

Fulminant myocarditis

Miocardite fulminante

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ABSTRACT

The myocarditis constitutes one of the most challenging diagnoses in cardiology because it is rarely recognized clinically. In addition, there is no gold standard exam for the diagnosis and current treatment remains controversial. The objective of this case report is to describe the importance of early diagnosis of acute fulminant myocarditis in the evaluation of patients with chest pain and clinical history that is suggestive that after appropriate treatment, presented favorable evolution.

Key words: Myocarditis; Magnetic Resonance Imaging; Heart Failure.

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RESUMO

A miocardite constitui-se em um dos diagnósticos mais desafiadores em Cardiologia, pois raramente é reconhecida clinicamente. Além disso, não existe um exame que seja padrão-ouro para o diagnóstico e o tratamento atual permanece controverso. O objetivo deste relato de caso é descrever a importância do diagnóstico precoce de miocardite aguda fulminante, na avaliação de paciente com dor torácica e história clínica sugestiva que, após tratamento adequado, apresentou evolução favorável.

Palavras-chave: Miocardite; Imagem por Ressonância Magnética; Insuficiência Cardíaca.

INTRODUCTION

Myocarditis or inflammatory cardiomyopathy has been recognized as one of the main determinants of dilated cardiomyopathy.¹ It is characterized by an inflammatory response of the myocardium, often as a result of an infectious primary assault on another site.² The inflammatory process can affect other heart structures, most commonly the pericardial sac (pericarditis). The involvement of this structure causes typical electrocardiographic alterations such as elevation of the ST-T segment and complaint of precordial pain, findings that are very similar to those in the acute coronary syndrome clinical framework, which excludes essential obstructive coronary disease in some cases.³

The most frequent aggressing agent is an infectious one, through a direct mechanism of the virus against cardiac myocytes and the myocardial supporting tissue, or through a post-infection viral autoimmune reaction.¹ Myocarditis may also be secondary to assaults by the immune system such as in peripartum myocarditis, radiotherapy, chemotherapy, and drug hypersensitivity. Among the infectious agents, the most common is viruses, especially enterovirus. Among these, the Coxsackie

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virus type B are most often responsible for myocardial inflammation although new studies found a wider spectrum of responsible viral genomes indicating a change from enteroviruses and adenoviruses to B19 parvovirus and herpes virus 6.⁴

Myocarditis is classified as fulminant, acute, and chronic (active and persistent). The clinical presentation is variable and may be asymptomatic or present frequent arrhythmias, sudden death, symptomatic or asymptomatic ventricular dysfunction, embolic and fulminant form events, which usually happens in young patients with no previous history of coronary heart disease. When fulminant, it begins within days after a viral infection well identified and is characterized by severe left ventricular dysfunction and often cardiogenic shock, with biopsies revealing serious inflammatory infiltrates and myocyte necrosis. Despite the severity of the framework, studies show that those patients with fulminant myocarditis are those who have more chance of full recovery^{5,6} and no morbidity and mortality related to the heart after the initial presentation.⁷ Surviving patients display histological resolution of their myocarditis and heart failure (HF) in the follow-up period.

Myocardial alterations can be demonstrated through ECG, which typically shows nonspecific changes in the ST segment and T wave. Thoracic radiology can show findings of heart failure in a more advanced form. Alterations in biomarkers of myonecrosis, mainly troponin I, are not commonly found only when the patient is in an acute phase, manifesting rapid myocardial deterioration. Another laboratory finding can be the presence of autoantibodies.⁷ Non-invasive examinations include scintillography with gallium, which presents only 36% sensitivity, but specificity of 98%; echocardiogram, which can help identify patients with fulminant myocarditis at the time of presentation; and cardiac magnetic resonance imaging (CMRI), which has between 100 and 90% specificity and 100% sensitivity.⁷

The diagnosis of viral inflammation is performed by an endomyocardial biopsy in the right ventricular based on the immunohistochemistry analysis with evidence of more than 14 lymphocytes and macrophages per mm² associated with positive HLADR.¹ If fulminant myocarditis is suspected, the endomyocardial biopsy should be performed in patients with newly established and not explained heart failure, lasting less than two weeks, in association with left ventricle of normal size or enlarged and with hemodynamic

involvement. In addition, the endomyocardial biopsy must be performed in patients with newly established and not explained heart failure, with duration of two weeks to three months, in association with dilated left ventricle and new ventricular arrhythmias or atrioventricular block (BAV) of second degree Mobitz type II or advanced BAV; and in patients not responsive to the usual treatment in one to two weeks. For patients who do not fit in these frameworks, the endomyocardial biopsy was not well established.⁸

In the view of the diversity of clinical presentations and the low specificity of laboratory markers, the diagnosis is based fundamentally on the high degree of clinical suspicion and, more recently, in the confirmation through CMRI findings. The degree of clinical suspicion increases with history of prior and viral disease and in the absence of preexisting heart disease associated or not to the sudden onset of arrhythmias or cardiac conduction disorder, as well as in the event of an increase in the cardiac area or symptoms of congestive heart failure without apparent cause.⁶

The myocarditis treatment still remains controversial. All patients with myocarditis should limit their physical activities and appropriately treated for HF.⁸ When indicated, arrhythmias must be treated besides preventing vascular spasm.⁷ These are supporting measures to reduce acute symptoms in the organism, with its immune system, eliminating the causal agent of infection. However, many current studies show a new therapy strand that works by minimizing the causal factors. This treatment aims to improve cardiac function and increase survival. The therapy is represented by immunosuppression (reduced inflammatory activity) and immune modulation (removal of immune complexes, autoantibodies, cytokines, and viral infection).¹

Several studies were executed in an attempt to show the benefits of the immunosuppressive therapy. In relation to adult patients with newly diagnosed cardiomyopathy and suspected myocarditis, the use of immunosuppressive therapy does not appear to be beneficial as its clinical course is usually of spontaneous recovery.⁷ The treatment of fulminant myocarditis may require short-term supporting measures such as intra-aortic pumping balloon or left ventricular assistance devices. In addition, these studies do not allow the indication of the routine way of using immunosuppressive or immunomodulatory therapy in dilated cardiomyopathies without clear etiology.¹ Therefore, the recommendation of immunosuppressive therapy requires histological confirmation of myocarditis and

is only indicated in those patients who have not improved with treatment for heart failure.⁷

CASE REPORT

J.R.S.N., male, 37 years old, was admitted in the ICU-B from the SEMPER Hospital in 11/5/2008, complaining of chest pain. He reported typical precordial pain with evident clinical and celebratory dyspnea at rest. He reported that the pain began two days ago, with periods of calm and exacerbation. The previous history just revealed abdominal pain associated with gastroenteritis in the previous week.

In the clinical examination he was ruddy, hydrated, cooperative, TA = 37.2 °C *without deficits and without systemic stasis*. PA = 137/74 mmHg. FC = 100 bpm. In the cardiac auscultation, rhythmic beats, B1 and B3 hypophonesis without outline. In the respiratory auscultation, decreased vesicular murmur, with bilateral basal crepitations. Free abdomen. No edema.

The complementary examinations revealed: chest x-ray with increased cardiac area and signs of pulmonary flow cephalization. Admission ECG: regular sinus rhythm, ST supra in D2, D3, avF, V4 to V8 (Figure 1). Troponin = 4.03; CKMB = 70; CPKT = 781.

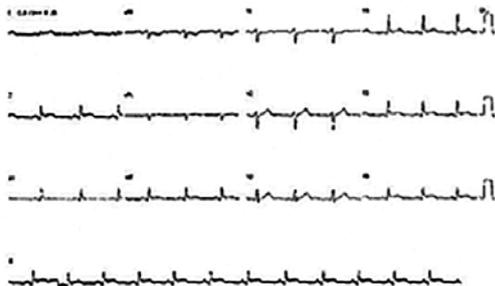


Figure 1 - Admission ECG performed on 11/15/08.

The patient's clinical association with important precordial pain, electrocardiographic alterations, and evidence of myocardial injury led to the hypothesis that it was a case of acute myocardial infarction. The catheterization conducted revealed normal coronary. Acute fulminant myocarditis was suspected because he was a young patient without comorbidities, not a user of illicit drugs, or with infectious prior history and primary ECG alterations.

The acute fulminant myocarditis (AFM) should be considered in patients with rapidly progressive cardiomyopathy and heart failure (HF), ventricular arrhythmias, or ECG alterations that indicate AFM but with normal coronary.⁴ In addition to ECG alterations and biochemical examination compatible with myocarditis, the patient evolved into cardiogenic shock on day 11/05 when he received dobutamine. Other complementary examinations for diagnostic confirmation were then requested including those that map inflammatory disease: PCR (daily values shown in the following graph): (Figure 2). Echocardiography showed left ventricular ejection fraction = 49%, increase in the atrium and left ventricle and diffuse hypokinesia indicating possible acute myocarditis (Figure 3). The clinical alterations associated with changes in the x-ray and ECHO allowed the diagnosis of left ventricular failure.

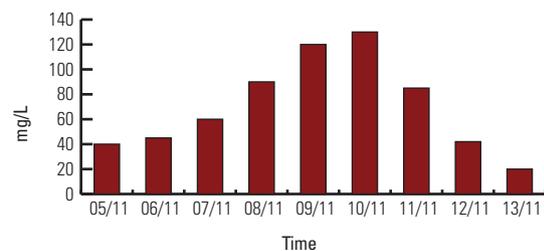


Figure 2 - Evolution of the patient based on PCR values.

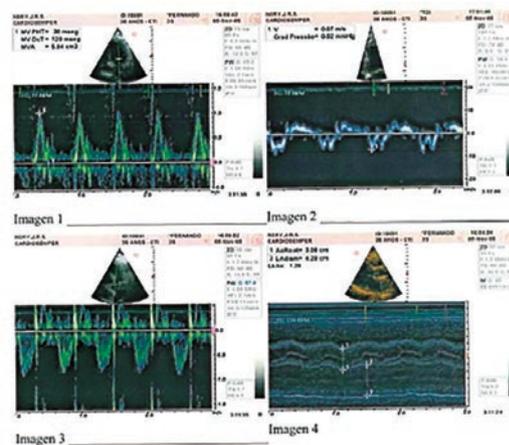


Figure 3 - Transthoracic Echocardiogram carried out on 11/5/08.

As shown in Figure 2 and x-ray from day 11/09 (Figure 4), representing the acute phase of the disease, the patient evolved with progressive worsening with severe left ventricular failure requiring non-invasive ventilation.



Figure 4 - Chest x-ray performed on 4/9/08 demonstrating severe pulmonary congestion.

The CMRI was performed on 11/11 and sealed the diagnosis of acute fulminant myocarditis. The examination revealed FE: 29.4%, increased left ventricular dimensions associated with important global hypokinesia as well as the presence of multiple areas of late enhancement; such findings are compatible with the inflammatory process (Figure 5).

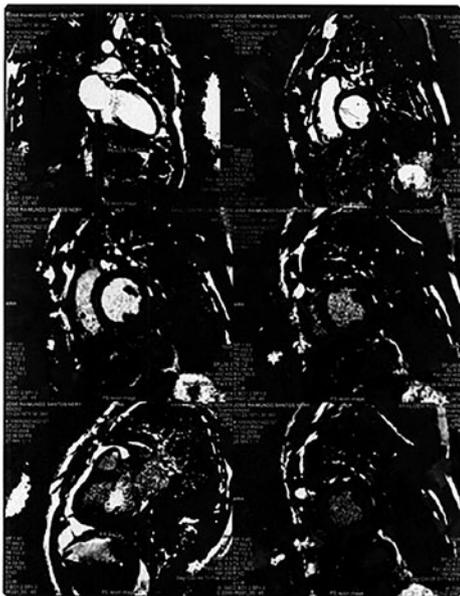


Figure 5 - CMRI held on 11/11/08 confirming the diagnosis of fulminant myocarditis.

The classic HF/left ventricular failure was the clinical treatment: IECA, beta-blockers, spironolactone, furosemide, and dobutamine (due to the cardiogenic shock), enoxaparin, VNI and water restriction. A framework progressive improvement was observed as shown in the radiography from day 11/13 (Figure 6). The patient was discharged from the ICU on 11/13/2008.

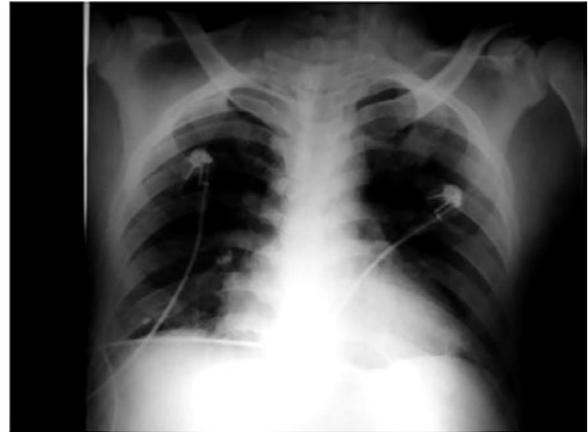


Figure 6 - Chest radiograph performed on 11/13/08 demonstrating intense pulmonary congestion improvement.

DISCUSSION

The diagnostic hypothesis initially suggested in this case was that of an acute coronary syndrome due to the clinical characteristics and observation of electrocardiographic and biochemical alterations consistent with AFM. However, the disease progression and a cardiac catheterization with normal report ruled out the possibility of an acute coronary episode. Thus, acute fulminant myocarditis was suspected, whose clinical diagnosis is made most often in young patients with a previous history of recent viral infection. Diffuse ST-T segment alterations and increase of myocardial injury markers can be observed. However, this form of clinical presentation is not a constant and often the diagnosis of myocarditis is not confirmed. Auxiliary methods in the diagnosis, such as laboratory tests and ECG are not specific and others, such as serology, scintillography, and endomyocardial biopsy, are not carried out in most cases, due to their complexity, need for specific contrasts, and an appropriate window of time for their execution. Thus, Cardiac MRI (CMRI) was requested, a method that can assess myocardial function, flows and fluids, and characterize areas of necrosis and fibrosis. The necrosis patterns found in the executed CMRI were compatible with fulminant myocarditis confirming the diagnosis. The patient evolved with rapidly progressive heart failure; the instituted treatment was the classic for HF and left ventricular failure. The treatment of myocarditis is controversial, no specific treatment have been established. However, studies show that all patients should receive random therapy for HF and that the immunosuppressive therapy does not lead to improved survival or car-

diac function in patients with fulminant myocarditis. Thus, the immunosuppressant treatment was not established for this patient and yet he evolved with progressive improvement after the introduction of treatment for CHF.

CONCLUSIONS

The early diagnosis of acute fulminant myocarditis, often difficult to carry out, was crucial for the positive progression of this patient. In this respect, the CMRI was essential enabling the precise assessment of the injured area. With its recent introduction in the clinical practice, the CMRI has been established as a mainstay in the diagnosis of AM, showing 100% sensitivity and 90% specificity. In this case, the patient evolved favorably after the implementation of the appropriate treatment for CHF, without the need for immunosuppressive therapy, which does not seem to be beneficial for adult patients with newly diagnosed cardiomyopathy and assumed fulminant myocarditis.

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