A Very di-Still-ed Diagnosis- Adult-Onset Still's Disease Presenting in a Middle-Aged Hispanic Patient

Background: Adult-Onset Still's Disease (AOSD) is a systemic inflammatory disorder characterized by daily high fevers, arthritis, evanescent rash, and leukocytosis (1). Patients can present without typical manifestations and pose a challenging differential. We present a case of a 52-year-old gentleman with a one-year history of recurring fever, lymphadenopathy, and weight loss diagnosed with AOSD. This case highlights the diagnostic challenge that AOSD poses and the strategies to help aid in the diagnosis.

Case Presentation: A 52-year-old gentleman presented to the ED for a 2-week history of fever associated with chills and bone pain. He reported that he has been having intermittent fever, weight loss, night sweats, and rash for the past year with prior workup being unrevealing. He endorsed swollen glands and fatigue but denied productive cough, chest pain, gastrointestinal, urinary, or neurological symptoms. On physical examination, the patient was febrile at 101.9 deg F, tachycardic 121 BPM, and RR 21 br/min. He appeared cachexic, with dry oral mucosa, palpable lymphadenopathy, and bilateral knee tenderness. Laboratories were remarkable for WBC 22.3 th/mm3, hemoglobin 11.1 gm/dL, platelet 513 th/mm3, sedimentation rate 120 mm/h, CRP 23 mg/dL, lactic acid 0.89 mmol/L, ferritin level 28,595.9 ng/mL, and LDH 603 IU/L. Peripheral smear revealed reactive neutrophilic leukocytosis. Infectious etiology, including SARS Covid-19PCR, HIV, blood cultures, lumbar puncture with CSF analysis, and QuantiFERON gold were negative. Autoimmune workup was unrevealing. CT Chest/Abdomen demonstrated moderate pleural effusions and reactive bilateral hilar, mediastinal, and retroperitoneal lymphadenopathy, and hepatomegaly. CT-guided biopsy of the left inguinal lymph node showed benign follicles with mixed B and T cells. Flow cytometry showed increased granulocytes and eosinophils without immunophenotypic abnormalities to suggest hematologic malignancy. After excluding infectious and malignant causes, rheumatology was consulted. Based on symptomatology, laboratory, and radiographic findings, a diagnosis of AOSD was entertained. Yamaguchi's criteria supported the diagnosis with four major and four minor criteria met (2). The patient was started on prednisone 1 mg/kg with excellent response.

Conclusion: AOSD is a diagnosis of exclusion, and the appropriate clinical scenario should warrant further investigation. AOSD should be in the differential after careful workup and excluding infectious etiology, malignancy, and other connective tissue diseases.

Citation:

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3. Efthimiou, Petros et al. "Adult-Onset Still's Disease in Focus: Clinical Manifestations, Diagnosis, Treatment, and Unmet Needs in the Era of Targeted Therapies." Seminars in arthritis and rheumatism 51.4 (2021): 858–874. Web.