Methods: CMR findings in 40 patients (mean age 45.43±14.7 years; male:female ratio 1:1) diagnosed with endomyocardial biopsy-proven VNLM at our Centre from January 2015 to December 2017, were retrospectively evaluated. CMR was performed at time of clinical presentation and before EMR. At contrast-enhanced CMR we analysed: ventricular function, T2-weighted sequences, short inversion time inversion recovery sequences (STIR), early (EGE) and delayed gadolinium enhancement (LGE). Demographic, clinical and histological data were correlated with CRM findings.

Results: Mean left ventricular (LV) ejection fraction (EF) was 52.5%±10.8%. 50% of patients had reduced LVEF (<55%). Mean right ventricle EF was 55.25%±7.2%. Akinetic or hypokinetic areas were detected in 3 (7.5%) and 17 (42.5%) patients, respectively. LV were identified in 39 patients (92.5%). LGE was the most common CMR finding. 20 patients (50%) met one LLC, 16 patients (40%) 2 LLCs. 1 patient met all 3 LLCs. STIR abnormalities were identified in the LV lateral wall in 8 (19%), septal wall in 9 (21.4%), LV inferior wall in 9 (21.4%), in the apex in 3 (7.1%), LV posterior wall in 3 (7.1%) and LV anterior wall in 2 (4.8%) patients.

Myocardial oedema on T2 images was detectable in 21 (52.5%), EGE in 1 (2.5%) and LGE in 36 (90%) patients. LV was intramural in 22 (55%), subendocardial in 23 (57.5%) and subendocardial in 4 (10%) patients. Multiple LGE patterns were disclosed in 10 (25%) patients. Pericardial effusion was detected in 9 patients (22.5%), 2 patients did not meet any LLC at CMR. When comparing CMR findings with Holter-ECG tape, clinical presentation, histology, biochemistry and autoimmunity results we found CRM oedema positively correlated with the detection of active myocarditis on EMB (11 vs 4, p=0.027). Serum anti-heart antibodies positively correlated with the detection of pericarditis at CMR (3 vs 1, p=0.050). Patchy/intramural LGE positively correlated with rhythm abnormalities at Holter-ECG tape (12 vs 6, p=0.037).

Conclusions: VNLM is an overlooked disease defined by EMB established histological, immunological and immune-histochemical criteria. Although the diagnostic power of CMR needs to be further investigated, it seems to correlate with EMB results and arrhythmic burden.

REFERENCE:

Disclosure of Interest: None declared


Abstract AB1136 – Table 1. Summary of the clinical characteristics of our patients.

Abstract AB1136 – Table 2. Comparison of with an Italian cohort of adult patients with PFAPA syndrome

Conclusions: PFAPA syndrome is one of the main causes of adults Chinese patients with recurrent fever of unknown origin and might be misdiagnosed. Clinicians, not only paediatricians, should take into account this clinical entity, especially for patients with pharyngitis, cervical adenopathy and aphthous stomatitis.

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Abstract AB1137 – Table 1. Clinical features of adult Chinese patients with periodic fever, aphthous stomatitis, pharyngitis, and adenitis: report from an single centre

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