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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 long-standing 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

Images

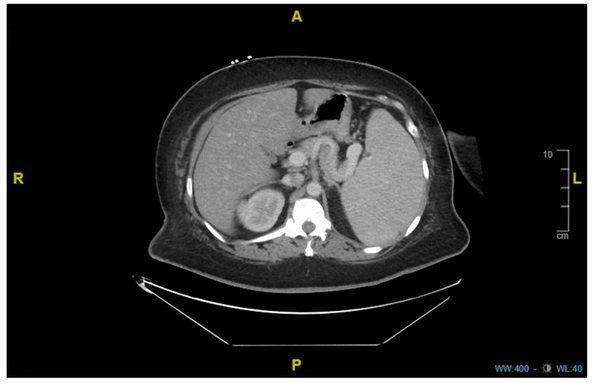

Figure 1: CT abdomen w/o contrast notable for hepatomegaly and splenomegaly (18cm)

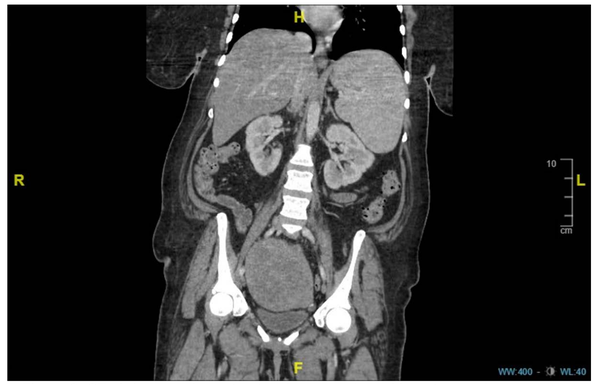

Figure 2: CT abdomen w/o contrast: splenomegaly and hepatomegaly

Test	Result
IgG	1300
IgA	189.0
IgM	90
Immunoelectrophoresis	See Hx
IEP Interpretation	
Rheumatoid Factor Scrn	Positive *
Rheumatoid Factor Titer	1:32 *
Cyclic Citrull Peptide	> 300.00 *
ANA Pattern	Negative
c-ANCA Antibody	< 0.2
Anti-Myeloperoxidase	< 0.2
SS-A/Ro Antibody	< 0.2
SS-B/La Antibody	< 0.2
Sm (Smith) Antibody	< 0.2
SM/RNP Antibody	< 0.2
Histone Antibodies	2.5 *
Anti-Cardiolipin IgG Ab	< 1.6
Anti-Cardiolipin IgA Ab	0.5
Anti-Cardiolipin IgM Ab	1.4
% CD3 Cells	69.4
Absolute CD3 Count	252 *
% CD4 Cells	35.0 *
Absolute CD4 Count	127 *
T-Help/Suppress Ratio	1.7
% CD8 Cells	20
Absolute CD8 Count	73 *

Table 1: Selected lab values

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

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