

Subacute Myocarditis with Malignant Progression in a Young Patient: Contribution of Cardiac Magnetic Resonance

Miocardite Subaguda de Evolução Maligna em Jovem: a Contribuição da Ressonância Magnética Cardíaca

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Introduction

Myocarditis can have a varied clinical presentation ranging from chest pain and mild dyspnea to cardiogenic shock and sudden death. It is defined as myocardial inflammation with myocyte necrosis or degeneration without ischemia and may result from infectious and non-infectious causes, with viral myocarditis being the most prevalent etiology.^{1,2} Myocarditis most commonly affects young male adults, being detected in up to 12% of patients aged under 40 years who had sudden death.³ Its diagnosis is fundamentally based on clinical suspicion due to recent viral infection associated with a current myocardial lesion, the absence of previous heart disease, and heart failure (HF) of no other apparent cause. Cardiac magnetic resonance (CMR) imaging findings can currently help confirm the diagnosis.⁴

The low availability of endomyocardial biopsy, the gold standard diagnostic method, brings major challenges to the management of myocarditis. On the other hand, the increased use of CMR provides important diagnostic and prognostic data. This report describes the clinical issues and CMR contribution in a case of myocarditis in a young patient.

Case report

A 21-year-old man without comorbidities had been complaining of chest pain and a dry cough, progressive dyspnea, and abdominal and lower limb edema for four weeks without fever. He was admitted to the emergency room on June 4, 2020 with a respiratory rate of 29 bpm, respiratory distress, and use of accessory muscles. He was pale (+/4+) and sweating. We noted the presence of B3 on cardiac auscultation, jugular turgency, and hepatojugular reflux, in addition to lower limb edema (2+/4+), heart rate of 141 bpm, blood pressure of 133/81 mmHg, and slow capillary refill. Diminished vesicular sounds were noted on the left with bilateral crackling rales; his oxygen saturation was 96% on room air.

Keywords

Myocarditis; Magnetic resonance imaging; Death, sudden.

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A COVID-19 polymerase chain reaction test was negative, as was the screening for autoimmune and Chagas disease. Laboratory tests showed a quantitative troponin T < 0.1 ng/L, leukocytes at 23,400/mm³ with 11% band neutrophils, and C-reactive protein at 103.2 mg/L. Findings of electrocardiography and chest X-ray performed on hospital admission were compatible with HF (Figure 1). Echocardiography showed eccentric hypertrophy and significant left ventricular systolic dysfunction due to diffuse hypokinesia and 24% left ventricular ejection fraction in addition to moderate right ventricular systolic dysfunction, global enlargement of the cardiac chambers, and absence of significant valvular dysfunction.

The patient progressed with low cardiac output in the first 24 hours and developed an acute kidney injury. The initial clinical management was difficult, with refractoriness for several days, the need for high doses of inotropes (dobutamine), and intolerance to drugs that reduce mortality in HF. After a prolonged hospitalization in the intensive care unit (ICU), clinical stabilization was achieved with the introduction of beta-blockers, sacubitril/valsartan, and spironolactone. CMR was possible after 44 days of hospitalization (Figure 2) and showed a 19% left ventricular ejection fraction, a 25% right ventricular ejection fraction, normal-sized atria, mesocardial fibrosis suggestive of non-ischemic etiology in 4% of the left ventricle, and the presence of linear delayed myocardial enhancement in the anteroseptal (basal and medium), inferoseptal (basal and medium), and inferolateral (medium) segments in addition to no signs of myocardial edema.

On July 22, 2020, the patient was discharged from the hospital with optimized pharmacological treatment and early outpatient scheduling. Before leaving the hospital, the patient experienced cardiopulmonary arrest (CPA) in ventricular fibrillation and received cardiopulmonary resuscitation maneuvers in the ward for 25 minutes. After spontaneous circulation returned, he was transferred to the ICU with mechanical ventilation assistance and noradrenaline and dobutamine intravenous injections. The patient presented refractory cardiogenic shock and required an intra-aortic balloon pump (IABP) implant. After two hours, he became refractory to the IABP, progressing with Interagency Registry for Mechanically Assisted Circulatory Support profile 2 classification. The authors opted for circulatory support with an INCOR biventricular assist device (Berlin Heart GmbH, Berlin, Germany) via a median sternotomy. After three hours, the shock worsened again associated with massive bleeding from the sternotomy due to a coagulation disorder. The patient progressed to CPA in asystole and died ten hours after the first CPA.

Case Report

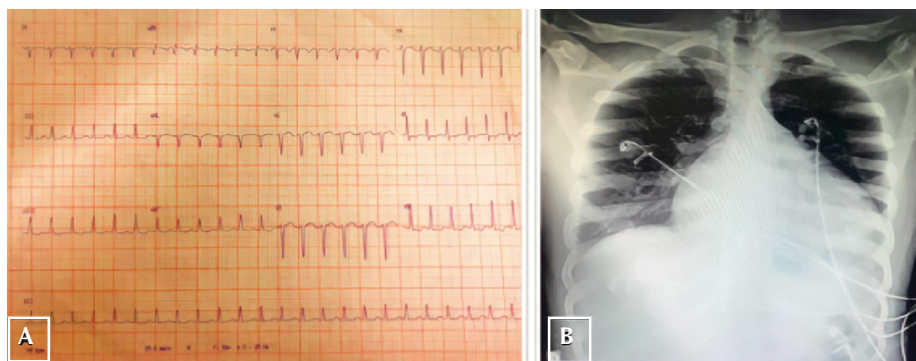


Figure 1 – (A) Electrocardiogram showing sinus rhythm, heart rate of 150 bpm, left atrial overload, absence of R wave progression in the anterior wall, and a nonspecific ventricular repolarization change. (B) Chest X-ray showing significantly increased cardiac area at the expense of the left and right chambers. Consolidation in the right lung base with mild bilateral pleural effusion.

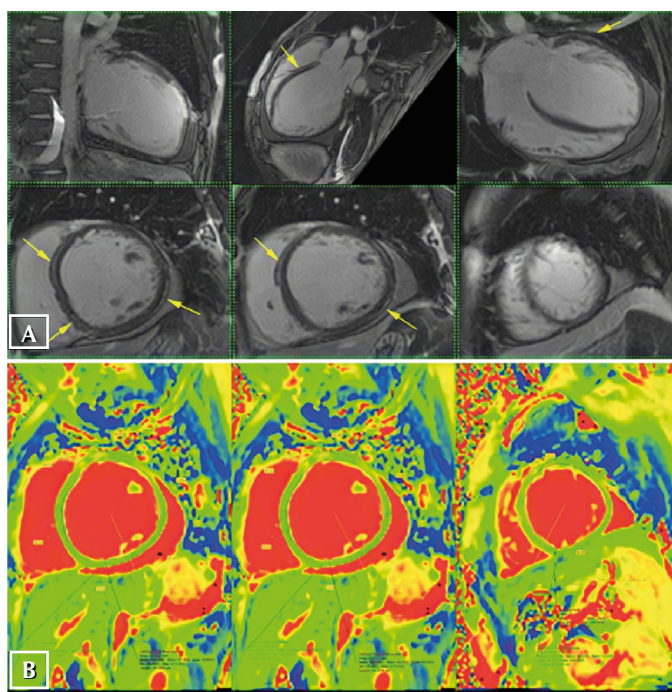


Figure 2 – Cardiac magnetic resonance. (A) Morphofunctional evaluation showing significant hypokinesia of the left and right ventricles. Delayed myocardial enhancement with a pattern of mesocardial involvement (yellow arrows) in the anteroseptal (basal and medium), inferoseptal (basal and medium) and inferolateral (medium) segments. (B) Parametric maps (T2) showing myocardial fibrosis with no signs suggestive of myocardial edema, evidencing the subacute phase of myocarditis.

Discussion

Myocarditis is often a mild and self-limited consequence of a systemic cardiotropic viral infection without clinically relevant residual damage. However, some patients present a rapidly changing clinical condition progressing with reduced cardiac function, hemodynamic compromise, severe arrhythmias, and sudden death.¹ The therapeutic approach is generally similar to the one used for a patient with decompensated HF.⁵ However, some specific situations may require treatment aimed at the cause of myocardial injury, such as infectious agents, systemic

autoimmune diseases, human immunodeficiency virus, hypersensitivity to drugs, and Chagas disease.⁶

A myocardial biopsy-guided approach is recommended for patients with a clinical presentation of acute HF with shock and adverse progression.² A biopsy was suggested in the present case, but logistical difficulties prevented its use. On the other hand, CMR can quantify myocarditis lesions, identifying myocardial inflammatory injury in the acute and subacute phases and scar lesions in the chronic phase.^{2,4,7} In addition to confirming the diagnosis, the risk of sudden

death can be stratified based on the presence or absence of delayed myocardial enhancement with gadolinium. Thus, CMR was used to investigate and guide the conduct, with the understanding that a myocardial biopsy could greatly contribute to cases of acute myocarditis, guiding a specific therapy based on its result.

The delayed enhancement pattern of myocarditis differs from the pattern of myocardial infarction, as it is often mesocardial, sparing the endocardium and possibly having epicardial and transmural distributions. Furthermore, the enhancement regions do not respect the coronary territory; rather, they are multifocal and heterogeneous. Despite the absence of increased serum troponin, which could be related to an unfavorable progression,² the presence of delayed myocardial enhancement is associated with a worse long-term prognosis⁴ that is compatible with the outcome of the reported case. The severe and unexpected ventricular arrhythmia at discharge and the unfavorable subsequent clinical progression was due to the minimal myocardial reserve.

As for the temporal progression, CMR T2 mapping assesses myocardial edema due to an inflammatory process that is usually reversible.^{8,9} However, this case did not present T2 images compatible with myocardial edema but rather with fibrosis, determining a subacute phase. Left ventricular dilation is also compatible with subacute myocarditis since the onset of negative remodeling occurs days or weeks after the initial myocardial injury.⁹ The acute phase, on the other hand, usually presents with considerably increased necrosis biomarkers and a normal cardiac area.

Cases of autoimmune, eosinophilic by hypersensitivity, sarcoidosis, or giant cell myocarditis would benefit from corticosteroid therapy but require biopsy and negative viral

testing. For these reasons, the use of corticosteroids is a frequent question in the management of myocarditis since patients with subacute presentations may present clinical worsening and not benefit from this intervention. The patient's clinical-laboratory scenario corroborated the CMR result, and the determination of the progression time aided with the decision-making.

Our patient showed a severe subacute stage of myocarditis that progressed to reversed sudden death and, subsequently, heart pump failure and multiple organ dysfunction. A common major challenge in refractory cases like this is choosing specific therapies, such as corticosteroid administration. In this situation, the definition of the time of myocardial aggression (acute or subacute) is essential. Clinical instability making CMR impossible and difficulties performing endomyocardial biopsy complicate the final diagnosis and management by the medical team. However, when available, CMR findings greatly contribute to decision-making, helping determine the temporal progression of myocarditis.

Authors' contribution

Research conception and design: Barbosa RR, Moreira GR, Curcio GM, Calil OA, Serpa RG, and Barbosa LFM; data collection: Moreira GR and Curcio GM; manuscript writing: Barbosa RR, Moreira GR, and Curcio GM; critical review of the manuscript for important intellectual content: Barbosa RR, Calil OA, Serpa RG, and Barbosa LFM.

Conflict of interest

The authors have declared that they have no conflict of interest.

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