Introduction: Systemic onset juvenile idiopathic arthritis (JIA) is an often severe and potentially life-threatening subtype of chronic arthritis starting under the age of 16 years. It is characterised by fevers, rash and arthritis, although this may not be apparent until later in the disease course, which can add difficulty to making the diagnosis. We present a case of systemic onset JIA with multi-organ impairment to showcase its mimicry of other diseases and highlight importance of awareness.

Case description: A 15-year-old girl presented with a 7 day history of fever, cough, sore neck, enlarged lymph nodes and widespread macular rash. She had been started on penicillin in the community for presumed scarlet fever, but was hospitalised due to worsening symptoms. Differential diagnoses discussed were lymphadenitis and EBV, due to ongoing fevers, high inflammatory markers, and raised liver function tests. She then developed an acute kidney injury and profound hypotension. She was treated with immunoglobulin for possible Kawasaki disease, broad spectrum antibiotics for possible toxic shock syndrome/sepsis, and transferred to the regional PICU for ongoing inotropic support and management of her multi-system illness. She was found to have a non-dilated cardiomyopathy with poor left ventricular function, low complement and positive anti-streptolysin titer. No positive cultures or focus of infection were found. Antibody tests including dsDNA were negative. She required CPAP for respiratory support and was treated with pulsed methylprednisolone for systemic onset JIA. She then received a weaning course of prednisolone over several months and made an excellent recovery.

Discussion: This was an interesting case for those involved because the initial symptoms, evolving signs and investigation results didn’t easily tie together to give one suitable diagnosis. Several avenues of treatment were explored, including a range of antibiotics and immunoglobulin, before methylprednisolone. The patient also remained remarkably ‘well’ looking despite her multi-organ impairment.

Key learning points: This case highlighted the importance of awareness of systemic onset JIA and its complications, particularly in a non-typical age group, for the general paediatrician. Early discussion with and involvement of tertiary rheumatology services served to optimise management and outcome, and will hopefully also improve long-term prognosis.

Conflicts of interest: The authors have declared no conflicts of interest.