

Case Report

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Transformation of Rheumatoid Arthritis to Adult Still's Disease

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Abstract

Adult Onset Still's Disease (AOSD) commonly affects young people. We observed an elderly woman with long standing seronegative rheumatoid arthritis and further transformation to systemic illness. A 74 year old woman was admitted to emergency room with multiple joints pain and rash, associated with a fever 40.5°C. The patient had a long standing rheumatoid arthritis presented as symmetric polyarthritis and treated with prednisone only. Multiple immunosuppressive and biological medications failed and she developed several severe bacteremia events. Her past ferritin levels were normal, except for a single elevation two years before.

Keywords: Adult Onset Still's Disease; Multiple immunosuppressive; Anti-cyclic citrullinated peptide

Case Report

On admission she was confused with a body itching rash. Breath sounds were absent in lower lung regions. Symmetric synovitis of elbows, wrists, small joints of the hands, and knees was revealed. Hematological analysis showed leukocytosis $19.3 \times 10^9/L$ (92% neutrophils), elevated liver enzymes (ALT 69 U/L, AST 70 U/L), elevated C-reactive protein (248 mg/L). There were extremely high levels of serum ferritin (56348 ng/ml). Anti-cyclic citrullinated peptide, anti nuclear antibodies and rheumatoid factor were negative. Repeated blood and urine cultures and serology were negative for pathogens. Hilar, axilar and iliac lymphadenopathy and bilateral pleural effusion were observed on CT examination. Echocardiography revealed no vegetations. Renal functions were normal, but with proteinuria 1.1 g/24 hours without active sediment. Bone marrow and bone biopsy were negative for malignancy, infection and macrophage activation syndrome (MAC). The patient developed a clinical picture with four major criteria of Yamaguchi (fever, arthritis, rash, leukocytosis) and three minor criteria (lymphadenopathy, liver enzyme elevation, negative RF, ANA), which are compatible with diagnosis of AOSD (Figure 1) [1]. She was started on methylprednisolone 100 mg IV/day followed by severe exacerbation of the itching rash. After two days of therapy with dexamethasone 16 mg/day IV the patient was still febrile (40°C) and confused. Because patient's history of recurrent severe bacteremia, along with supportive literature about the efficacy of IVIG

in refractory cases of AOSD [2,3] and our experience showing only very high dose of corticosteroid therapy may be helpful for aggressive AOSD, we decided to start dexamethasone 16 mg \times 3/day IV along with intravenous immunoglobulines 25 g/day for 5 days. Of note, IG level before therapy was within normal range with mildly elevated IGA (428 mg/dl, N=70-350). Two days later confusion state resolved, fever and rash disappeared, pain and swelling of joints diminished, and the patient become cooperative and oriented. Ferritin level taken one week after therapy decreased to 651 ng/ml (Figure 1). We are planning a maintenance therapy with IVIG 30 g/day for 4 days monthly for 6 months along with gradual reduction of corticosteroid dose. Alternative therapy is also considering: anti-IL-6 (Actemra-Tocilizumab). The patient was discharged with prednisone 60 mg/day. In conclusion, we described an unusual case and its features were as follow:

1. Elderly onset of AOSD
2. Transformation of rheumatoid arthritis into systemic disease: AOSD
3. Extremely high ferritin level 56348 ng/ml
4. Itching rash not in line with usual evanescent rash described in AOSD. It may be explained by unusually severe intra-dermal inflammation with massive perivascular mononuclear infiltrates of helper-T cells [3].
5. Exacerbation of the rash after methylprednisolone injection. It is still an unclear phenomenon.
6. Non-nephrotic proteinuria and bilateral pleuritis, showing internal organ risk due to aggressive AOSD.

References

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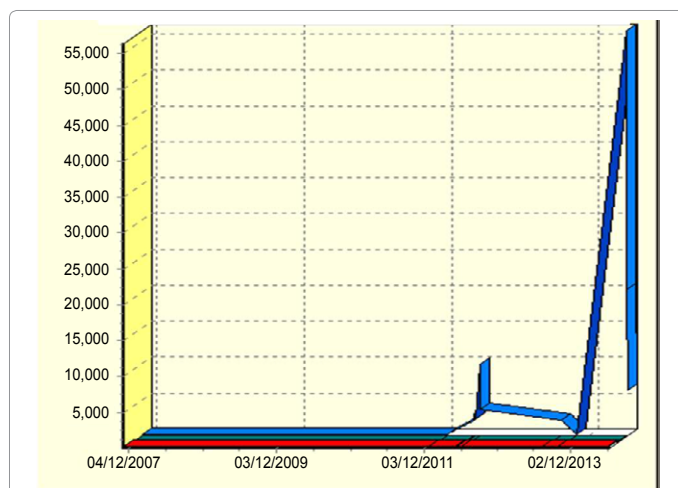


Figure 1: Dynamic of ferritin in patient with Still's transformation of rheumatoid polyarthritis.

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