

Systemic-onset juvenile idiopathic arthritis and incomplete Kawasaki disease may belong to a single clinical syndrome within a spectrum of severity

Sirs,

We read with great interest the case report by Keskindemirci *et al.* (1) regarding systemic-onset juvenile idiopathic arthritis (SoJIA) with macrophage activation syndrome (MAS) and coronary artery lesions (CALs) misdiagnosed as incomplete Kawasaki disease (KD). SoJIA is sometimes confused with incomplete KD because both diseases have overlapping clinical features and can be accompanied with CALs and/or MAS (1-3). Therefore, both KD and SoJIA should be considered in the differential diagnosis of prolonged fever, rash and lymphadenopathy (1). Here, we wish to make some comments by presenting our own experience.

A 19-month-old girl was transferred to our paediatric rheumatology unit due to KD refractory to intravenous immunoglobulin. Although there were no definite abnormalities on echocardiography, she was diagnosed with incomplete KD on the basis of the clinical and laboratory findings (4). After pulse steroid therapy, her fever subsided and C-reactive protein value began to decline. However, after one week of afebrile period, fever recurred and her laboratory data worsened: platelet count, 68,000/mm³; aspartate transaminase, 964 IU/L; alanine transaminase, 1,099 IU/L; ferritin, 57,100 ng/mL; triglyceride, 272 mg/dL;

and fibrinogen 175 mg/dL. Bone marrow aspiration revealed haemophagocytosis. She was diagnosed with KD with MAS, and underwent MAS therapy with dexamethasone and cyclosporine for 8 weeks (5). Her clinical and laboratory findings were improved and there was no recurrence during therapy. Four weeks after terminating MAS therapy, she was hospitalized again with fever, rash and arthritis. At that point she was finally diagnosed with SoJIA with MAS.

Initially we also thought that our patient had been misdiagnosed with KD with MAS, as Keskindemirci *et al.* (1) did. However, considering the current diagnostic criteria, both our and their patient should be diagnosed with KD with MAS (4). With their genetic susceptibility to SoJIA, both patients exhibited clinical manifestations of SoJIA with MAS (1). Otherwise, they would have presented with KD with MAS. In other words, recurrent, longer-duration inflammation will be observed in a patient predisposed to SoJIA, while single, shorter-duration inflammation will be observed in those predisposed to KD. Differentiating SoJIA from incomplete KD is based on the chronicity of the disease. Given the large degree of clinical, laboratory, and therapeutic overlap between the two diseases, SoJIA and incomplete KD may be considered a single clinical syndrome within a spectrum of severity. In practice, both diseases share common phenotypes of severe inflammation, such as CALs or MAS. Our hypothesis may help explain the distinctive characteristics of SoJIA and incomplete KD compared to other JIAs and typical KD (4, 6).

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