Systemic-onset juvenile idiopathic arthritis or incomplete Kawasaki disease: a diagnostic challenge

Sirs,

As being part of a tertiary paediatric rheumatology centre in Turkey, we read with great interest the article by Dong *et al*. (1) concerning the incidence and presentation differences between patients with presumed Kawasaki disease (KD) with subsequent diagnosis of systemic onset JIA (SoJIA) and sole KD. In the light of our two similar, cases we aimed to recap our experience about this issue.

Both patients (first case: 9 month-old girl, second: 20 month-old-boy) were referred us with development of macrophage activation syndrome after incomplete KD, despite treatment with IVIG. While initial symptoms in both were fever for two weeks and evanescent rash, pericardial effusion and hepatosplenomegaly were detected only in the first case. Pancytopenia and fever, namely MAS improved with concomitant use of IV bolus prednisone and cyclosporine A. However, the girl re-admitted at six months with long lasting fever and rash and the boy had developed second flare with fever and polyarthritis when he was 4-year-old. After these flares anti-interleukin (IL)-1(anakinra or canakunimab) therapy was introduced into therapy with a great success in both patients with a flare free period of 4 and 12 months, respectively.

In conclusion, this challenging problem is experienced by other paediatric rheumatologists (2-4), as well. While IL-18 has recently been proposed as a useful marker in differentiating SoJIA from incomplete KD (4), the diagnosis is still clinical. However, there are some hints:

1. Flares are mostly related to SoJIA.
2. While fever and rash in KD are continuous, fever in SoJIA is intermittent and rashes waxes with fever (3).
3. While the child looks almost normal during the afebrile period of SoJIA, patients look sick and agitated all time in KD.

We further suggest the use of anti-IL-1 agents in both diseases which, according to our experience, brings about an excellent control of flare-ups.

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