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THU0600 A CASE OF SYSTEMIC SCLEROSIS COMPLICATED BY RENAL CRISIS: POTENTIAL ETIOPATHOGENETIC ROLE OF CYTOMEGALOVIRUS AND TREATMENT

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Background: Scleroderma renal crisis (SRC) is a rare complication of systemic sclerosis (SSc), which can be triggered by viruses, such as Cytomegalovirus (CMV). SRC presents as a new-onset accelerated-phase hypertension with/without rapidly progressive renal failure.
Objectives: Here we describe the case of a patient developing SSc complicated by the appearance of SRC after a recent episode of acute Cytomegalovirus infection.
Methods: A 66-year-old male was referred to our Scleroderma Unit in March 2019. He presented with widespread skin rash, exertional dyspnoea and peripheral oedemas. He reported a myocarditis due to CMV occurred in October 2018. Antibodies anti-CMV IgM were detected in his serum. The patient developed a progressive cutaneous involvement characterized by diffuse oedema, sclerosis and melanoderma. Subsequently, Raynaud's phenomenon, puffy hands and pitting scars occurred. Laboratory tests showed positive ANA in a titer of 1:640 in a nucleolar staining pattern. Additionally, persistence of anti-CMV IgM was found. Skin biopsy showed scleroderma-like finding. Nailfold capillaroscopy revealed a SSc pattern. Chest high resolution computed tomography displayed basal interstitial thickening and subpleuric ground-glass opacities. Therefore, the patient was diagnosed with SSc. Three weeks later he developed severe hypertension and a rapid, progressive renal impairment. Serum creatinine increased (up to 4.15 mg/dl), glomerular filtration rate impaired (25 ml/min). Renal biopsy (picture A, B) revealed acute thrombotic microangiopathy. A diagnosis of thrombotic thrombocytopenic purpura was excluded. The patient was diagnosed with SRC and we started therapy with ACE-inhibitor and loop diuretic. Even if the dosage of ACE-inhibitor was increased up to the maximum tolerate dose, his renal function did not improve and the blood pressure control was inadequate. Consequently, the patient underwent plasma exchange (PEx) sessions. Two weeks later there was an improvement of renal function and blood pressure normalized. Six months later the disease was controlled: glomerular filtration rate was 41 ml/min and blood pressure was within the normal range. The patient was treated with ACE-inhibitor and underwent fortnightly apheretic sessions. Treatment for scleroderma vasculopathy is ongoing.
Results: Viral infections may be responsible for SSc. A brief interval between an acute viral infection and the onset of SSc may suggest CMV as a possible trigger for the disease. Similarly, other infectious agents could be involved in the multistep and multifactorial mechanism of SSc. This case sheds light on the potential and intriguing role of CMV in SSc. Moreover, it leads us to hypothesize a CMV possible direct role in sclerodermal kidney damage. Use of ACE-inhibitor significantly reduced the mortality rate due to this complication. Exact therapeutic mechanism of PEx in the treatment of SSc is unclear.
Conclusion: In our case the integrated ACE-inhibitor-PEx approach has showed effectiveness and safety in the management of SRC.
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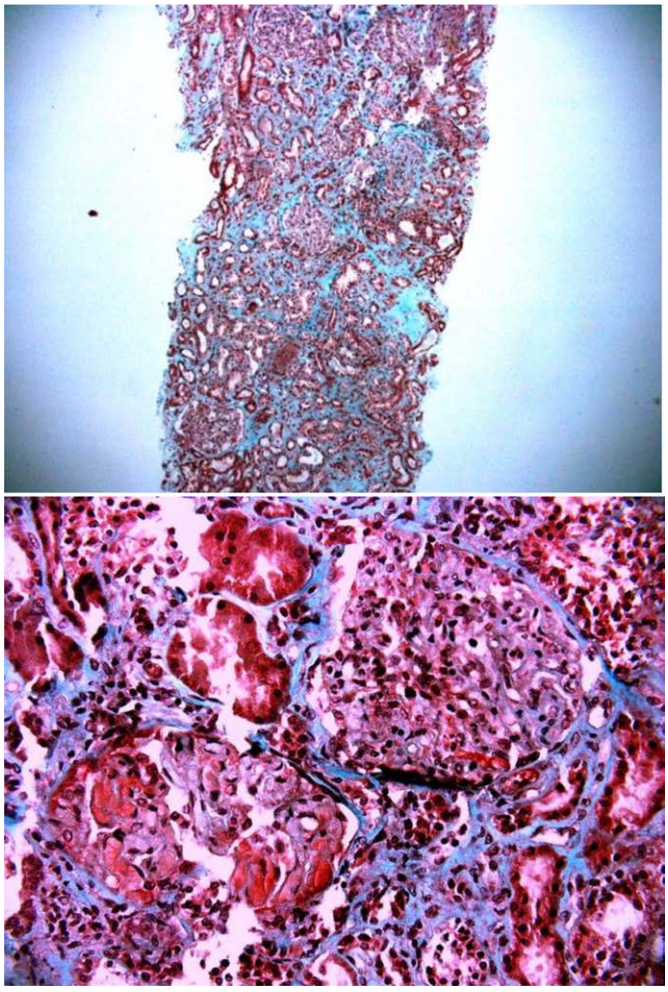

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THU0601 REFRACTORY ACUTE CORONARY SYNDROME AND CARDIOGENIC SHOCK AS ATYPICAL ONSET OF EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS

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Background: Eosinophilic Granulomatosis with Polyangiitis (EGPA) is an ANCA-associated vasculitis, characterized by eosinophilic infiltration in tissues, intravascular and extravascular granuloma formation. It is a rare disease, affecting between 0 and 4 per million population per year. The cardiac involvement occurs in 15-60% of EGPA patients (generally ANCA-negative); it is often insidious, underestimated and it has a poor prognosis. The disease usually shows a prodromal phase characterized by asthma and allergic manifestations.
Objectives: We report the case of a young patient with acute coronary syndrome (ACS) complicated by cardiogenic shock as the first manifestation of EGPA.
Methods: A 36 year old Indian male patient, with a previous history of asthma, rhinitis, Raynaud syndrome and allergy to ketoprofen, presented to the emergency department with a complaint of chest pain and dyspnea. Cardiac troponin was elevated and he was admitted to intensive care unit with a diagnosis of ACS. The patient conditions rapidly deteriorated due to acute cardiogenic shock and an urgent coronary angiogram was performed. An occlusion of the left main coronary artery was treated with angioplasty and two drug-eluting stents. Echocardiography showed severe left ventricular dysfunction requiring inotropic and intra-aortic balloon pump support. A mild dermatitis after salicylic acid administration resolved with intravenous hydrocortisone 1 g. The thrombophilia