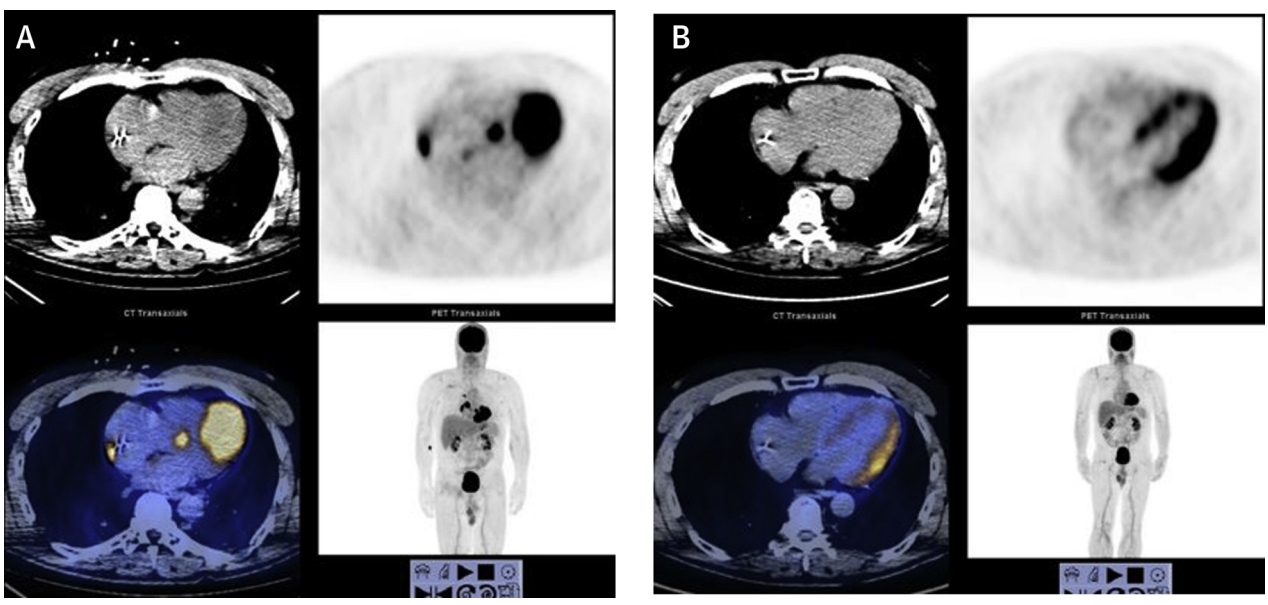
A contrast echocardiogram at the time of diagnosis demonstrated heterogeneous left ventricular masses involving the apex and anterior walls **(blue arrows)**. The inferoseptum and inferior walls with heterogeneous thickening all concerning for malignant invasion of the heart **(red asterisks)**. **(A)** Apical 4-chamber view, **(B)** apical 3-chamber, and **(C)** apical 2-chamber views.

FIGURE 4 Histopathology of the Endomyocardial Biopsy Specimen



Endomyocardial biopsy of the cardiac tumor. **(A)** Hematoxylin-eosin showing spindled malignant cells without associated pigment. **(B)** Positive immunohistochemical staining for S-100 in the tumor supportive of diagnosis of amelanotic melanoma.

FIGURE 5 Positron Emission Tomography/Computed Tomography Images



(A) Positron emission tomography computed tomography images at presentation demonstrated the largest lesion in the left ventricular apex with probable pericardial involvement. **(B)** A follow-up scan with resolution and residual physiological uptake.

JACC: CASE REPORTS, VOL. 1, NO. 4, 2019
DECEMBER 2019:638–42

The apical tumor was initially misinterpreted as apical hypertrophic cardiomyopathy, and a secondary prevention ICD was implanted. A magnetic resonance nonconditional ICD was scanned using our institutional protocol involving pre- and post-scan device programming and continuous monitoring by a cardiologist. The CMR-based diagnosis led to the timely initiation of the immunotherapy with pembrolizumab. Follow-up scans are being used to monitor the patient's clinical course and response to therapy.

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KEY WORDS cardiac magnetic resonance, immunotherapy, metastatic cancer, ventricular tachycardia

APPENDIX For supplemental videos, please see the online version of this paper.