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VESSEL DISEASES

278 Cardiac involvement in a 23 years old patient with granulomatosis with polyangiitis (GPA)

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Aims: Granulomatosis with polyangiitis (GPA) is a systemic necrotizing vasculitis, which could potentially affect any organ system. However, there have only been a few reports on cardiac involvement. In fact, it most commonly involves the sinuses, lungs, and kidneys with necrotizing granulomatous vasculitis. In 12% of a large series of patients with GPA there was cardiac involvement, largely manifested by pericarditis and coronary arteritis.
Methods and results: We describe a rare case of a 23-year-old girl, with no pathological history, at exception of a recent flu-like syndrome for which she carried out the search for SARS-CoV-2 RNA through nasopharyngeal swab, results negative. After a month, she went to the emergency department for a syncopal episode and subsequent head trauma. On this occasion, echocardiogram performed showed the presence of left ventricular systolic dysfunction due to hypokinesia of the middle distal segments; CT angiography of the chest revealed the presence of pulmonary embolism. For this reason, the patient was admitted to the cardiac intensive care unit, where EKG shown anterolateral myocardial infarction with ST elevation and immediately was performed coronary angiography, that evidenced two-vessel disease, with subsequent ineffective attempt to angioplasty. Due to the intercurrent appearance of hyposthenia and paraesthesia in the left upper limb, CT angiography of the brain was performed with detection of lower right pre central frontal hypodensity, suspected for recent ischaemic lesion and hypodensity of the right carotid artery as recent thrombosis. In light of the multi-organ involvement of ischaemic nature and the young age of the patient, rheumatological evaluation was carried out, with execution of a laboratory tests that showed the presence of positivity for ANCA anti-PR3 antibodies, on the basis of which was diagnosed GPA, and rituximab therapy was immediately initiated, with clinical benefit.
Conclusions: Cardiac involvement of GPA was first reported by Wegener in 1936. Classical or generalized GPA is characterized by necrotizing granulomatous vasculitis of the upper and lower respiratory tract together with glomerulonephritis. Widespread disseminated vasculitis involving both small arteries and veins occurs to a greater or lesser degree as the disease progresses. A localized form of GPA limited primarily to the upper and lower respiratory tracts has been described. Despite histopathological diagnosis of GPA, with autoantibodies against to circulatory neutrophilic cytoplasmic antigens, we can diagnose GPA easily and early. GPA must be kept in mind as the differential diagnosis of new onset cardiomyopathy, especially in the existence of pulmonary and renal pathologies. The clinical presentation of GPA can be so diverse that the list of differential diagnoses is vast, ranging from infections (fungal, bacterial, and mycobacterial) to other vasculitides, including Henoch–Schönlein purpura, sarcoidosis, Behcet syndrome, and malignancies. Despite that involving the heart is well described, significant cardiac complications occurring during the course of the disease are rare.