AB1023  CARDIAC MRI IN HYPERFERRITINAEMIC DISEASE STATES REVEALS MYOCARDIAL INFLAMMATION NOT IDENTIFIED BY ECHOCARDIOGRAPHY

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Background: Acutely unwell adult patients with hyperferritinaemic disease states are typically challenging to diagnose. Case series suggest that cardiac involvement may be common (up to 20%) but the phenotype has not been well characterised.

The elevation of cardiac biomarkers suggests cardiac involvement, but are non-specific in acute illness. Cardiac MRI (CMR) offers the ability to characterise the myocardium and identify inflammation, and modern motion-corrected sequences now allow the assessment of patients who may struggle to breath-hold in the recovery from acute illness.

Objectives: We report 3 patients who underwent CMR in the acute phase of illness with raised cardiac biomarkers.

Methods: Case records of acutely ill patients with hyperferritinaemia from two major London centres were reviewed and cases who had undergone CMR in the acute phase of illness were identified.

Results: 3 cases were identified from a cohort of 22. We report CMR findings from differing aetiologies of hyperferritinaemic states:

Case 1: A female in her 60s presented acutely unwell with fever, swollen joints and pink rash. Ferritin was raised at 50544/L (20-300L); troponin I 384ng/L (<34ng/L) and Brain Natriuretic Peptide (BNP) 324ng/L (<159ng/L). Echocardiography was normal. However CMR with T2 mapping revealed several small areas of raised signal consistent with myocardial inflammation. A diagnosis of systemic Adult Onset Stills Disease (AOSD) was made. She received IV methylprednisolone and anakinra with normalisation of cardiac biomarkers.

Case 2: A male in his 20s with known SLE was unwell due to end stage renal failure requiring transplant. He had a previous prolonged admission secondary to HLH. He presented with chest pain and concave shaped ST elevation on ECG. Troponin peak 2168ng/L, BNP 1334ng/L. Peak ferritin 1300ug/L. He responded to colchicine with improved troponin, and was discharged with close follow up.

Case 3: A male in his 20s presented with septic shock attributed to meningococcal septicaemia requiring ITU admission. Troponin was elevated at >9000ng/L. Bloods demonstrated raised ferritin and features consistent with HLH were identified. CMR reported elevated native myocardial T1/T2 signal of the lateral and mid-anterior walls in keeping with myocardial oedema. Pericardium adjacent to the anterolateral wall had elevated T1/T2 signal with hypenhancement on delayed enhancement imaging. Tissue characterisation was in keeping with an acute myopericarditis process.

In addition to broad spectrum antibiotics to treat his underlying infection, he received therapy for HLH including methylprednisolone, anakinra and IVIG. He subsequently made a good recovery to treatment.

Conclusion: CMR in acute illness with hyperferritinaemia reveals abnormal tissue characterisation with myocardial inflammation, even when echocardiography is normal. We suggest CMR may be a useful test to expand our understanding of hyperferritinaemic disease states.

References:

Disclosure of Interests: None declared

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AB1024  EVALUATION OF CLINICAL FEATURES IN PATIENTS DIAGNOSED WITH JUVENILE AND ADULT-ONSET FAMILIAL MEDITERRANEAN FEVER

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Background: Familial Mediterranean Fever (FMF), which is more common in groups in the Mediterranean basin, is a monogenic auto inflammatory disease characterized by recurrent attacks of febrile peritonitis, pleuritis and arthritis.

Objectives: The aim of this study is to investigate the clinical features of patients diagnosed with juvenile and adult-onset Familial Mediterranean Fever (FMF).

Methods: Patients with FMF were included in the study consecutively without sample collection. Data about age, sex, disease duration (month), symptom duration, age at diagnosis, diagnosis delay time, comorbid diseases, and medications were noted. Patients with onset of symptoms ≤ 20 years old were classified as juvenile-onset, those > 20 years old were classified as adult-onset FMF. The frequency and characteristics of attacks and the presence of amyloidosis will be recorded.

Results: The mean age of 86 patients (63 female, 23 male) with FMF was 38.38±11.6 years. The patients with juvenile-onset FMF were 26.7% of the patients.