Recurrent lupus pericarditis treated with anakinra – a case report

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To the Editor,

Pericarditis is the most common cardiac involvement in systemic lupus erythematosus (SLE)[1, 2]. It usually presents in patients with other forms of SLE serositis, namely pleuritis and peritonitis, and as part of multisystem disease activity[2,3]. Possible complications include recurrence, pericardial tamponade and constrictive pericarditis[4-6]. Non-steroidal anti-inflammatories, colchicine and corticosteroids are commonly used in pericarditis, with overall good efficacy[2,3,7]. Immunomodulator therapies are often necessary in severe or refractory cases, or to control multisystem disease activity[3,7].

We report the case of a 32-year-old female patient diagnosed with SLE for six years with cumulative symmetric polyarthritis, malar rash, photosensitivity, anemia, leucopenia, pleural effusion, class III lupus nephritis, ANA positivity with 1:1280 titer, and homogeneous pattern on immunofluorescence, hypocomplementemia, and positive anti-dsDNA antibody), undergoing prednisolone, hydroxychloroquine, and mycophenolate. Our patient was admitted for acute sharp chest pain that worsened with deep inspiration and in the supine position. There were no symptoms suggestive of respiratory infection (namely fever, cough, sputum, or recent viral condition), inflammatory parameters were only slightly altered (erythrocyte sedimentation rate 24 mm/1st h and C-reactive protein 9.1 mg/L), and troponin I was 0.001 ng/ml. Electrocardiogram (EKG) revealed diffuse ST-segment elevation in precordial leads. Transthoracic echocardiogram showed no signs of effusion or altered contraction. Further laboratory tests revealed elevated anti-dsDNA titer and C3 and C4 consumption. Acute pericarditis was diagnosed and therapy with high dose acetylsalicylic acid (1500mg/day) and prednisolone 30mg/day was initiated, with complete pain resolution in the next three days.

One year later, she was admitted for recurrent acute pericarditis and treated with high dose acetylsalicylic acid, colchicine, and prednisolone, with clinical improvement in a few days. Additional immunomodulator therapy with rituximab was then initiated to prevent new episodes. After the second rituximab cycle (fourth infusion), the patient was newly admitted twice in 3 months for acute pericarditis, when tapering the prednisolone dose below 10mg/daily. Serial echocardiograms performed were all unremarkable. A literature review prompted switching from rituximab to anakinra. Two years after commencing anakinra, no more

episodes of acute pericarditis were observed and serial follow-up monitoring with EKG and echocardiogram consistently revealed normal results.

Anakinra is an interleukin-1 receptor antagonist used in the treatment of idiopathic recurrent pericarditis; its use is based on the observation that pericarditis, in some diseases, may be an autoinflammatory process[2]. However, in other secondary forms of pericarditis, including SLE-associated, data are scarce[8,9]. Shaukat *et al.*, described the use of anakinra in 44 patients (only two with SLE) with non-idiopathic recurrent pericarditis intolerant or refractory to colchicine and corticosteroids, with improvement in symptom relief during hospitalization and decreased recurrence risk.8 Similar findings were observed in a larger cohort, including 224 patients (21 patients with autoimmune diseases including SLE, no data was shown regarding individual autoimmune disease diagnosis).9 Both studies found anakinra use safe in the patients included[8,9].

In our literature review, we found only one case reporting anakinra efficacy in a SLE patient with recurrent pericarditis[10]. Interestingly, the patient had fever accompanying the remaining pericarditis symptoms, which led to the hypothesis that IL-1 could be driving the inflammatory process[10]. Fever was not present in our patient. The fact that there are only two cases depicting anakinra efficacy in SLE pericarditis makes these cases exceptional Further studies are needed to shed light on the pathogenesis of recurrent pericarditis associated with SLE, and the potential benefit of anakinra in different subsets of patients.

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