**MACROPHAGE ACTIVATION SYNDROME IN CHILDHOOD AUTOIMMUNE DISEASE; A RARE COMPLICATION AND A DIAGNOSTIC DILEMMA**

Jayasekara NP1, Herath HMSY1,Dissanayake S1, Dandeniya C1, Abeygunawardhana AS1, Thalgahagoda RS1

1.Teaching Hospital Peradeniya

Introduction

Macrophage Activation Syndrome(MAS) is a rare complication of childhood autoimmune disease(cAD). The overlapping nature of MAS and disease activation in cAD, together with the added possibility of sepsis, pose a diagnostic dilemma with therapeutic challenges.

Case presentation

Case 1 - A 14-year-old girl with systemic lupus erythematosus and Class-2 lupus nephritis, since 10-years of age, presented with fever, reduced appetite and arthralgia of 5 days duration. She was ill looking, febrile and pale. Moderate splenomegaly was noted without hepatomegaly and lymphadenopathy. Investigations revealed, pancytopenia (WBC 1000/mm3;Haemoglobin 8.6g/dl;Platelet 61000/mm3) with ESR 25mm/hr, CRP 0.6mg/dl, lactate-dehydrogenase 2736U/L, Ferritin of 65956ng/ml, Triglycerides 424mg/dl, AST 358U/L, ALT 144U/L, UFR protein 2+, occasional red cells, Serum-Creatinine 52micmol/l. Haemophagocytosis on bone marrow biopsy confirmed MAS. There was no evidence of a significant renal flare.

Case 2 - An 11-year-old girl with undifferentiated autoimmune disease since 4 years of age, with a history of anti-NMDAR encephalitis, pyrexia of unknown origin, autoimmune haemolytic anemia and sub-nephrotic range proteinuria presented with high grade fever for 3 days. Examination revealed cervical lymphadenopathy and hepatosplenomegaly. Investigations revealed, WBC 5000/mm3, haemoglobin 10.3g/dl, platelets 150000/mm3, ESR dropped from 70mm/hr to 25mm/hr, CRP 13mg/dl, Lactate-dehydrogenase 435U/L, Ferritin 3100ng/ml, Triglyceride 309ml/dl, Fibrinogen 1.6g/L, AST 47U/L;ALT 68U/L, UFR- normal, Serum-Creatinine 44micmol/l. Haemophagocytosis on bone marrow biopsy confirmed MAS.

Both of these patients had a remarkable recovery following steroids.

Conclusion

The diagnosis of MAS in the presence of cAD is challenging. Early detection and prompt treatment are the key to reducing morbidity and mortality.