Introduction

Rheumatic fever (RF) is a prevalent condition in developing countries, accounting for a cardiac mortality rate of 7.9% in Brazil (DATASUS). Cases of rheumatic fever are often underdiagnosed and recognized only after permanent valve damage due to previous carditis. Secondary prevention of RF aims to limit the development and/or progression of valvular lesions and their complications. However, even on therapy with benzathine penicillin every 21 days, patients may experience recurrence of acute RF, making diagnosis a greater challenge. The development of new diagnostic methods can contribute to the early detection of rheumatic carditis (RC) considering that 90% of cases are subclinical. In addition, not only the usual gallium-67 scintigraphy, but also tests such as cardiac nuclear magnetic resonance imaging (MRI) and positron emission tomography (PET-CT) may have the potential to be diagnostic tools in acute rheumatic carditis. However, more studies are needed to validate these methods.

Case Report

A 23 years-old woman was admitted with a history of permanent atrial fibrillation, cardiac sequelae of RF and four cardiac surgeries (mitral, aortic and tricuspid valve repair in 2006 and 2007; aortic valve replacement with a biological prosthesis in the 2014). The patient has been treated with benzathine penicillin for secondary prophylaxis. Two years before, the patient was admitted to the emergency room with New York Heart Association functional class II. She had signs of right and left congestion, aortic systolic murmur, tricuspid regurgitation murmur, and silent mitral auscultation. A transthoracic echocardiogram (TTE) revealed decreased left ventricular ejection fraction (LVEF) (from 56% to 26%), and mitral and aortic prostheses dysfunction. The hypothesis of recurrence of acute RF was made mainly because of the reduction in LVEF. There was evidence of inflammatory activity (leukocytes 13,620/mm$^3$, erythrocyte sedimentation rate of 36 mm/h, lactic dehydrogenase 297 U/ml, C-reactive protein 2.5 mg/L). A gallium-67 scintigraphy revealed increased concentration of the radiopharmaceutical in the cardiac area, with a diffuse pattern of uptake (Figure 1). Oral prednisone (1mg per kilogram per day) was started with good clinical response. MRI was conducted 72 hours after initiation of steroids, which showed LVEF of 44%, non-ischemic focal myocardial fibrosis with native T1 and extracellular volume, leading to the hypothesis of inflammatory cardiomyopathy (Figure 2). Seven days after prednisone, another TTE showed a significant improvement in LVEF, to 50%. Then, a fluorodeoxyglucose-(18F-FDG) PET-CT was performed, which confirmed myocardial uptake (Figure 3). Finally, the patient was discharged to finish corticosteroid therapy and referred for elective mitral and aortic valve replacement.

Discussion and Conclusion

RF is more prevalent in low- and middle-income countries, reflecting the low level of primary preventive care especially in children and young adults. In Brazil, there are about 30,000 new cases of RF per year, of which approximately 15,000 may have...
cardiac involvement. Cardiac complications are the most feared complications of RF due to their chronic and sometimes disabling sequelae. RF is an often underdiagnosed condition since when a patient needs surgery it reflects an infection which occurred 10 or 20 years before. Medical diagnostic is challenging, as there is no gold-standard complementary test available to confirm an acute RF case.

RC can present as mild, moderate or severe disease. Besides, the most frequent presentation is subclinical which is defined as a functional or structural change in the heart, seen on echocardiography, consistent with
rheumatic heart disease, many times in the absence of a pathological heart murmur.⁷

There are diagnostic imaging methods that are useful to define a suspected RF carditis. Currently, endomyocardial biopsy, an invasive test with reduced sensitivity, is less used.⁸ Scintigraphy with gallium ⁶⁷, cardiac magnetic resonance and positron emission tomography (PET-CT) are important complementary tools.

Recent studies have evaluated the use of gallium scintigraphy in the diagnosis and prognosis of this disease and shown a high sensitivity (91-100%) of the method.⁹,¹⁰ Gallium scintigraphy allows the detection of RC activity and therapeutic follow-up and has its best diagnostic performance in the first three months of clinical presentation. In addition, it is a reliable method to distinguish acute from chronic and inactive RC.¹¹ PET-CT, despite its low sensitivity, has shown a positive predictive value close to 100. Finally, cardiac MRI has shown an 85% accuracy. Both methods have great potential to become standard tests for the diagnosis of acute RC.¹¹-¹⁴

This case report demonstrates that PET-CT and MRI are useful tests to diagnose acute RC. These recent imaging tools could not only establish the diagnosis, but also assess the effectiveness of the treatment. Gallium scintigraphy was also essential to reveal the cardiac inflammatory process, which was confirmed by MRI findings, suggesting inflammatory cardiomyopathy. In this context, PET-CT was used as a control test after treatment.

In this case report, we demonstrated the importance of multiple diagnostic imaging tools to early diagnose RF carditis, particularly after an unexpected reduction in the LVEF in a patient with valvular heart disease.

In conclusion, similarly to gallium-67 scintigraphy, MRI and PET-CT can be of high value in the diagnosis and follow-up of acute or recurrent RF.

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