A rare and serious cause of pancytopenia in a patient with systemic lupus erythematosus: haemophagocytic lymphohistiocytosis

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DESCRIPTION

A 60-year-old woman with a history of hypertension, rhupus (systemic lupus erythematosus (SLE) and rheumatoid arthritis), anemia who presented to an outside hospital with generalized weakness, worsening anaemia, recurrent vomiting and had a complicated medical course including septic shock due to *Escherichia coli* bacteraemia with acute renal failure, acute liver injury, acute respiratory failure and pancytopenia. Patient was receiving continuous renal replacement therapy, mechanical ventilation and immunosuppression with intravenous methylprednisolone and azathioprine. Patient was weaned from mechanical ventilation and transferred to our hospital for further evaluation and treatment. Her labs were consistent with pancytopenia, positive antidouble stranded DNA, anti-SSA/RO, anti-SSB/LA, elevated serum creatinine and elevated aspartate aminotransferase, alkaline phosphatase, reduced serum complement (C3 and C4) levels and extreme hyperferritinaemia (ferritin of 60 800) concerning for haemophagocytic lymphohistiocytosis (HLH). Further labs revealed an elevated interleukin two soluble receptor and elevated triglyceride levels. Bone marrow biopsy and aspirate were consistent with haemophagocytosis as displayed in figure 1. Patient was started on a standard protocol of therapy that included ciclosporin, etoposide and dexamethasone. This was followed by clinical and biochemical improvement in patient's overall condition, however, the patient made the decision to transition to comfort care. HLH is known as a relatively rare complication in autoimmune diseases, particularly,

SLE.¹ Mortality rates may exceed 20%, according to some reports.² Older age, the presence of infection, maximum ferritin elevation and elevated CRP are associated with increased mortality.^{3–5} There are no validated treatment protocols, current protocols are largely based on retrospective case series and case reports.⁶

Learning points

- ▶ It is important to maintain a high index of suspicion for a timely diagnosis and earlier treatment.
- ▶ Bone marrow biopsy is usually diagnostic of haemophagocytic lymphohistiocytosis (HLH), but haemophagocytosis is a late feature of HLH.
- ▶ There are no validated treatment protocols for HLH, current protocols are largely based on retrospective case series and case reports.

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Figure 1 A bone marrow aspirate revealing phagocytosis by histiocytes of erythrocytes, leucocytes, platelets and their precursors.

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