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Acute rheumatic fever in a 61-year-old patient

Surto de febre reumática aguda em paciente de 61 anos

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ABSTRACT: The study and description of Acute Rheumatic Fever (ARF) in adults is scarce as its occurrence is rare over the age of 40 years old. We describe an ARF episode in a 61-year-old woman that presented with a one-month history of retrosternal pain, dyspnea, orthopnea and nonproductive cough. Her doppler echocardiogram revealed new findings of mitral valve thickening, calcification, reduced mobility and significant regurgitation, in addition to an ejection fraction of 39% and increased left ventricular and left atrium diameters. Cardiac magnetic resonance confirmed important left ventricular systolic dysfunction and enlargement, with severe mitral regurgitation and no delayed myocardial enhancement or edema. PET/CT with ^{18}F-FDG showed increased uptake in the left ventricular wall and papillary musculature. Endomyorcadial biopsy confirmed mild lymphohistiocytic myocarditis. Treated with Prednisone and Azathioprine, besides secondary prophylaxis with Penicillin G and prescription optimization, the patient presented significant clinical improvement on follow-up.

Keywords: Rheumatic fever; Middle aged; Myocarditis; Positron emission tomography computed tomography; Azathioprine.

RESUMO: O estudo e a descrição da febre reumática aguda (FRA) em adultos é escasso, dado que sua ocorrência é rara acima dos 40 anos de idade. Descrevemos um episódio de FRA em paciente feminina de 61 anos com uma história de um mês de dor retrosternal, dispnéia, ortopnéia e tosse não-produtiva. Dopplerecocardiograma revelou achados novos de espessamento da valva mitral, calcificação, mobilidade reduzida e regurgitação significativa, além de uma fração de ejeção de 39% e aumento dos diâmetros do ventrículo esquerdo e do átrio esquerdo. Ressonância magnética cardíaca confirmou importante disfunção e aumento de diamêtro sistólico do ventrículo esquerdo, com insuficiência mitral grave e ausência de realce tardio ou edema miocárdico. PET/CT com ^{18}F-FDG mostrou aumento de captação na parede ventricular esquerda e na musculatura papilar. A biópsia endomiocárdica confirmou miocardite linfohistiocitária leve. Tratada com prednisona e azatioprina, além de profilaxia secundária com penicilina G e otimização da prescrição, a paciente apresentou melhora clínica significativa no seguimento.

Descritores: Febre reumática; Meia-idade; Miocardite; Tomografia computadorizada com tomografia por emissão de pósitrons; Azatioprina.

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INTRODUCTION

Acute rheumatic fever (ARF) is the leading cause of acquired valvular heart disease in developing countries[1]. An ARF episode often occurs in children and young adults, especially from 4 to 15 years. Its most severe event is carditis, leading to chronic rheumatic heart disease (RHD)[1]. The study and description of ARF in adults is scarce as its occurrence is rare over the age of 40 years old, what leads to few attention paid by the doctors to this diagnosis in older patients[2]. The aim of this case report is to the describe an ARF episode in a 61-year-old woman and raise awareness to this differential diagnosis for patients above the most common age of presentation, especially in developing countries where rheumatic fever has still a considerable incidence. Searching on PUBMED for the last 10 years, we have found only two cases presenting with ARF above sixty years old[2,3].

CASE REPORT

A 61-year-old woman with a history of a previous ARF event 14 years ago was admitted in our hospital with a one-month history of retrosternal pain, dyspnea, orthopnea and nonproductive cough. She went to the emergency service when the symptoms began; a chest radiograph revealed signs of pulmonary congestion and a bilateral pleural effusion. Electrocardiography revealed only ventricular extra systoles, left bundle branch block and left atrial hypertrophy. The patient then received prescriptions of Furosemide and Carvedilol and was referred to our service.

On physical examination, she presented normal vital signs, bilateral rales, a 3+/6 mitral systolic murmur and a gallop rhythm (S3). She was followed as an outpatient and had no murmur on previous examinations. She had no arthritis and no skin rashes. Laboratory data: NT-proBNP 1117 pg/mL, ASLO normal. A chest CT showed bilateral pleural effusion and interlobular septal thickening, compatible with pulmonary congestion; a small pericardial effusion was present. Doppler echocardiogram showed an ejection fraction of 39%, left atrium (LA) 44 mm, left ventricular (LV) diffuse hypokinesia, LV systolic diameter 54 mm, LV diastolic diameter 63 mm. Mitral valve presented mild thickening, points of calcification on its ring and reduced mobility on the posterior leaflet, with significant regurgitation and maximum and average diastolic gradient estimated respectively as 11,8 mmHg and 5,8 mmHg (Figure 1). Previous echocardiogram from one year before had an ejection fraction of 57%, LA 35 mm, LV systolic diameter 34 mm, LV diastolic diameter 49 mm, no LV functional impairment or hypokinesia, mitral valve with minimal thickening, no calcification and minimal regurgitation.

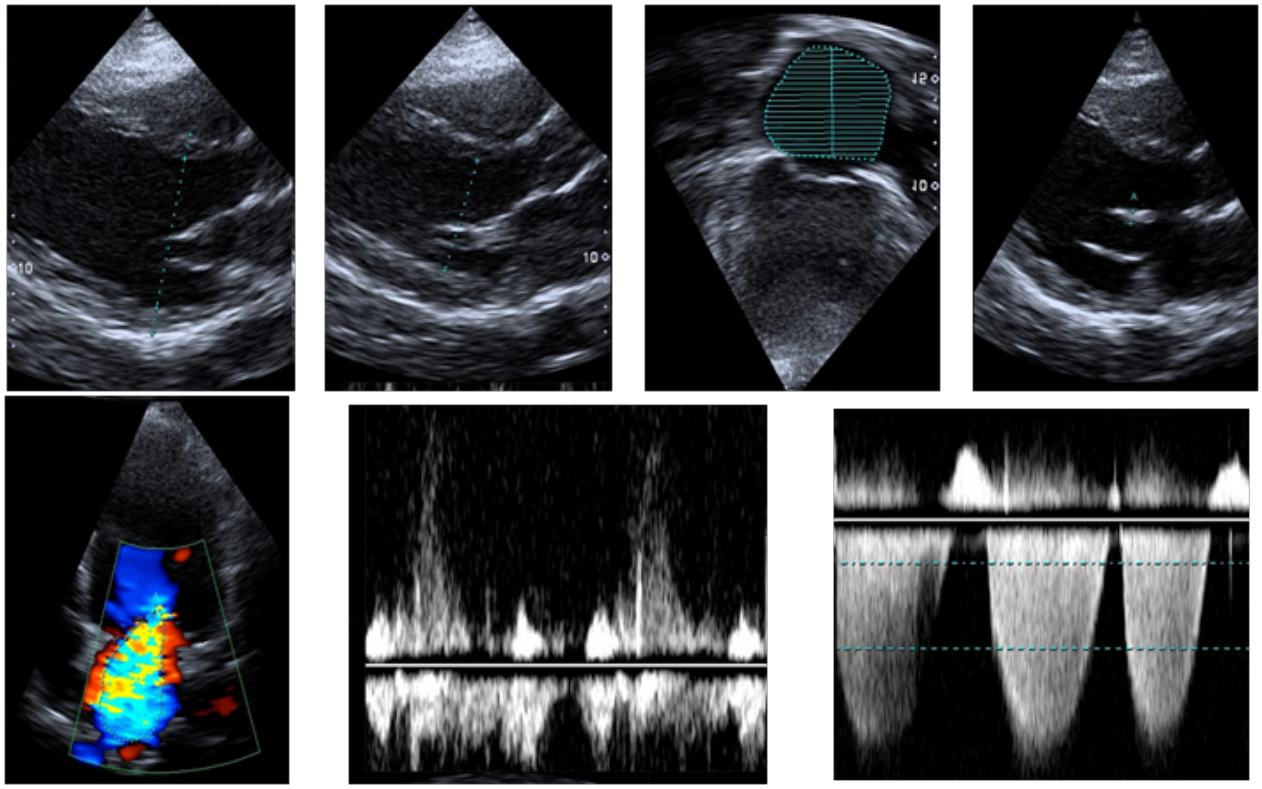

Figure 1 - Echocardiographic images representing, on the superior row from left to right: left ventricular diastolic diameter, left ventricular systolic diameter, left atrial volume and mitral valve thickening. On the inferior row: 4-chamber color doppler showing important mitral regurgitation; continuous-wave doppler from pre and post-ARF mitral regurgitation

Cardiac magnetic resonance confirmed important left ventricular systolic dysfunction and enlargement, with severe mitral regurgitation (43% of the stroke volume). There was no delayed myocardial enhancement or edema, making less likely the hypothesis of infarction or viral myocarditis (Figure 2).

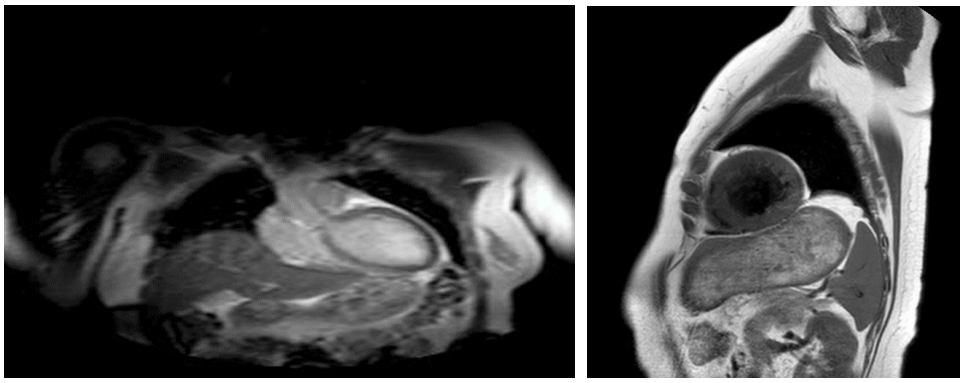

Figure 2 – Cardiac magnetic resonance image showed important increase in left ventricular dimensions, decreased left ventricular ejection fraction (30%), important mitral regurgitation (43% of the stoke volume), diffuse hypokinesia, dyssynchrony of the interventricular septum and absence of myocardial edema or delayed enhancement

Cardiac catheterization revealed normal pressures on the pulmonary artery and on the right chambers. PET/CT with ^{18}F-FDG was performed 24 h after specific diet to avoid physiological myocardial glucose uptake and showed a heterogeneously diffuse increased uptake of fluorodeoxyglucose in the left ventricular wall and papillary musculature, attributed to active rheumatic carditis (Figure 3). Gallium scintigraphy showed a positive study for active cardiac inflammatory process as well.

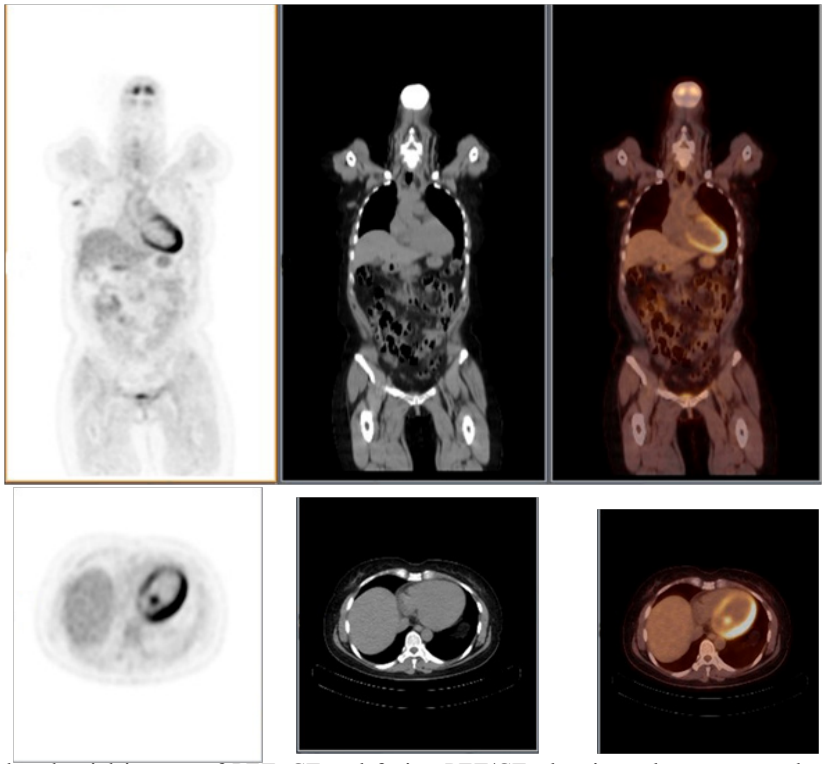

Figure 3 – Coronal and axial images of PET, CT and fusion PET/CT, showing a heterogeneously diffuse increased uptake of fluorodeoxyglucose in the left ventricular wall and papillary musculature

An endomyocardial biopsy, performed together with the cardiac catheterization, revealed discrete cardiomyocyte hypertrophy, interstitial edema, mild perivascular inflammatory infiltrate and absence of Aschoff nodules on hematoxylin-eosin staining. Immunohistochemical reactions for T lymphocytes (CD3) and macrophages (CD68) showed focal positivity (less than 24 cells/mm2) – (Figure 4).

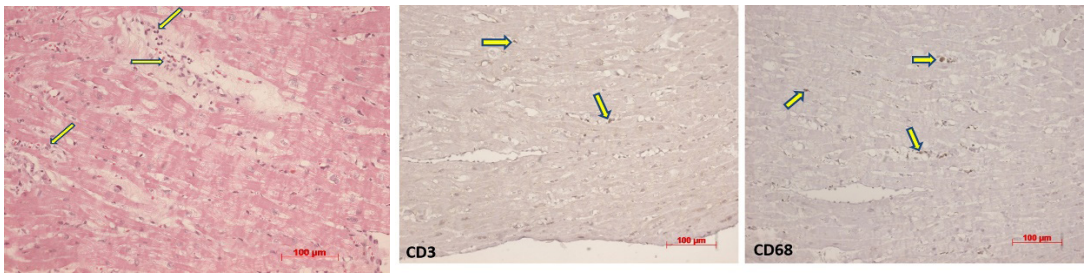

Figure 4 - Endomyocardial biopsy showing discrete cardiomyocyte hypertrophy, interstitial edema and mild perivascular inflammatory infiltrate (arrows). Absence of Aschoff nodules. Hematoxylin-eosin staining. Immunohistochemical reactions for T lymphocytes (CD3) and macrophages (CD68) showing focal positivity (less than 24 cells / mm2). Counter-staining by Harris hematoxylin

We then suspended Carvedilol and introduced Prednisone, Azathioprine, Enalapril, Spironolactone, Digoxin, Penicillin G plus fluid restriction, with the presumptive diagnosis of ARF. The patient presented rapid and significant clinical improvement on follow-up.

DISCUSSION

The Jones criteria is classically used for the diagnosis of Rheumatic Fever. However, it has been described considering mostly the presentation of first ARF events in children and young adults[4-6]. Recurrent attacks in older adults can often present with atypical clinical features, considering the classical parameters of the Jones criteria[7,8]. As such, clinical judgement and a strong differential diagnosis is essential for adults presenting with features that could be explained by an ARF event.

As our patient presented miocarditis, valvulitis, previous history of ARF and no features of other differential diagnosis, our final diagnosis was a recurrent attack of ARF. We optimized the prescription and introduced, besides secondary prophylaxis with Penicillin G, Prednisone and Azathioprine, a new approach for rheumatic carditis treatment, considering a recent review discussing the need for using new drugs in addition to glucocorticoids[9].

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