Background: We present a case of pure red cell aplasia (PRCA) secondary to adult-onset Still's disease (AOSD) to raise awareness of this rare complication. To our knowledge, only seven cases have been reported. RA and SLE have established associations with PRCA, but only anecdotal evidence exists linking PRCA with AOSD. Early recognition of this rare, but life-threatening complication may prevent delay in diagnosis and successful treatment.

Methods: N/A.

Results: A 24-year-old female with no previous medical history presented to the Manchester Royal Infirmary in June 2014 with recurrent high fevers, widespread arthralgia, leucocytosis and an intermittent salmon-pink skin rash consistent with AOSD, according to the Yamaguchi criteria. Her serum ferritin was 8000 μg/l. ANA and RF were negative and her haemoglobin was 130 g/dl. She initially responded well to regular NSAIDs. One month later she was readmitted under general medicine with collapse. Her haemoglobin was very low, at 23 g/l, with a mean corpuscular volume of 70 fl. There was no obvious gastrointestinal blood loss. Her CRP was 155 mg/l and serum lactate dehydrogenase was moderately elevated at 868 IU/l. Her reticulocyte count was low (0.5%). On examination, she was tachycardic, with moderate hepatosplenomegaly and synovitis of the right ankle and left knee.

Her infective screen was negative for hepatitis B, cytomegalovirus and HIV. Her bone marrow biopsy showed an absence of mature erythroblasts with normal white cells and platelet maturation, consistent with PRCA. She required a transfusion of three units of red cells and was given pulsed methylprednisolone 500 mg for 3 days, followed by prednisolone 30 mg daily. She was then established on ciclosporin therapy. Her haemoglobin subsequently improved without any further episodes of PRCA.

Conclusion: Haemophagocytic syndrome/macrophage activation syndrome was a close differential, particularly with the very high serum ferritin levels. However, PRCA was the more likely diagnosis, given the bone marrow biopsy findings, the very low haemoglobin and the low reticulocyte count. PRCA differs from aplastic anemia in that production of white cells and platelets is normal. It is also important to exclude other causes of red cell aplasia such as leukaemia, and infections such as HIV and parvovirus. High-dose steroid therapy is the mainstay of treatment. There is anecdotal evidence to support the use of immunosuppressants such as ciclosporin, AZA, CYC and rituxumab.

Disclosure statement: The authors have declared no conflicts of interest.