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CASE REPORT

Fulminant myocarditis—Case report☆

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KEYWORDS
Fulminant myocarditis;
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Abstract A 46-year-old woman was admitted due to diplopia because of ophthalmoplegia, which improved with corticosteroid therapy. Eight days later, she was admitted with fulminant myocarditis in cardiogenic shock, with severe left ventricular dysfunction and frequent episodes of nonsustained ventricular tachycardia. As there was no clinical improvement, an endomyocardial biopsy was performed that revealed inflammatory infiltrate, vasculitis, and PCR positive for cytomegalovirus, Epstein–Barr virus, parvovirus B19 and enterovirus. Left ventricular function recovered with heart failure treatment and corticosteroids. Three months later, after progressive withdrawal of prednisolone, there was recurrence of myocarditis and left ventricular dysfunction, which was successfully treated by restarting corticosteroid therapy. One month later she was readmitted with fulminant myocarditis which again responded to steroids. She intermittently presented cutaneous purpura lesions. At this time the provisional diagnosis was vasculitis and she started monthly cycles of cyclophosphamide. Before the second cycle, she was admitted with pneumonia and ventricular dysfunction and died.

PALAVRAS-CHAVE
Miocardite fulminante;
Corticoterapia;
Vasculite

Miocardite Fulminante – a propósito de um caso clínico

Resumo Mulher de 46 anos foi internada por diplopia devido a oftalmoplegia, que melhorou com corticoterapia. Oito dias depois, iniciou quadro de miocardite fulminante associada a choque cardiogénico por disfunção ventricular esquerda grave e episódios frequentes de taquicardia ventricular não mantida. Por não apresentar melhoria clínica, foi submetida a biópsia endomiocárdica que revelou infiltrado linfocitário, sinais de vasculite e PCR positiva para citomegalovírus, vírus Epstein-Barr, enterovírus e parvovírus. Após tratamento da insuficiência cardíaca e corticoterapia, recuperou a função ventricular. Três meses depois e após a suspensão de prednisolona, teve recorrência da miocardite com disfunção ventricular, tratada com sucesso após reinício de corticóides. Um mês depois, foi reinternada com o mesmo quadro que respondeu ao aumento da dose de corticóides. Apresentou intermitentemente lesões cutâneas

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tipo púrpura. Colocou-se a hipótese de vasculite e iniciou tratamento com ciclos mensais de ciclofosfamida. Antes do segundo ciclo, foi internada com pneumonia associada a disfunção ventricular e faleceu.

Introduction

According to the Dallas criteria, myocarditis is characterized as an inflammatory infiltrate of the myocardium. The most common cause is viral infection; less often it can be secondary to a non-viral infection, hypersensitivity to drugs or toxins, autoimmune disease, giant cell myocarditis or sarcoidosis.[1] Clinical manifestations range from asymptomatic electrocardiographic (ECG) alterations, through non-specific systemic symptoms such as fever, myalgia, palpitations or exertional dyspnea, to cardiogenic shock and sudden death.[2] In 1991, Lieberman et al.[3] proposed a clinicopathologic description that classified myocarditis as fulminant, subacute, chronic active or chronic persistent. The clinical diversity of myocarditis makes it impossible to determine its real incidence, but it is estimated to cause 8.6–12% of sudden deaths in young adults and 9% of cases of dilated cardiomyopathy.[1,2] Its prognosis and treatment depend on the underlying etiology and the clinical and hemodynamic repercussions.

Case report

A 46-year-old woman, working in air traffic control, with a history of smoking and right peripheral facial paralysis 15 years previously (with complete recovery), was admitted in June 2009 to the neurology department of Faro Hospital due to horizontal diplopia of sudden onset. She developed limited eye movement and bilateral ptosis but with preserved pupillary reflexes. Etiologic study included magnetic resonance imaging (MRI) of the cranium and orbits, electromyogram, and analysis of cerebrospinal fluid, including serology for Epstein–Barr virus (EBV), cytomegalovirus (CMV), herpes simplex 1 and HIV 1 and 2, and VDRL, none of which showed abnormalities. The chest CT was also normal and excluded thymoma. Immunologic study for ANA, ANCA, ECA and rheumatoid factor was negative, but was positive for skeletal muscle antibodies. She was treated with three 1-g methylprednisolone pulses and she was discharged two weeks after admission, with only right eye ptosis.

The provisional diagnoses were rapidly progressive external ophthalmoplegia, ocular myasthenia and mitochondrial myopathy. During this hospitalization, ECG, transthoracic echocardiography (TTE) and troponin I and BNP measurement were not performed.

Eight days after discharge she was admitted to the cardiology department of the same hospital with fulminant myocarditis in cardiogenic shock and with frequent episodes of nonsustained ventricular tachycardia (VT) (Figure 1).

Laboratory test results included troponin I 12 ng/ml (normal <0.1), BNP 2635 pg/ml (normal <100) and CRP 76 mg/dl (normal <5). The ECG showed sinus rhythm with incomplete right bundle branch block and symmetric negative T waves in V1–V6. TTE (Figure 2) revealed nondilated chambers, severe left ventricular (LV) dysfunction (LV ejection fraction [LVEF] 25%) with hypokinesia of multiple segments, and no pericardial effusion. Corticosteroid therapy was restarted (IV prednisolone 1 mg/kg). On the fifth day, although no longer under IV inotropic support, she continued to present dyspnea at rest and frequent episodes of nonsustained VT and was therefore transferred to the advanced heart failure care unit of Coimbra University Hospitals with suspected giant cell myocarditis (GCM). Cardiac catheterization showed no angiographically significant coronary lesions; endomyocardial biopsy (EMB) of five specimens (Figure 3) revealed infiltrations of T lymphocytes, macrophages and CD20-positive B lymphocytes and signs of vasculitis, but did not detect multinucleated giant cells. Polymerase chain reaction analysis of the biopsy specimens was positive for parvovirus B19, enterovirus, EBV and CMV.

Serology for CMV, herpes simplex and EBV was positive for IgG but negative for IgM, and was negative for HIV, hepatitis C virus, *Borrelia* and *Coxiella*. Tests for smooth muscle antibodies were positive but were negative for skeletal muscle antibodies, unlike the result during the first hospitalization. The patient showed gradual clinical improvement and recovery of LV function under treatment with prednisolone and was discharged one month later with a diagnosis of fulminant myocarditis of probable autoimmune origin.

She remained in NYHA functional class I until the end of October 2009, when three days after suspension of corticosteroid therapy for the purpose of a muscle biopsy, she was readmitted for recurrence of myocarditis, severe LV dysfunction, cardiogenic shock and renal and liver failure. Steroids were restarted, leading to clinical improvement and recovery of LV function, and she was discharged under heart failure treatment and oral prednisolone. She underwent muscle biopsy as an outpatient under prednisolone 50 mg/day, which showed type II fiber atrophy, compatible with steroid-induced and/or disuse myopathy. Control TTE showed nondilated chambers and good LV global systolic function (LVEF 57%). In order to reduce steroid use, azathioprine 100 mg/day was introduced.

In mid-December, coinciding with a reduction in prednisolone from 50 to 40 mg/day, she was readmitted to the cardiology department with hemodynamically tolerated sustained VT, which was converted to sinus rhythm by amiodarone. TTE again showed severe LV dysfunction. A 50-mg IV prednisolone bolus was administered and the oral dose was increased to 1 mg/kg/day, and the daily dose of azathioprine was increased to 150 mg/day.