Systemic lupus erythematosus presenting to haematology with pancytopenia and features of macrophage activation syndrome

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DESCRIPTION

This bone marrow biopsy (figure 1) shows haemophagocytosis consistent with macrophage activation syndrome (MAS) secondary to previously undiagnosed systemic lupus erythematosus (SLE).

A 44-year-old woman had been unwell for 4 weeks with fever, weight loss and an aphthous ulcer. There were no other clinical features of SLE.

There was pancytopenia (platelet count 65×10^9/L; neutrophil count 0.5×10^9/L; haemoglobin 107 g/L). The reticulocyte count was 20×10^9/L. Parvovirus and Epstein-Barr virus IgM were not detected.

A very high ferritin level of 3717 µg/L in the context of cytopenias was suggestive of MAS, a life-threatening hyperinflammatory state.1 Clinical features of MAS include fever, lymphadenopathy and hepatosplenomegaly. Laboratory markers include pancytopenia, altered liver function and coagulopathy, with raised lactate dehydrogenase (LDH) and triglycerides.2

This patient had raised aspartate transaminase and alanine transaminase levels (154 and 145 U/L, respectively). The LDH level was high (458 U/L). The triglyceride level was 2.07 mmol/L. There was no coagulopathy. The C-reactive protein level was 3.2 mg/L. There was a raised urine protein to creatinine ratio (>200).

Antinuclear antibody titres were raised (1/160–640). The anti-double-stranded DNA level was high at >379 IU/mL, which alongside ulceration, cytopenias and proteinuria yielded a diagnosis of SLE. Complement levels were low (C3 0.3 g/L and C4 0.1 g/L), indicating active disease. The cytopenias and clinical features responded well to prednisolone.

While the cytopenias may have been autoimmune in aetiology, the high ferritin level and bone marrow biopsy findings make MAS a likely contributing factor.

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Competing interests None declared.

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