21. SEPSIS
Hannah Zacharias1, and Tony Chan1
1Rheumatology, Royal Berkshire Hospital, Reading, United Kingdom

Introduction: Sepsis is a major cause of mortality and there have been many patient safety initiatives to improve the outcome such as ‘Think Sepsis’ and ‘Sepsis 6’. It is essential to consider mimics of sepsis in those presenting with fever, particularly when there is no improvement despite antimicrobials. Macrophage activation syndrome (MAS) is a complication of uncontrolled inflammation from underlying connective tissue disease resulting in a sepsis-like condition. Delay to the detection and treatment of this condition can also result in significant morbidity and mortality.

Case description: We report a 28-year-old female presenting with shortness of breath, chest pain, bilateral leg swelling and fevers. Relevant past medical history included a recent diagnosis of Graves’ disease for which she had been commenced on carbimazole. On presentation CRP 53 mg/L and d-dimer 14100, chest X-ray; cardiomegally and bilateral pleural effusions, CT thoracic aorta angiogram; large pericardial effusion with maximum depth 5.9cm and compression of the cardiac chambers. Echocardiogram; preserved ejection fraction of 55–60%. The patient was commenced on intravenous antibiotics, furosemide and pericardial drain removed 1.5L.

The patient was referred to the rheumatology team on day 10. She reported episodic arthralgia, recurrent mouth ulcers, anaemia; with no evidence of haemolysis and pericardial effusion (serositis). Serology: ANA 1:1280 fine speckled pattern, dsDNA > 200, ENA positive (RNP and Smith antibodies) and low complement; C3 (0.35) and C4 (0.005), ferritin 1400, CRP 40 and ESR 62; consistent with a diagnosis of systemic lupus erythematosus (SLE). The patient was pulsed with IV methylprednisolone 500mg on Day 13 and started on prednisolone 40mg and azathioprine.

Day 14; the patient developed a new pyrexia and profound hypotension resulting in admission to intensive care unit for inotropic support and escalation of antibiotics. Bedside echo revealed a small volume pericardial fluid; no tamponade, but significant new biventricular failure. Repeat ferritin was 8489, Triglycerides 5.5, LDH 331. The patient had persistent fevers and blood dyscrasias consistent with macrophage activation syndrome (MAS) with lupus myocarditis. The patient was pulsed with a further two doses of IV methylprednisolone 750mg and commenced on ciclosporin 3mg/kg.

Day 18; the patient became increasingly tachycardic with increasing oxygen demands and a worsening metabolic acidosis despite renal replacement therapy. Based on the rapidity of deterioration, and lack of response to intervention the patient was transferred for extracorporeal membrane oxygenation (ECMO).

Discussion: This case was of interest due to the diagnostic difficulties the team faced as an underlying autoimmune condition was not initially considered. Distinguishing the need for antibiotics versus immunosuppression can be daunting for general medical teams. This case highlights the importance of involving rheumatology early in the course of a patient presenting with fever not responding to conventional therapies.

Lupus myocarditis was confirmed by cardiac biopsy after the patient went into cardiogenic shock with a LVEF 10% due to lupus myocarditis requiring ECMO support showing the severity of disease activity. The use of ECMO remains rare in our practice and there are only five centres within the UK offering this service.

The patient had made an excellent response to treatment and is well maintained on mycophenolate, methotrexate and hydroxychloroquine; most recent echocardiogram May 2018 shows resolved effusion with normal biventricular size, ejection fraction 50 to 55%.

Key learning points: MAS secondary to underlying autoimmune conditions can present with fever and features that mimic sepsis. An awareness of MAS is needed as early diagnosis and prompt initial treatment are both key factors for a favourable outcome. This case resulted in a change to clinical practice. A Trust-wide guideline on the management of MAS has now been produced to guide teams on the management of MAS in the patient with sepsis who is not improving. This case also shows the importance of collaborative team working (physicians, ICU, tertiary centre).

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