Myocardiopathy and Rheumatoid Arthritis. A Patient Awaiting a Heart Transplant

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Cardiovascular pathology is common in rheumatoid arthritis. However, myocardial affection is unusual and clinical disease is rare. We report a case of dilated cardiomyopathy in a patient with rheumatoid arthritis and progressive heart failure that required inclusion into a heart transplantation list.

Key words: Rheumatoid arthritis. Cardiomyopathy. Heart transplantation.

Miocardiopatía y artritis reumatoide. Una paciente en lista de trasplante cardíaco

La enfermedad cardiovascular es común en la artritis reumatoide. No obstante, la afección miocárdica es inusual y suele cursar de forma asintomática. Presentamos un caso de miocardiopatía dilatada en una paciente con artritis reumatoide e insuficiencia cardíaca progresiva que requirió su inclusión en lista de trasplante cardíaco.

Palabras clave: Artritis reumatoide. Miocardiopatía. Trasplante cardíaco.

Introduction

Rheumatoid Arthritis (RA) is a systemic disease with extra-articular manifestations. Cardiovascular disease is common in RA and is the most frequent cause of death in these patients, but its incidence is not very superior to the one in the general population. Myocardial affection is uncommon and is usually an autopsy finding because in most cases it is asymptomatic. Cardiac tissue granulomas suggest a specific myocardial affection.[1] We present the case of dilated cardiomyopathy in a patient diagnosed with RA with progressive heart failure that lead to her inclusion on a waiting list for heart transplant.

Clinical Case

A 46-year old woman was diagnosed with RA after presenting with symmetrical polyarthritis predominat in the hands, with a positive rheumatoid factor, treated over the course of her disease with non-steroidal anti-inflammatory drugs (NSAID) and oral corticosteroids at variable doses, with a good progression regarding the joints. After 25 years since the onset of disease the patient was hospitalized after a 6-week episode that included progressive dyspnea, culminating in dyspnea during rest, associated to orthopnea, and edema. Upon physical examination there was a systolic murmur at the apex, bilateral crepitant rales, and pitting edema on both pretibial areas, and the laboratory results showed an increase in cardiac enzymes with a maximum total creatin kinase of 1184 and an MB fraction of 53, an ESR of 54, and negative cardiotrophic virus scan. On the EKG, sinus rhythm was 65 beats/min and a complete blockage of the left His bundle was evident. Ecocardiogram manifested a dilated left ventricle with a poor systolic function (LVEF, 45%). After being diagnosed with congestive heart failure, acute myocarditis, and dilated cardiomyopathy with moderate systolic dysfunction, she was discharged and loop diuretics and Amiodarone are added to her previous treatment of enalapril.

She returns to the hospital after 5 months due to myocarditis and elevated heart enzymes and an EKG that showed atrial fibrillation with ventricular extrasystoles, without ecocardiographic changes, initiating treatment with digoxin and coumadin. A myocardial scan is carried out (Figure 1) observing inferior hypoperfusion compatible with myocarditis. After 4 years of being clinically stable she presents worsening rest dyspnea with hemodynamic repercussion (blood pressure, 90/40 mm Hg), an increase in respiratory rate, crepitant rales, edema, and liver

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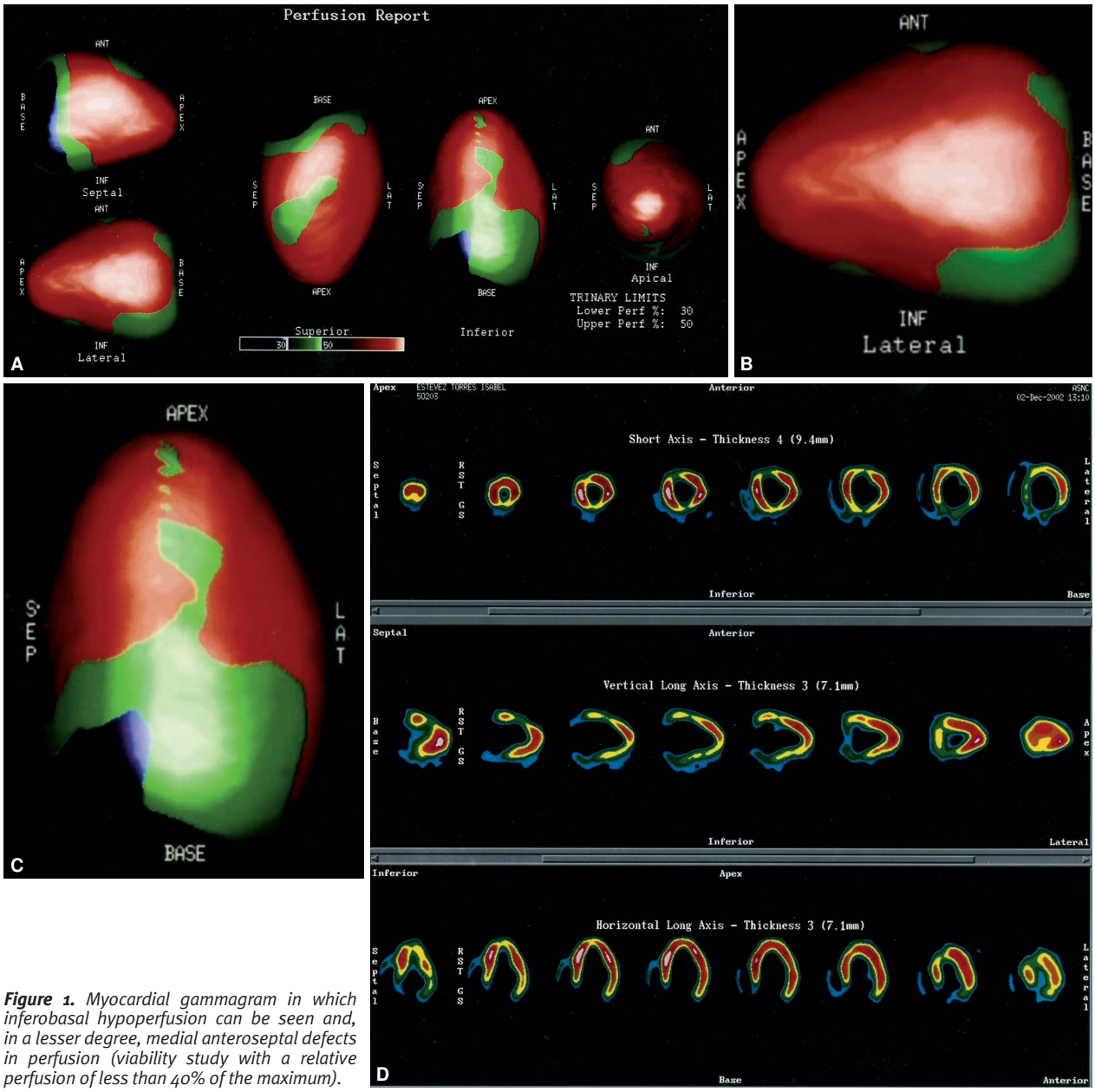

Figure 1. *Myocardial gammagram in which inferobasal hypoperfusion can be seen and, in a lesser degree, medial anteroseptal defects in perfusion (viability study with a relative perfusion of less than 40% of the maximum).*

enlargement, and in the ecocardiogram (Figures 2-4) a dilated left ventricle with a poor function (LVEF, 12%) was observed. A cardiac catheterization and coronariography was carried out and it showed normal coronary arteries: CI, 1.7 L/min/m^2; PVR, 2.5 U Wood/m^2, and PAP, 27 mm Hg. She was diagnosed with congestive heart failure with a functional class of III-IV according to NYHA secondary to dilated cardiomyopathy, including her in a waiting list for cardiac transplant.

The extracted anatomical piece did not have coronary atheromas nor were there areas of recent, or previous infarction on the myocardium. The atrioventricular valves, ant the aortic and pulmonary valve did not have any structural alterations. The left ventricle was very dilated and white patchy lesions in the endocardium of both ventricles, more evident on the entry and exit tracts of the left ventricle, corresponding with areas of subendocardial fibrosis and geographically with areas of poteroinferior akinesia in the ecocardiogram and of inferobasal hipoperfusion in the myocardiac scan (a viability study with a relative perfusion of less than 40% of the maximum) and in a lesser degree in the medial anteroseptal level.

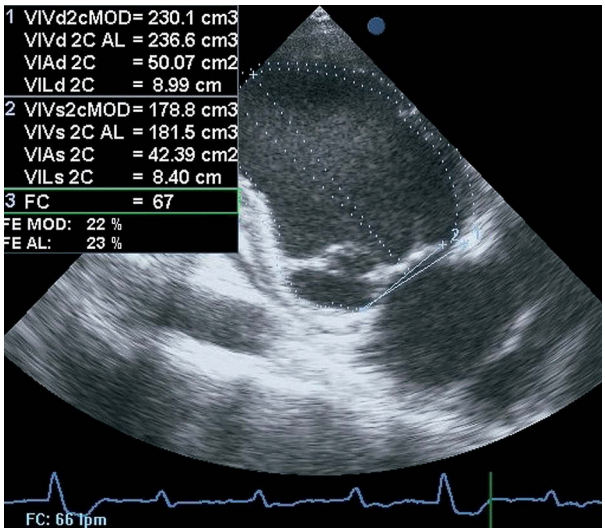

Figure 2. *Bidimensional image showing a very enlarged left ventricle with poor general contractility (systolic dysfunction) and marked akynesia in the posterior wall of the ventricle.*

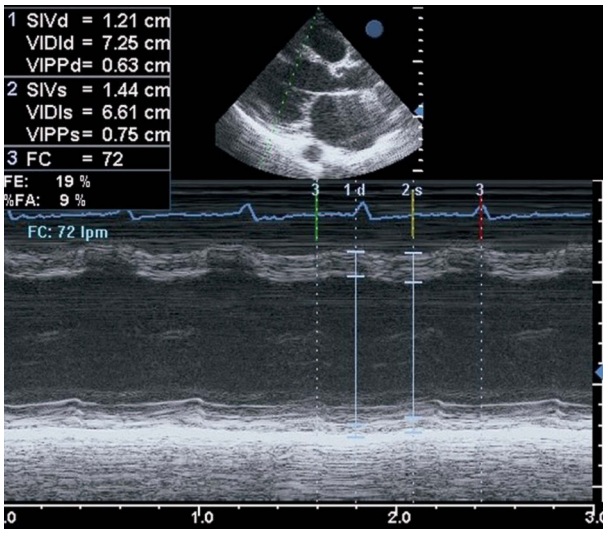

Figure 4. *M mode image (space and time) in which a dilated left ventricle with marked generalized hypokinesia can be seen.*

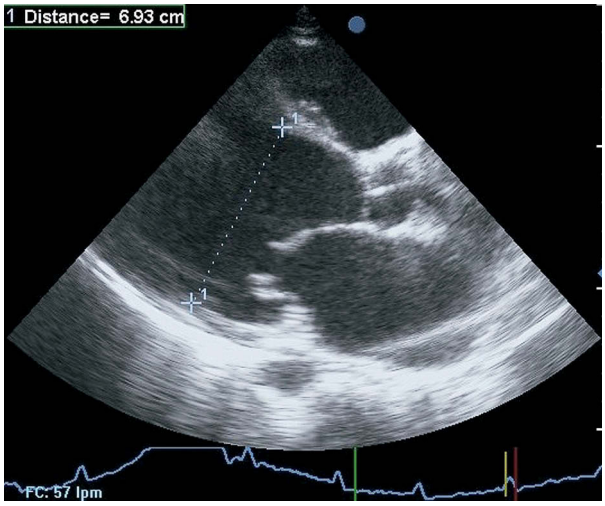

Figure 3. *Increased ventricular teledyastolic diameter (69.3 mm; normal up to 58 mm).*

Discussion

Cardiovascular manifestations in RA are found in relation to granulomas in the cardiac tissue or in the development of vasculitis.[2] The forms of presentation are pericarditis, myocarditis, endocarditis, valvular disease, conduction defects, or coronary lesions.[3] Autopsy findings frequently find cardiac lesions, but only a small proportion has clinical consequences.[4] The most frequent cause of death in RA is cardiovascular disease and accelerated atherosclerosis due to vasculitis, which increases the serum concentration of lipoproteins and causes hypofibrinolysis. In general, cardiomyopathies are divided according to their primary causes into those of unknown cause and those due to secondary causes or associated to other diseases or conditions in other organs and systems (infections, metabolic, deposit, toxic, periparthum, connective tissue disease, associated to deficiency). Because establishing a causal relationship is frequently difficult, we commonly rely on the use of the physiopathological classification in dilated, restrictive, and hypertophic. Ecocardiographic, gammagraphic, coronariographic, and radionuclide scanning differences can be found in these three types of myocardiopathy (Table).

Myocardiopathy in RA is uncommon. Viral, genetical, and autoimmune factors have been implicated in its pathogeny.[5] Animal models have shown us that the presences of certain inflammatory cytokines, found also in large quantities in the synovium of RA patients, are associated to congestive heart failure un myocardiopathy.[6] There are two described forms of myocardiopathy in RA. One is unspecific, more frequent, with an inflammatory infiltrate and the other is specific, granulomatous, and associated to rheumatoid nodules. Clinically they can lead to the development of dyspnea, congestive heart failure, arrhythmia, and alterations in heart conduction, as was the case in our patien.[1]

Even if the ecocardiographic study allows us to perform the diagnosis of myocardiopathy, for a definite diagnosis an endomyocardial biopsy must be performed. Early treatment must be established with steroids and immunosupressants (cyclophosphamide). In case of severe dysfunction of the ventricular function, it might be even necessary to consider myocardial transplant.[10]

Ecocardiographic, Gammagraphic, and Coronariographic Differences Between Different Types of Myocardiopathy

	Dilated	Restrictive	Hypertrophic
Echocardiogram	Dilatation and dysfunction of the left ventricle	Increase in the width of the left ventricle wall	Asymmetric hypertrophy of the septum
Gammagraphy	Dilatation and dysfunction of the left ventricle	Normal or slightly decreased systolic function	Energetic systolic function; defects in perfusion
Catheterism	Dilatation and dysfunction of the left ventricle; increase in left filling pressures; reduction in cardiac output	Normal or slightly decreased systolic function; increase in left and right filling pressures	Energetic systolic function; dynamic obstruction to the flow of the left ventricle; increase in left and right filling pressures

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