CASE REPORT

Non-articular Felty's syndrome: An uncommon diagnosis

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ABSTRACT

Felty's syndrome is a triad of rheumatoid arthritis, neutropenia, and splenomegaly. We hereby report an unusual case of non-articular Felty's syndrome and its management along with discussing the importance of appropriately ruling out alternate causes of neutropenia with splenomegaly.

KEYWORDS

Felty's syndrome, rheumatoid arthritis, neutropenia

What was known on this topic?
Felty's syndrome is a triad of rheumatoid arthritis, neutropenia, and splenomegaly.

What does this case add?
We present a case of severe neutropenia and mild splenomegaly in a patient with high titres of rheumatoid factor and anti-CCP with no signs of synovitis. After ruling out alternate causes of neutropenia and splenomegaly, a diagnosis of non-articular Felty's syndrome was made.

INTRODUCTION

Felty's syndrome is a triad of rheumatoid arthritis, neutropenia, and splenomegaly. We report an unusual case of moderately severe asymptomatic neutropenia and splenomegaly with positive serologies (rheumatoid factor and anti-citrullinated protein antibody) but no joint involvement, i.e. non-articular Felty's syndrome.

CASE REPORT

A 73-year-old Caucasian female sought medical attention because of an incidental finding of leukopenia and neutropenia. She had a past medical history of essential hypertension, ischaemic cardiomyopathy with an ejection fraction of 45-50% (status post-implantable cardioverter defibrillator) and ventricular tachycardia on amiodarone. She was found to have leukopenia (1800 cells/µl, normal = 3800-10,600 cells/µl) and neutropenia (700 cells/µl, normal = 1800-7700 cells/µl) on routine blood testing which persisted despite discontinuation of the amiodarone for three to four months. Review of the systems was negative for morning stiffness, joint pain, joint swelling, oral-nasal or genital ulcers, rash or any family history of autoimmune diseases. She was a non-smoker with no alcohol or recreational drug use.

Physical examination was unremarkable except for splenomegaly. There was no obvious synovitis or limited range of motion in any of her joints.

Imaging confirmed splenomegaly (18 cm) with no hepatomegaly or adenopathy and was otherwise unremarkable. She subsequently underwent bone marrow biopsy and multiple peripheral smears which were negative for alternate causes of neutropenia. There was no immunophenotypic evidence of either a mature T or B lymphoid neoplasm in the flow cytometry analysis.

Autoimmune work-up revealed a normal direct anti-globin test, anti-nuclear antibodies, anti-dsDNA antibody, anti-SSA, anti-SSB, C3, C4, c-ANCA, p-ANCA, extractable nuclear antigen antibodies and monoclonal protein evaluation. Serology for syphilis, brucellosis,

infectious mononucleosis and viral hepatitis was negative. Tuberculosis skin test, quantiFERON-TB gold test, human immunodeficiency virus testing and cytomegalovirus polymerase chain reaction were negative. However, rheumatoid factor (180 IU/ml, normal < 15 IU/ml), cyclic citrullinated peptide (CCP) (45 IU/ml, normal < 7 IU/ml) and erythrocyte sedimentation rate (65 mm/h, normal 0-20 mm/h) were significantly elevated. Imaging of wrists, hands, knees, ankles and feet showed no evidence of erosions or joint space narrowing.

A final diagnosis of non-articular Felty's syndrome was proposed. She was started on daily prednisone 40 mg daily with subsequent improvement in the leukocyte (4100 cells/µl) and neutrophil (1700/µl) counts within ten weeks, at which time it was tapered down to 20 mg daily and subsequently to 10 mg daily and finally discontinued.

DISCUSSION

The differential diagnosis in our case vignette included myeloproliferative syndromes, systemic lupus erythematosus, large granular lymphocyte syndrome, tuberculosis and sarcoidosis, all of which were ruled out by appropriate testing.

Rheumatoid arthritis is a chronic inflammatory arthritis with significant extra-articular manifestations. Felty's syndrome is a severe extra-articular feature of rheumatoid arthritis. Felty's syndrome is characterised by the triad of rheumatoid arthritis, neutropenia, and splenomegaly.[1] The lifetime risk of Felty's syndrome for a rheumatoid arthritis patient is less than 1%.[1]

Felty's syndrome usually develops late in rheumatoid arthritis. Arthritis almost always appears first and has typically been present for ten years or more before neutropenia is recognised. In very rare cases, neutropenia appears before or without arthritis.[1]

The articular disease in rheumatoid arthritis associated with Felty's syndrome is usually severe in terms of both erosions and deformity. In some affected individuals, Felty's syndrome may develop during a period when the symptoms and physical findings associated with rheumatoid arthritis have subsided or are not present. In such cases, it may remain undiagnosed. Also, as a result of neutropenia, affected people are increasingly susceptible to certain infections such as *Staphylococcus aureus, Pseudomonas aeruginosa, Escherichia coli, Herpes zoster* and fungi.[2]

Anti-CCP has a very high specificity for rheumatoid arthritis. The combination of rheumatoid factor and anti-CCP has a specificity of 99.5% for rheumatoid arthritis.[3] There is no specific diagnostic test for Felty's syndrome. It is a clinical diagnosis in rheumatoid arthritis with unexplained neutropenia and splenomegaly.

The treatment of neutropenia in Felty's syndrome is mainly comprised of disease-modifying anti-rheumatic drugs including methotrexate, hydroxychloroquine, cyclophosphamide, azathioprine, glucocorticoids, and G-CSF.[4]

CONCLUSION

Manifestations of Felty's syndrome without clinical but only with laboratory features of rheumatoid arthritis are extremely rare. We present a case of severe neutropenia and mild splenomegaly in a patient with high titres of rheumatoid factor and anti-CCP with no signs of synovitis. The current vignette highlights the importance of appropriately ruling out alternate causes of neutropenia with splenomegaly and to recognise an uncommon presentation of Felty's syndrome.

DISCLOSURES

There are no disclosures and no author had any relationship with the industry. The authors declare no conflicts of interest.

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