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Life-saving treatment with the use of anakinra in severe relapse of Still disease with perimyocarditis and aseptic pneumonia

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A 27-year-old man with a history of systemic Juvenile Rheumatoid Arthritis (sJIA), remaining in remission for 13 years (Pouchot score of 0), presented urgently to the Cardiology Department due to myopericarditis with a typical chest pain, a pericardial effusion on transthoracic echocardiography (Figure 1A), a reduction of the PQ segment on the electrocardiogram and high troponin T (up to 1352 pg/ml, reference range <13 pg/ml). He also reported a sore throat, a fever up to 39°C and arthralgia in shoulders and knees. Laboratory tests revealed severely elevated inflammatory parameters with a ferritin up to 13641 ng/ml (reference range: 30–400 ng/ml) and leukocytosis up to 22 x^3/ul (reference range: 4.26–10.57x10’3/ul) with 95% neutrophils and a neutrophil/lymphocyte ratio of 32.

Rheumatoid factor, antinuclear antibodies and anti-neutrophil cytoplasmic antibody tests were negative. Viral infections and other triggers were excluded. Based on Yamaguchi criteria (patient fulfilled all minor criteria and 3 major criteria besides rash) a suspicion of Still disease was raised by the consulting rheumatologist. Despite intravenous administration of methylprednisolone (1100 mg) the patient's condition deteriorated with a sudden dyspnea, desaturation, fever (up to 41ºC) and a pink maculopapular rash (Figure 1B), with a computed tomography (CT) image of left-sided pneumonia and left pleural effusion (Figure 1C). The patient was urgently transferred to a Pulmonology Department, where a non-infectious lung involvement was confirmed by thoracentesis and bronchoscopy. After readmission to Rheumatology Department a treatment with high doses of intravenous methylprednisolone pulses (cumulative 3 g), followed by oral prednisone 60 mg daily and methotrexate 15 mg per week orally were introduced. After initial temporary improvement, the patient's condition deteriorated with reapparition of fever, sore throat, skin changes and rising ferritin level. CT scan revealed a pleural, pericardial and peritoneal effusion, splenomegaly and right hilar lymphadenopathy (Figure 1D). The macrophage activation syndrome and leukemia were excluded by bone marrow biopsy. Paroxysms of atrial flutter followed by bradycardia were observed. Echocardiography revealed reduced left ventricular global longitudinal strain (Figure 1E) and cardiac magnetic resonance imaging showed enhancement of the thickened pericardium (Figure 1F). Life-saving therapy with anakinra (an interleukin-1 antagonist) 100 mg daily was administered, with a quick resolving of fever, pulmonary and skin changes, with a normalisation of the inflammatory parameters and ferritin level. The patient was discharged from the hospital on a maintenance treatment with anakinra, methotrexate and tapered prednisone and remains asymptomatic at the 3 months follow-up.

Nowadays sJIA and Adult Onset Still disease are considered to be a continuum of Still disease [1]. This rare (1–10 per million annually) autoinflammatory disorder may differ in its clinical

manifestation and course – from mild to life-threatening [2]. Our case of glucocorticoid-resistant flare of Still disease with a multi-organ involvement shows that a failure to respond to the traditional treatment in the Still disease should raise suspicion of its potentially life-threatening course and encourage implementation of biological treatment without delay. From the diagnostic point of view, it shows also that Still disease should be taken into consideration in the initial workup of myopericarditis.

Article information

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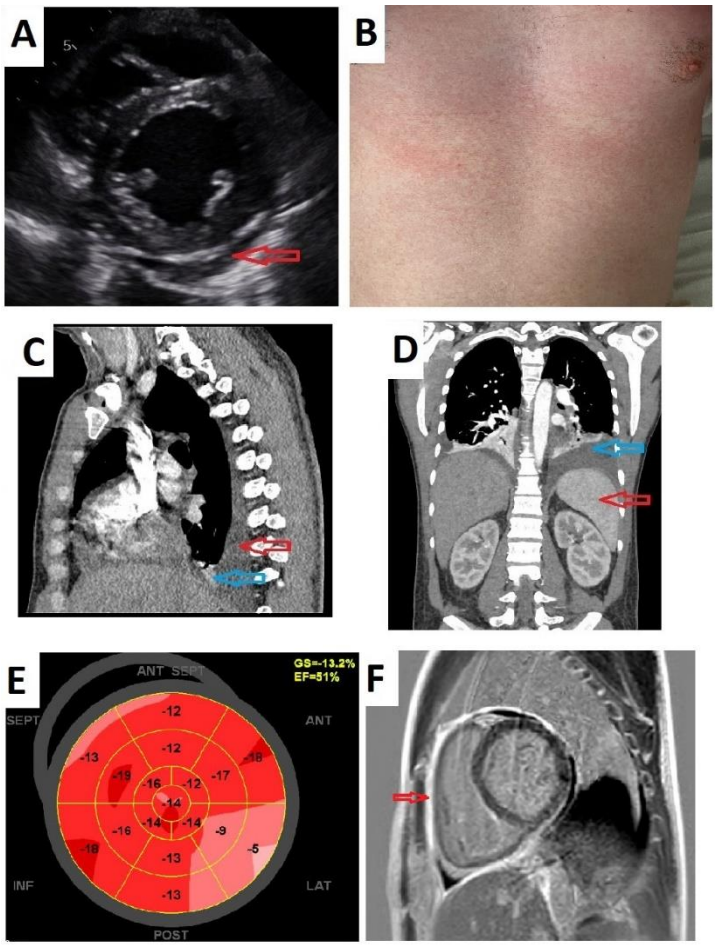

Figure 1 A – Transthoracic echocardiogram showing the pericardial effusion (red arrow); **B** – photograph showing the pink maculopapular rash on the patient's chest; **C** – computed tomography angiography of the chest showing left-sided pneumonia (blue arrow) and left pleural effusion (red arrow); **D** – computed tomography of the abdomen showing splenomegaly (red arrow) and left pleural effusion (blue arrow); **E** – transthoracic echocardiogram showing the reduced left ventricular global longitudinal strain; **F** – magnetic resonance imaging of the heart showing the enhancement of the thickened pericardium (red arrow)

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