Suplemento da Revista da
SOCESP
Sociedade de Cardiologia do Estado de São Paulo

CASE HISTORY OF RIGHT VENTRICULAR ENDOMYOCARDIAL FIBROSIS IN A NONAGENARIAN PATIENT

RELATO DE ENDOMIOCARDIOFIBROSE DE VENTRÍCULO DIREITO EM PACIENTE NONAGENÁRIA

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Received on 10/09/2018,
Accepted on 05/09/2019,

ABSTRACT

Endomyocardial fibrosis is a rare, recently-described, restrictive cardiopathy. It was first described in 1938 by Williams and studied from an anatomopathological perspective by pathologist Davies in South Africa, receiving the eponym Davies Disease in 1948. In terms of the natural history of this disease, there is a higher incidence of diagnoses in the third and fourth decades of life, at a mean age of 32 years. We want to raise the awareness of the scientific community by reporting the case of a woman who was first diagnosed with endomyocardial fibrosis at 90 years of age and pose questions about the incidence and subdiagnosis of this disease, as well as about its evolution.

Keywords: Endomyocardial Fibrosis; Nonagenarians; Heart Failure; Aged, 80 and over.

RESUMO

A endomiocardiofibrose é uma cardiopatia restritiva, pouco comum, com descrição recente. Foi descrita pela primeira vez em 1938, por Williams e estudada do ponto de vista anatomopatológico pelo patologista Davies, na África do Sul, recebendo o epônimo Doença de Davies em 1948. Considerando a história natural dessa doença, há maior incidência de diagnósticos nas terceiras e quartas décadas da vida, com média de 32 anos. Queremos chamar a atenção da comunidade científica, relatando o caso de uma senhora com diagnóstico inicial de endomiocardiofibrose aos 90 anos e indagar sobre a incidência e o subdiagnóstico dessa doença, bem como sua evolução.

Descritores: *Fibrose Endomiocárdia; Nonagenários; Insuficiência Cardíaca; Idoso de 80 anos ou mais.*

INTRODUCTION

Endomyocardial fibrosis (EMF), a rare disease first described by Williams in 1938, is characterized by fibrous myocardial thickening and restrictive underlying endocardium.[1]

Davies, in 1947, recognized the significance of the lesions by successive autopsies on young patients with mural fibrosis. In his work with Ball, and later with Connor, he described the histological and macroscopic characteristics found in EMF that were defined as Davies Disease.[1]

It can occur in any gender or race, and is more frequent in young adults, with two incidence peaks, one from 10 to 15 years and the other around 30 years,[1] with an average age of 32 years.[2] In Brazil, the first report dates from 1966[3] and women predominate five times more often than men.[4]

Although the date of the first publication was advanced, a clear pathophysiological mechanism has not yet been clearly established.

It is a disease prevalent in the tropics and economically underdeveloped regions.[5]

EMF is a rare heart disease that affects only the heart and can affect one or both ventricles, being right ventricular (RV) involvement is a minor incident form.[6]

Its insidious evolution allows disease progression in periods represented by few symptoms, but later evolves with severe hemodynamic repercussion due to heart failure.

In the field of therapy, surgical treatment is not yet the best option, since it does not prevent disease progression,[7] being clinical therapy the main pillar.

We bring this report in order to show a unique incidence that is beyond the previously published ranges, showing the favorable evolution of the patient through clinical treatment.

CASE REPORT

We will report the case of a female patient, 92 years old, caucasian. With a history of two pregnancies, uneventful and with no history of heart disease. Patient sought medical service due to dyspnea on moderate exertion accompanied by weakness of lower limbs. During outpatient investigation, the patient evolved with worsening functional class and was

then referred to the local emergency room. Admitted to NYHA functional class III, no complaints of chest pain.

At transthoracic Doppler echocardiogram, left ventricular septum and wall with normal thickness and movement, as well as left ventricle (LV) cavity with preserved size and function. Left atrium (LA) increased to a significant degree. This examination also revealed mitral, aortic, and tricuspid valves with moderate insufficiency accompanied by grade III LV diastolic dysfunction, a restrictive pattern, and moderate grade pericardial effusion, drawing attention to the presence of echogenic imaging within RV of unclear etiology.

Continuing the investigation of echocardiographic results, cardiac magnetic nuclear resonance (MRI) presented the following morphofunctional parameters: LV with a fraction of 69%, while RV had an ejection fraction of 38%, with apical obliteration suggestive of myocardial and subendocardial fibrosis accompanied by thrombus. and/or calcification in its apical portion. The volumes and thickness of ventricular walls were preserved except for ventricular tip. MRI also included dilation of the inferior vena cava, significant enlargement of the right atrium and mild pericardial effusion. Thus, the results obtained were compatible with isolated RV endomyocardial fibrosis. (Figure 1)

After drug optimization, the patient evolved well, responding to the proposed treatment for heart failure, with improvement of all symptoms related to hospitalization, being discharged with clinical follow-up. Since then, after two years of hospitalization, the patient remains stable, with no further compensation to date.

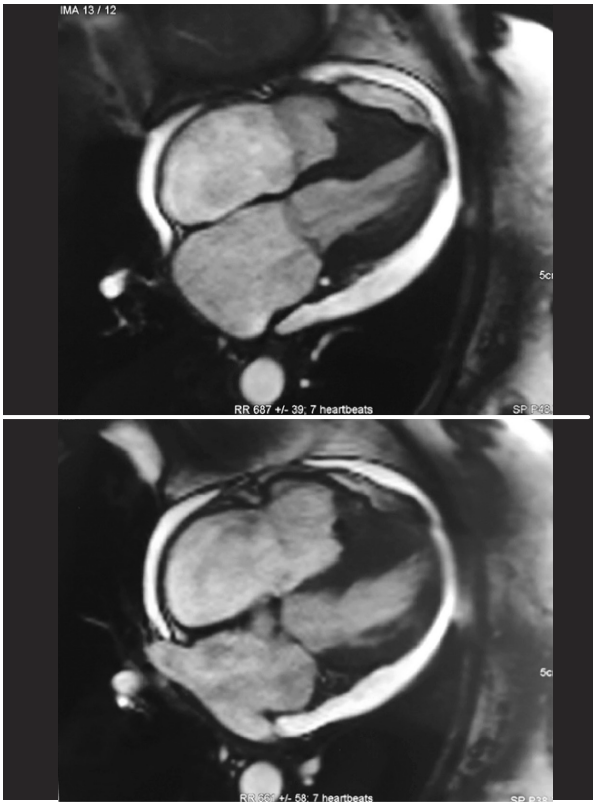

Figure 1. Cardiac MRI shows obliteration of the right ventricular apex and significant enlargement of the right atrium.

DISCUSSSION

In 1948, Davies, through anatomopathological study characterized EMF as a fibrous thickening of the myocardium and endocardium that could occur in any gender or race, being more frequent in young adults. In Brazil, the first report dates from 1966[3], with women predominating five times more often than men.

Regarding the affected age group, EMF has a bimodal frequency at 10 and 30 years of age.[8] In the study by Fernandes et al.[2], the average age was 32 years and on the study by Barreto et al.[9], the maximum age in the sample was 65 years. In the review by Fagundes et al.,[5] EMF is predominantly in women, adults and young people, ranging in age from two years to 56 years of age. In the study by Mady et al.,[17] the age of the subjects ranged from nine to 65 years, while the registration of the patient in this report is dated 1926, an age rarely reached by a patient with the same diagnosis.[2,9] There was opening of the heart failure condition at 92 years of age, exceeding the upper limit of 50 years in Iglezias1 study sample and well above the highest age reported by a Brazilian article.[10]

Although the date of the first publication was advanced, no etiological mechanism has yet been clearly established. Some hypotheses proposed by Davies[11] range from viral infections to antigen-antibody reactions and malnutrition. Toxoplasma gondii infection was suggested, but results without statistical values overturned the thesis.[12,13]

Eosinophilia had also been hypothesized, since in some series, laboratory tests show eosinophilia of up to 10% in one study[1,4,5,9,14] however with no confirmed hole.[15]

It is a predominant disease in Africa as the main cause of HF, extending mainly through the tropics and economically underdeveloped regions.

EMF is a heart disease with a well-defined pathological finding known as fibrous endocardial thickening. It focuses exclusively on the heart and may affect one or both ventricles, and RV involvement is a minor incident form of the disease. Isolated LV involvement is more frequent in Brazil,[1] and isolated right ventricular involvement occurs in less than 10% of patients.[5]

Depending on the degree of fibrosis, it can affect papillary muscles and consequently the valves without, however, causing direct injury to the leaflets.[1]

Its insidious evolution allows disease progression in periods represented by few symptoms, and may later evolve with severe hemodynamic repercussion.

Clinical onset may be oligosymptomatic, with the onset of right or left heart failure depending on the degree and intensity of fibrosis and the affected chamber.[7] In the case of RV involvement, the symptoms presented may be lower limb edema. mild to severe ascites, associated with pericardial effusion, dyspnea, and hepatomegaly that worsen according to the degree of fibrosis.[1,4,5,9,15]

Chest X-ray can be used as a diagnostic aid, presenting cardiomegaly of varying degree, with increased cardiothoracic index (ICT). In right ventricular EMF, the Cardiothoracic Index is usually greater than 0.7 and, in biventricular cases, is greater than 0.5. This test may help in the perception of mono or biventricular involvement, as in the study by Fernandes et al.[2,4,5]

Electrocardiogram is usually nonspecific, but may show low QRS voltage, first-degree atrioventricular block, and atrial

fibrillation, especially in right ventricular involvement, helping to identify the type of ventricular involvement. Bradycardia or bradi-tachy syndrome may be manifestations of sinus node involvement.[4,5,16]

Echocardiogram is the exam of choice for establishing the diagnosis, showing ventricular involvement. Basal inferior wall thickening and apical obliteration by fibrosis are the findings of the disease, which may involve the papillary muscles. There is usually thrombus deposition or even fibrosis-associated calcification, with apical contractility usually preserved. There are varying degrees of atrioventricular valve insufficiency, ventricular size is normal, and the atria are dilated. Systolic function is preserved and diastolic function is altered, ranging from the relaxation reduction pattern to the restrictive pattern. The ejection fraction is usually preserved. The presence of mild and moderate pericardial effusion can also be found.[4,17]

After the examination, the following echocardiographic criteria described by Mocumbi et al.[18] should be analyzed (Table 1)

Diagnosis requires two major criteria including apex obliteration and right ventricular thrombus and two minor criteria.[4]

In the field of therapy, surgical treatment is not yet the best option, as it does not prevent disease progression. A report of four cases showed ventricular fibrosis recurrence after surgical treatment of EMF, of which two deaths due to low debt. Thus showing the high risk of surgical treatment,[7] making clinical therapy the main pillar of EMF treatment.

Disease prognosis depends on the location and degree of cardiac involvement. Survival is two years after diagnosis in 95% of cases due to sudden death or heart failure, according to Roberts et al., and depends on the degree of cardiac function and the affected chamber.[3] The presence of atrial fibrillation is associated higher prevalence of tricuspid regurgitation and evolving with worse prognosis.[19,20]

Table 1. Echocardiographic criteria.

Major	Minor
Endomyocardial plaques >2 mm	Ventricular wall affected by endomyocardial mass
Thin endomyocardial thickness <1 mm in more than one ventricular wall area	Restricted flow in atrioventricular valves
Ventricular apex obliteration	Diastolic opening of the pulmonary valve
Thrombus without severe ventricular dysfunction	Slight thickening of the anterior mitral leaflet
Right ventricular apex notch	Enlarged atrium with normal size ventricle
Atrioventricular dysfunction secondary to ventricular wall adhesion	Interventricular septum M movement and posterior wall FLAT
	Increased density of the middle or other ventricular band

Given the above, we should ask ourselves if such a pathology could not present itself in older patients, out of the ranges previously published and, therefore, still underdiagnosed in this population.

Given the fact, this article aims to show the scientific society that such a pathology may be more incident and insidious than we currently know. Showing the importance of reporting new cases in order to emerge more information about the disease so that we can increasingly optimize its treatment.

CONCLUSION

According to literature data, EMF is still a rare disease and little investigated despite the adverse prognosis.

What makes this case really unique is the incidence that evades all standards previously published. In addition to good response to clinical therapy.

CONFLICTS OF INTEREST

The author declares that he has no conflicts of interest in this work.

AUTHORS' CONTRIBUTIONS: Each author contributed individually and significantly to the development of the manuscript. KFSI and RNA were the main contributors in the writing of the manuscript. KFSI and RNA followed the patient, gathered clinical data, performed the literature search and discussed the data. RNA, KFSI, FRL and HIAE reviewed the manuscript and contributed to the intellectual concept of the study.

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