A Case of Adult Onset Still's Disease in a Patient with Ankylosing Spondylitis

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Introduction: Adult Onset Still's Disease (AOSD) is a rare auto-inflammatory disease. AOSD is a disease of exclusion: diagnosis is possible only when there is no evidence of infections, malignancies, and connective tissue diseases. The Ankylosing spondylitis (AS) is an inflammatory joint disease characterized by inflammation and stiffness of the joints and spine, but it isn`t accompanied by high fever as an extra-cutaneous symptom. There are a few cases reported that AOSD combined with AS. Here we report the case of a 45-year-old man diagnosed as AOSD combined with AS.

Case presentation: A 45-year old male with no underlying disease presented fever, pharyngitis, and oligoarthritis of both knees. He had received antibiotic treatment, but fever sustained, a salmon-colored macular rash appeared. Laboratory data demonstrated that WBC 19.32K with neutrophil predominance 86.9%. C-reactive protein 152 mg/L and procalcitonin 0.2 ng/mL. Liver enzymes were elevated, and ferritin was above 2000.0 ng/mL. Cultures from blood, throat, urine, and synovial fluid from both knees were all negative. Computed tomography showed multiple enlarged lymph nodes in mediastinum and splenomegaly. Blood protozoa examination, Dengue PCR, and Antinuclear Antibody (ANA) titer were all negative. The test of Anti-cyclic citrullinated peptide antibodies and Rheumatoid factor (RF) were within normal limits. According to the Yamaguchi criteria of AOSD, he met all major criteria: arthralgia, fever above 39℃, salmon-colored rash, leukocytosis, and he met all minor criteria: sore throat, lymphadenopathy, splenomegaly, abnormal liver function test, negative tests for RF and ANA. He also complained the symptoms of inflammatory back pain. Bone scan showed increased uptake in both sacroiliac joints, and the sequential study revealed bilateral sacroiliitis (Figure 1) and positive for the human leukocyte antigen B27assay, which met the diagnostic criteria of AS.

Conclusion: Only one case of AOSD combined with AS was reported in Korea. This report remarks our patient meets the typical diagnostic criteria for AOSD and AS, including HLA-B27. A pathophysiologic link between AOSD and AS should be considered.

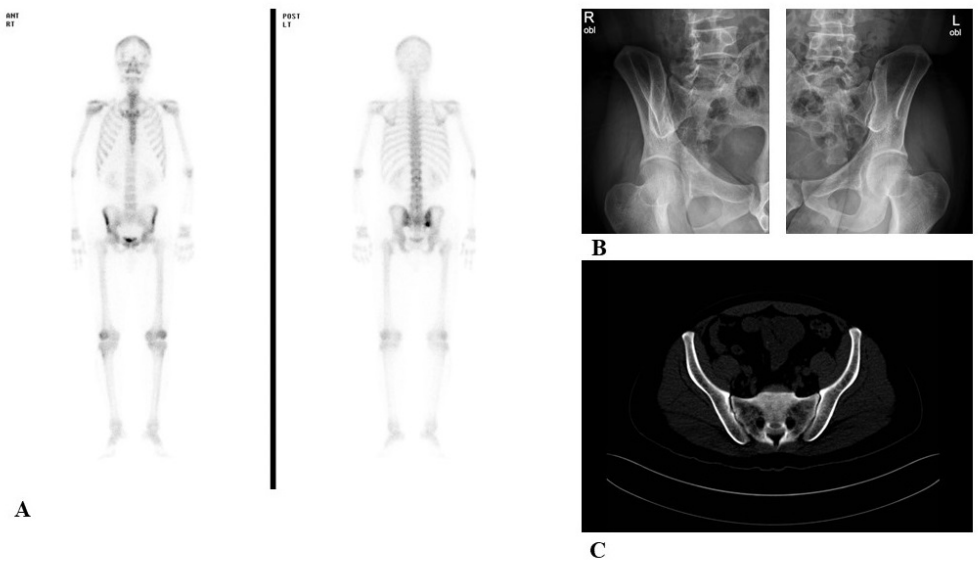

Figure 1. (A) The whole body bone scintigraphy demonstrated the increased uptake in both sacroiliac (SI) joints. (B-C) An anteroposterior radiograph (B) and computed tomography (C) of the pelvis revealed the bone sclerosis and bone erosion of the right SI joint (Grade 3) and irregularity of the articular surface with sclerosis of the left SI joint (Grade 2).