CARDIOVASCULAR FLASHLIGHT

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Lethal enterovirus myocarditis in a patient with granulomatosis with polyangiitis following rituximab and high-dose steroid therapy

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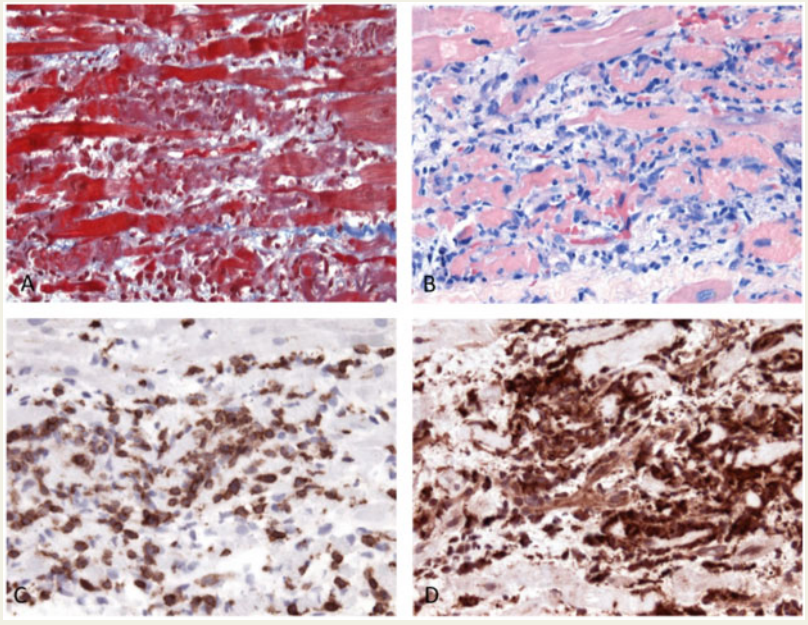
A 42-year-old woman was diagnosed with granulomatosis with polyangiitis (GPA) revealing positive cANCA and proteinase- 3 antibodies. The computed tomography of the thorax revealed a persisting 7 cm large lesion in the right upper lobe of the lung. A remission-maintaining therapy with 500 mg rituximab every 6 months in combination with low-dose prednisolone was performed. Two years later, recurrent palpitations, nausea, and fatigue occurred. ECG showed a 3rd-degree AV block. Laboratory results revealed elevated heart enzymes (troponin I 2.6 µg/mL, CK 534 U/L). CHD was ruled out. Echocardiography did not show any relevant changes. A DDD pacemaker was implanted without complications. A few days after discharge, the patient presented with cardiogenic shock. On Day 3, the patient had to be resuscitated and extracorporeal life support was performed. Echocardiography showed a severely reduced ejection fraction of both ventricles. Assuming a cardiac manifestation of GPA, high-dose steroid administration and repeat administration of 650 mg rituximab were performed. Multi-organ failure with severe coagulation disorder occurred. Meanwhile, endomyocardial biopsies (EMB) were performed revealing a fulminant lymphocytic myocarditis with extensive myocardial necrosis (see *Panels A–D*). RT-PCR revealed a high load of enteroviral RNA in the myocardium; sequencing of the PCR product identified coxsackievirus B2 RNA. After interim clinical improvement, the patient died on the 11th day of immunosuppressive treatment. This case report stresses the relevance of EMB for the correct diagnosis of acute heart failure, thus avoiding the lethal immunosuppressive therapy of enteroviral myocarditis by rituximab and steroids.



Fulminant lymphocytic myocarditis. (*Panel A*) Masson Trichrome and (*Panel B*) Giemsa staining reveal myocyte necrosis and inflammation with mononuclear infiltrates. Immunohistochemical stainings demonstrate high amounts of (*Panel C*) CD3+ T lymphocytes and (*Panel D*) CD68+ macrophages (×400).

Data are available from the authors.

Conflict of interest: The authors have submitted their declaration which can be found in the article Supplementary Material online.