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Case Report

# Two Edges of Immune Dysregulation: Adult Onset Still's Disease in a Patient with a History of Kaposi Sarcoma

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**Abstract:**

We describe a very unique case of emergence AOSD in a patient with a history of KS. A 52-years old woman presented with fever, arthralgia, sore throat and skin rash that started 1 year ago. Patient's medical history was noteworthy because 1.5 years ago she was diagnosed with Kaposi's sarcoma where she presented with a low extremity rash and swollen lymph nodes. Lymph node biopsy demonstrated HHV-8 positivity and atypical vascular proliferation and atypical endothelial cells that were interpreted as Kaposi sarcoma. She receieved paclitaxel chemotherapy and with this treatment Kaposi sarcoma was in remission. Current physical examination revealed a fever of 38.5º C, salmon colored rash in anterior chest wall, pharyngeal erythema, cervical and axillary lymphadenopathy and polyarthralgia in wrists, ankles and small joints of hand and feet. Laboratory examinations revealed neutrophilic leukocytosis, anemia, a very high ferritin level (16815 mcg/l) and elevated acute phase reactants. Rheumatoid factor and antinuclear antibody were negative. Infections and mlaignancies were ruled out and patient was diagnosed with adult onset Still's disease. Patient responded dramatically to the combination treatment of 2*16 mg methylprednisolone and 15 mg/week oral methotrexate. Clinical symptoms improved dramatically, acute phase reaactants and ferritin normalized and hemoglobin increased. As soon as the symptoms were controlled, methylprednisolone tapering was initiated and it was eventually stopped. Patients continues to receive methotrexate. She is under follow up for 27 months with both her Kaposi sarcoma and adult onset Still's disease in remission. To our knowledge, this is the first case of AOSD that developed in a patient with KS history. The patient whose immune system initially succumbed to opportunistic pathogen HHV-8 and suffered from KS received chemotherapy. Once KS was in remission, a different form of immune dysregulation- this time immune system hyperactivation, with a predominant innate immune system activation occurred and she suffered form AOSD.

**Key words:** adult onset still's disease; immunosuppression; kaposi sarcoma

## Introduction

Kaposi sarcoma (KS) is a low-grade vascular tumor associated with human herpesvirus 8 (KSHV/HHV8) infection [1]. Adult-onset Still's disease (AOSD) is a multigenic autoinflammatory disease characterized by fever, arthritis, rash, and hyperferritinemia [2]. Herein, we describe a unique case of emergent AOSD in a patient with a history of KS.

## Case

A 52-years old woman presented with fever, arthralgia, sore throat, and skin rash that started 1 year prior to presentation. The patient's medical history was noteworthy because 1.5 years ago she was diagnosed with Kaposi's sarcoma, which presented with a lower extremity rash and swollen lymph nodes. Lymph node biopsy demonstrated HHV-8 positivity and atypical vascular proliferation, and atypical endothelial cells, which were interpreted as Kaposi sarcoma. She received paclitaxel chemotherapy, and the Kaposi sarcoma was in remission. Her symptoms began a few months after the Kaposi treatment was complete. She described an erythematous rash that became more pronounced with fever, spiking fever that reached as high as 39.5°C, inflammatory joint pain that was accompanied by a morning stiffness of at least 30 minutes, and sore throat. Prior to her presentation to the rheumatology outpatient clinic, she

was hospitalized in the infectious disease department, where she underwent extensive clinical, laboratory, and imaging examinations that ruled out infectious pathologies and malignancies. Physical examination revealed a fever of 38.5°C, a salmon-colored rash in the anterior chest wall, pharyngeal erythema, cervical and axillary lymphadenopathy, and polyarthralgia in the wrists, ankles, and small joints of the hands and feet. Laboratory results were as follows: Leukocytes 13.66*103/µL, neutrophil 9.96*103/µL, hemoglobin 86 g/L, platelet 351*103/µL, ferritin 16815 mcg/L, C-reactive protein, 54 mg/L, erythrocyte sedimentation rate, 73 mm/hour, RF-negative, ANA negative, and anti-CCP negative. The patient's creatinine level and liver function test results were normal, and urine analysis was unremarkable. Recent imaging modalities have demonstrated only cervical and axillary lymphadenopathy. The patient fulfilled the Yamaguchi criteria and was diagnosed with adult-onset Still's disease. Before initiating immunosuppressant therapy, consent was obtained from the patient's oncologist. The medical oncologist stated that KS was in remission and immunosuppressants could be administered with close follow-up. Oral methylprednisolone (2 × 16 mg) and methotrexate (15 mg/week) were administered. The patient's fever, rash, sore throat, arthralgia, and morning stiffness dramatically improved by the end of the first month. CRP level regressed to 1 mg/L and the erythrocyte sedimentation rate regressed to 11 mm/hour. Hemoglobin increased to 126 g/L and ferritin regressed to 162 mcg/day in the second month of treatment. At the end of the first month, once the symptoms were adequately controlled, tapering of methylprednisolone was initiated; at the end of 6 months methylprednisolone was discontinued. Currently, the patient receives only 15 mg/day of oral methotrexate. The patient was followed-up at the 27th month of rheumatology. AOSD did not recur and, during the entire follow-up period, there were no signs or symptoms suggestive of Kaposi sarcoma. Her follow-up in the rheumatology and medical oncology departments continues.

## Discussion

The HHV-8 virus alone is not sufficient for KS to emerge, and there needs to be some degree of host immune dysfunction (1). Our patient was HIV negative and did not undergo immunosuppressive treatment before KS diagnosis. This patient was also epidemiologically interesting because KS predominantly affects men (3). The patient contracted KS despite the absence of any trigger for immunosuppression. This case is especially noteworthy because the patient later developed AOSD. To our knowledge, this is the first reported case of AOSD in a patient with a history of KS. A patient whose immune system initially succumbed to the opportunistic pathogen HHV-8 and developed KS received chemotherapy. Once KS was in remission, a different form of immune dysregulation, immune system hyperactivation, with a predominant innate immune system activation, occurred and the patient suffered from AOSD. Steroid treatment and methotrexate were administered cautiously with the consent of the patient's medical oncologist, where we walked in a fine line between adequately suppressing the immune system to control AOSD and trying to avoid excessive immunosuppression to prevent the reemergence of KS. The patient was closely followed up during the treatment period. Once the patient's symptoms were adequately controlled, prednisolone treatment was rapidly tapered off and stopped. However, after 27 months of follow-up, the patient's immune system was under control, with remission of both Kaposi sarcoma and adult-onset Still's disease.

## Informed consent:

Obtained from the patient.

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