Felty's Syndrome: Triad Not Always Evident

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Felty’s Syndrome: Triad Not Always Evident

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Introduction

- Felty’s Syndrome (FS): characterized by neutropenia, splenomegaly, and seropositive rheumatoid arthritis (RA)
- Usually presents in patients with established longstanding RA
- Absolute neutrophil counts below 2000/uL required for diagnosis
- Neutropenia should be persistent and unexplainable by other processes
- Pathogenesis not entirely known
- First line treatment: methotrexate
- Second line: rituximab
- Third line: glucocorticoids

Case Presentation

- 48 year old African American female with two weeks of worsening nausea and vomiting
- Past medical history: DM2, HTN, GERD, obesity
- Pancytopenia w/ neutropenia (absolute neutrophil count 83.6 cells/uL), fever, and splenomegaly (CT abdomen)
- Found to have Staph aureus rhinosinusitis and ESBL UTI
- 10 months of recurrent admissions for neutropenia and fatigue. Extensive workup done during that time.
- Two bone marrows seven months apart: normal cellularity and adequate iron storage
- Flow cytometry: no immunophenotypic evidence of acute leukemia, high-grade myelodysplasia or T-cell lymphoproliferative disorders
- Immunological, serological, tumor markers negative

Discussion

- This case was an unusual presentation of Felty’s Syndrome
- This patient did not have obvious signs and symptoms of longstanding RA
- Key features in medical history aided in diagnosis
- Family history of maternal grandmother and grandmother w/ RA
- Mild joint pains in hands and knees
- Hand x-rays did not suggest degenerative/erosive joints
- Rheumatoid factor titer 1:32, anti-CCP antibody over 300U/mL, anti-histone antibodies 2.5U
- 2010 ACR/EULAR diagnostic criteria for RA met (8 points)
- Patient was started on weekly methotrexate and transferred to outside facility w/ inpatient rheumatology
- This case illustrates the importance of pursuing alternative diagnoses even when symptoms and signs of the primary disease process are not obviously present

References

1. Jakez-Ocampo, Juan MD; Atisha-Fregoso, Yemil MD; Llorente, Luis MD. Pancytopenia as Manifestation of Nonarticular Felty Syndrome. JCR: Journal of Clinical Rheumatology: October 20, 2018 - Volume Publish Ahead of Print - Issue - p