G15 Systemic Lupus Erythematosus and Fatal Cardiac Failure Due to Pancarditis in a Young Man

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The goal of this presentation is to present a case of sudden cardiac failure and death in a 28-year-old Caucasian male, with reactivation of Systemic Lupus Erythematosus (SLE). A complete methodological forensic approach by means of autopsy, histological, and immunohistochemical examinations lead investigators to conclude an acute congestive heart failure due to pancarditis as cause of death.

This presentation will impact the forensic science community by discussing a definitive diagnosis of acute congestive heart failure with dilated cardiomyopathy after pancarditis was made, as a fatal and rare complication of Systemic Lupus Erythematosus.

SLE is an inflammatory, autoimmune disease of unknown etiology, characterized by the production of autoantibodies and the deposition of immune complexes in various organs. Cardiac involvement occurs frequently, although it is often mild enough not to cause clinical concern. Pericarditis is most commonly seen, with a reported prevalence of 60%. Myocardial involvement is present in only a minority of patients and valvular abnormalities can be demonstrated in an increasing number of patients. Although most of the valvular lesions will be present without any symptoms, valve incompetence can result in congestive heart failure. Myocardial involvement usually accompanies other cardiac lesions. Isolated myocarditis, or dilated cardiomyopathy, is a rare and usually late clinical manifestation of SLE. Autopsy series in diagnosed SLE patients showed 62% pericardial involvement, 50% valvular involvement (Libman-Sacks lesions and infective endocarditis) and 40% myocarditis, but all have been underdiagnosed clinically.

A 28-year-old Caucasian man, with systemic lupus erythematosus (SLE) treated with hydroxychloroquine and systemic glucocorticoids, was admitted to the emergency department for an arm-ache after an accidental fall. Admission radiographs revealed a spiroid diaphyseal humeral fracture at the mid-distal third, which was treated by surgical internal fixation with a locked antegrade intramedullary nail, and then it was replaced by an external fixation. An ECG showed sinus bradycardia (58/min), QRS axial left deviation in the frontal plane, incomplete right bundle branch block, marked ST-T segment elevation.

After few days he was discharged to continue anticoagulant and antibiotic therapy at home, but three days later he was admitted again to the same hospital for high fever (39.5–40.5°C). The clinical examination revealed pharyngeal hyperaemia, cervical lymphadenopathy and the classical “butterfly” erythematosus rash on the face and on the neck. Hematologic studies revealed anaemia, neutropenia, lymphopenia and thrombocytopenia; the morphological examination of peripheral blood and the research for viruses with cardiac and lung tropism were negative. On the eighth day the diagnosis of reactivation of SLE was made and higher doses of glucocorticoid, antipyretic, and antibiotic therapy were administered.

On the fourteenth day, an echocardiography was performed showing normal atrioventricular and semilunar valves, the ventricles were dilated and hypocontractile, with a 33% ejection fraction; the Doppler examination revealed the mitral valve regurgitation. He was transferred to the Department of Cardiology but few hours later he suddenly collapsed; blood gas analysis revealed metabolic acidosis. Vasoactive drugs (dopamine and noradrenaline), bicarbonate, and fluids were administered. The next morning he collapsed again but cardiopulmonary resuscitation was unsuccessful and the man was pronounced dead.

A postmortem examination was performed 48 hours after death. The external examination revealed only malar erythematosus cutaneous rash. Internal examination was unremarkable except for heavy lungs and reddish colored foam on trachea and the main bronchi and a cerebral edema.

The heart had a normal shape (15x13x5cm) and a weight of 495g. The left ventricular wall thickness was 1.9cm and the right ventricular wall thickness was 0.8cm. The atrial chambers were normal, the ventricules ones were dilated, and the myocardium was flaccid. Cross sectioning of extramural coronary arteries showed no significant stenosis or thrombotic occlusion. The atrioventricular and semilunar valves were normal except for mitral valve, which showed abnormal leaflet thickening with a decreased mobility.

The histological examination of the heart was performed using haematoxylin-eosin (H&E) and revealed pericardial spots (lymphocytic infiltrates); the myocardium showed focal and rare lymphocytic infiltration in perivascular areas, patchy fibrosis, rare foci of irreversible hypercontraction with myofibrillar break and anomalous cross band formation, and focal interstitial hemorrhages in subendocardial layers (reflow areas). The mitral cusps showed diffuse fibrosis and lymphocytic infiltrates.

The immunohistochemical examination of the heart specimens revealed a positive reaction in cardiac myocytes for antibodies anti-TNF- α and IL-8, and a stronger positive reaction for antibodies anti-IL-15 and IL-10.

Furthermore, the expression of CD-4 and CD-8 showed a strong positive reaction in pericardium, valvular endocardium, and less positive in myocardial specimens.

Examination of the other organs was unremarkable except for cytotoxic cerebral edema, massive

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pulmonary edema and polyvisceral stasis.

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Lupus, Pancarditis, Dilated Cardiomyopathy

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