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Is there any familial predisposition of adult onset still disease with spondyloarthritis? (predisposition of still disease with spondyloarthritis)

Abstract

We have recently observed two Adult Onset Still Disease patients whose first degree relatives have sponyloarthritis with marked axial involvement. One of the patient's father has Ankylosing Spondylitis with marked axial involvement and second patient has two sons who have psoriatic arthritis.There is few data exist about the association of Still Disease and Sponyloarthritis.Our aim is to take attention to these two entities.

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Introduction

Adult Onset Still Disease (AOSD) is one of the rare diseases and %15 of this rare disease is complicated with macrophage activation syndrome (MAS).[1] We have recently observed two AOSD patients (One of them is complicated with MAS) whose first degree relatives have sponyloarthritis with marked axial involvement. Adult Onset Still Disease with MAS patient's father has Ankylosing Spondylitis (AS) with marked axial involvement and second AOSD patient has two sons who have psoriatic arthritis. One of the sons has marked axial involvement and he is under anti-TNF treatment. In fact it is not exactly known if there is any association between these two entities according to the literature. Here, we present two cases with AOSD whose first degree relatives have spondyloarthritis. We want to take attention to the familial predisposition of two diseases.

Case 1

Eighteen years old male patient admitted to the hospital with fever, sore throat, cough, fatigue, weight loss with transient macular-hyperemic lesions. The lesions was dissolving concurrently with the normalization of body temperature. During the differential diagnosis procedure, antibiotherapy with seftriaxon and vancomycin were started empirically with the suggestion of infectious disease specialist. We could not reach any clinical nor laboratory achievement with antibiotherapy. Leucocyte count 23.900/µl with neutrophil 21100/µl Erythrocyte sedimentation rate (ESR): 84mm/hour, CRP:192mg/dL, procalcitonin: 34.09ng/mL ferritin:166956ng/mL.Hepato-splenomegaly was also detected. During the follow up marked anemia and thyrombocytopenia were observed and bone marrow aspiration and biopsy was performed.The phagocytized cells in the bone-marrow were seen on the stained aspiration material. At the same time liver function tests raised very high levels (AST:1226IU/L ALT:716IU/L). Adult Onset Still Disease complicated with macrophage activation syndrome was diagnosed. The treatment was as follows IVIG (0.4gr/kg/day) 5 days, pulse steroid therapy (1000mg/day) 3 days. A complete clinical and laboratory remission has achieved. When we checked the family medical history we have noticed that his father has ankylosing spondylitis (AS) phenotype and have learned that he had diagnosed AS years ago.

Case 2

Seventy-six years old women additted to the hospital fever, sore throat, macular rash on her trunk, knee arthritis, and ferritin levels >2000ng/mL CRP:221mg/dL, ESR:46mm/hour AST/ALT:122/39IU/L .She has also asthma and parkinson's disease with birth defects of cleft lip. She was diagnosed as AOSD and have reached remission with glucocorticoid therapy (1mg/kg/day). When we checked the family history we learned that she has two sons with psoriatic arthritis. One of them has been receiving infliximab therapy for marked axial involvement for years. The other son has been newly diagnosed psoriatik arthritis with bilaterally ankle involvement. The co-existence of AOSD with spondyloarthritis (SpA) was shown before with case series and the authors suggested that patients with AOSD should be also investigated for the typical features of SpA but there are just very few cases exist in the literature.[2,3] Sacroiliitis and SpA also can be seen with an another auto-inflammatory disease Familial Mediterranean fever (FMF).[4] Adult Onset Still Disease tought to be multigenic autoinflamatory disease and affect both of the immun system arms (innate and adaptive).[1,2] Spondyloarthritis are associated with HLA-B27 and in contrast to other rheumatic diseases, affect the both gender equally and there is no defined specific autoantibody. It is proposed that the SpA should be one of the mixed pattern diseases between autoimmune and autoinflamatory diseases.[5] Both of our patient's relatives have marked phenotypical axial involvement with anatomical changes and bilaterally grade IV sacroiliitis. One of the first degree relatives is ankylosing spondylitis and the other one is psoriatic arhritis with prominently axial involvement. The occurence of two entities among first degree relatives may show the need of further studies to lighten the autoinflamatory part and innate arm immunopathology interactions for sponyloarthropaties.

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Conflict of interest

The author declares that there is no conflict of interest.

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