Eosinophilic granulomatosis with polyangiitis, a new recurrent feature in an extremely rare disease

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Introduction: Eosinophilic granulomatosis with polyangiitis (EGPA) is an extremely rare necrotizing vasculitis affecting small- and medium-sized vessels. EGPA may affect the heart leading to myocardial inflammation and necrosis. Although, only a few cardiological based studies have been conducted.

Purpose: This study aimed to investigate the prevalence and clinical impact of cardiac-specific involvement (CSI+) and to give an update on EGPA cardiological manifestations.

Methods: This is a single-centre study. Cardiological evaluation included ECG, blood test, echocardiography, global longitudinal strain (GLS), cardiac magnetic resonance (CMR).

Results: We prospectively enrolled 52 consecutive EGPA patients, between October 2018 and October 2019, mean age 59±3 years, 30 (57%) female. We identified 13 (25%) CSI+ patients: 6 myocarditis, 2 pericarditis, 1 coronaritis, 1 Prinzmetal angina, 2 LV apical thrombosis, 1 unexplained wall motions abnormalities (WMA) in the absence of coronary artery disease.

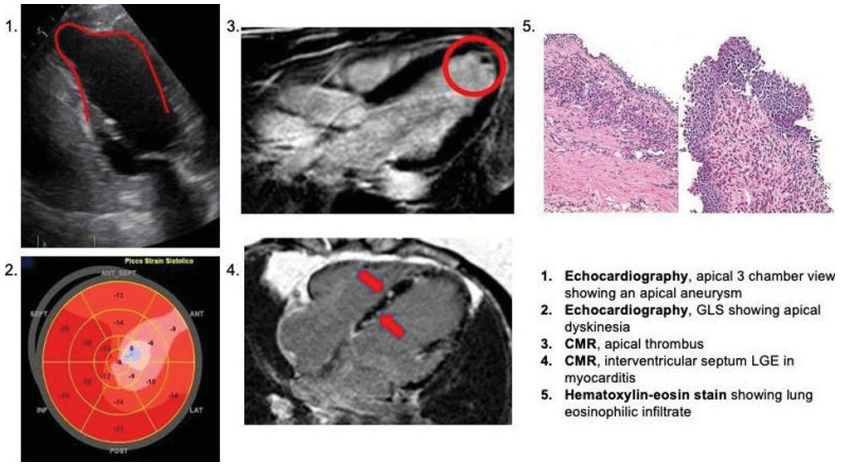
Twelve-leads ECG revealed abnormalities in 11 (85%) CSI+ vs 9 (23%) CSI–, p=0.0001; ECG abnormalities identified CSI+ with 85% sensitivity, 77% specificity, 94% negative predictive value.

Median troponin level in CSI+ 9 ng/L (IQR 6–11) vs CSI– 11 ng/L (IQR 6–25), p=0.2548; NT-pro-BNP value in CSI+ 210 pg/L (IQR 175–484) vs CSI– 159 ng/L (IQR 66–299), p=0.0576.

Echocardiographic left ventricular end diastolic volume in CSI+ 62±4 ml/m^2 vs CSI– 52±1 ml/m^2, p=0.0116; LV ejection fraction in CSI+ 57%±2 vs CSI– 66%±1, p=0.0002. In CSI+ patients GLS was –15±1 vs CSI– GLS –21±0.4, p<0.0001. Echocardiography identified WMA in 8 (61%) CSI+ vs 1 (3%) CSI–. In 7 (54%) CSI+ patients, apical segments showed WMA and among them 5 demonstrated the presence of apical aneurysm.

Twelve patients underwent CMR, it showed non ischaemic late gadolinium enhancement in 7 patients; in 5 patients we identified akinetic or dyskinetic segments without associated LGE. Two patients showed LV apical thrombus.

Conclusions: EGPA showed a high prevalence of CSI. We repeatedly found a tropism for apical involvement in WMA that often manifested themselves as apical aneurysm. These may be a new clinical feature, previously not described, in such a rare condition.



Cardiac involvement EGPA

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