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Case Report Open Access

Cellulitis in the Presentation of Felty's Syndrome: A Case Report

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Abstract

Felty's syndrome is an uncommon but severe extra-articular manifestation of rheumatoid arthritis. Felty's syndrome is characterized by the triad of rheumatoid arthritis, neutropenia, and splenomegaly. The lifetime risk of Felty's syndrome for a rheumatoid arthritis patient is less than 1%. We present a case which is a classical presentation of Felty's syndrome with the triad of RA, neutropenia and splenomegaly. We present a case of 51-year-old woman on chronic RA treatment who presented with cellulitis. The work up showed splenomegaly and neutropenia which support Felty's syndrome diagnosis. Patients with Rheumatoid arthritis (RA), who developed neutropenia and splenomegaly, should be suspected of developing Felty's syndrome as a complication of Rheumatoid arthritis.

Keywords: Felty's syndrome; Cellulitis; Rheumatoid arthritis

Introduction

This case is a presentation of a unique condition that is not easily identified in regular basis. The patient case was escalated from being a cellulitis patient with rheumatoid arthritis to the diagnosis of rare Felty's Syndrome. The pharmacy intervention with the choice of best antibiotic treatment was a challenge.

We report the case of a 51-year-old female with a known medical history of seronegative rheumatoid arthritis, presents to the ED with a worsening left groin wound that started around 4 days ago after self-administering an Enbrel shot. It started as a small blister, which popped open and later became ulcerated and painful. She also described some pus coming out of the area. She is having a discharge from an abdominal wall wound. Patient states that she is having mandibular pain that she never had before. She complains of pain in the right eye with clear discharge and also redness in the left eye; denies any visual disturbances. She complains of low-grade fever and chills at home. She also complains of pain and swelling on the left chin. She describes her situation as generalized weakness and pain and states that overall she feels sick. She complains of nausea but no vomiting but reports anorexia. Denies any diarrhea, problem urinating, cough, chest pain, or shortness of breath. She also complains of a mild intermittent headache. Patient has chronic polyarticular pain, mostly on her left knee and ankles. She refers on and off swelling even on medication and also states limited head movement that has been that way for a long time. She denies smoking or drinking and family history is noncontributory. Patient also reports allergies to zosyn and sulfa. Past medical and surgical history includes rheumatoid arthritis, shingles, hepatitis C, GERD, status post appendectomy, and knee surgery. Her home medications were Enbrel, methotrexate, prednisolone, tramadol, doxepin, folic acid and omeprazole.

Physical examination shows a chronically ill looking woman, not in acute distress but in discomfort with severe itch. She has a 2 × 1 cm oval wound along left groin, some drainage on the gauze and very minimal cellulitis, Splenomegaly was clearly appreciated. Musculoskeletal evaluation reveals tenderness on bilateral knees and ankles without any swelling or warmth. Bilateral hands with chronic deformities and no deviation as seen with RA patients.

The laboratory was significant for low WBC's (1.4×10^3 cells/μL), and a low hemoglobin and hematocrit (Hgb was 9.5 g/dL, hematocrit 27.6%). The microbiological wound culture positive for Methicillin-Resistant Staphylococcus aureus (MRSA); there was no growth from blood culture.

Assessment

- Rheumatoid arthritis with probable Felty's syndrome
- Pancytopenia, unclear etiology, possibly due to infection versus drug-induced, which includes methotrexate versus underlying rheumatoid arthritis
- Left mandibular pain, poor dentition and chronic steroid use, possible cause of focal infection and abscess
- Leukopenia, possibly secondary to her medications for RA, but still has an ANC of more than 1000 cells/μL
- Cellulitis

Plan

ID was consulted and she was started on IV vancomycin, but started complaining of severe itch and rash all over her body. Her current medications is IV ceftaroline 600 mg IV q12H (switched from vancomycin), tramadol PRN, pantoprazole 40mg daily, famotidine 20 mg BID, folic acid 1mg daily, and prednisone 10 mg daily.

In this case, the patient presented with an ulcerated, infected wound to the left groin wound that started after self-administering an Enbrel shot. Treatment for cellulitis was initiated with IV vancomycin after wound culture showed positive growth for gram methicillin resistant staphylococcus aureus species (MRSA). The patient has significant history of RA. Treatment for MRSA in the setting of leukopenia became limited because patient developed redman's syndrome with vancomycin, the side effect profile of linezolid (neutropenia,

pancytopenia,), synercid (pancytopenia, rash). Daptomycin has a milder hematological side effect profile than the three mentioned above however the infectious disease consultant chose ceftaroline which has the safest side effect profile of all the anti-MRSA agents [1]. In the CANVAS trial comparing the vancomycin with ceftaroline, ceftaroline was non-inferior to vancomycin plus aztreonam in treated patients with ABSSSI caused by gram positive and negative pathogens. Consistent with safety profile of the cephalosporin class, ceftaroline is well tolerated [1].

The case was diagnosed as Felty's syndrome as it has history of rheumatoid arthritis, Splenomegaly, and leukopenia. This limited the treatment choices for her cellulitis and the choices of antibiotics were tailored toward those who did not interfere with the disease state and would not worsen the leukopenia. The only working option was selected of cefaroline without any serious consequences.

Discussion

Rheumatoid arthritis (RA) is a chronic, systemic, inflammatory disorder of unknown etiology that primarily involves joints. RA has significant extra-articular manifestations [2,3]. Felty's Syndrome (FS) is a severe extra-articular feature of RA. FS is characterized by the triad of RA, neutropenia, and splenomegaly. The lifetime risk of FS for a RA patient is less than 1% [3]. Over 95% of FS patients are positive for RF with high titers [2,3]. FS usually develops after a long course of RA [4]. Arthritis almost always appears first and typically has been present for 10 years or more before neutropenia is recognized [5]. The articular disease in FS is usually severe in terms of both erosions and deformity [3,5]. The exact cause of neutropenia in this patient has not been confirmed however her immunosuppressive therapies of MTX and Enbrel are suspected. Leukopenia can be defined as a leukocyte count of less than 3×10^9/L, with an absolute neutrophil count of less than 2.0×10^9/L. There is an increased frequency of bacterial infections in FS. Neutropenia is believed to be the main cause of this increased rate of infection. The FS patients who develop recurrent infections have significantly lower neutrophil counts. Other risk factors for infections include severe disability, a high level of immune complexes, hypocomplementaemia, neuropathy, skin ulcers, syndromes and steroid treatment [3] Most often the infections affect the skin, mouth, and upper and lower respiratory tract [3].

The clues that led to the provisional diagnosis of Felty's syndrome are the patient's history of rheumatoid arthritis, presentation with splenomegaly, and neutropenia. Neutropenia is certainly the most common and most important feature of Felty's syndrome. Treatment options for neutropenia in FS include splenectomy, colony stimulating factors G-CSF, disease modifying anti-rheumatic drugs (DMARDs) and non-steroidal anti-inflammatory drugs NSAIDS [5]. NSAIDS are not favored because of their potency for worsening neutropenia. The goal of treatment is for a hematological response which will result in a permanent rise of ANC 1000 per ml [1,2]. The patient in this case is being evaluated for the best options to treat her symptoms.

DMARDS are considered first line therapy in these patients; however the patient in this case has been on two DMARDs (Methotrexate and Enbrel) as for quite some time, therefore her treatment options will have to be carefully evaluated. Recently, there has been a growing interest in the biologic agent rituximab in the treatment of Felty's syndrome [6,7] however there has not been many reported cases of beneficial treatment of Felty's syndrome with rituximab. Global analysis of all cases of Felty's syndrome only suggests the use of rituximab as second line therapy in all patients with refractory Felty's [7]. The determination of Felty's syndrome in this patient is just suggested, therefore it is safe to conclude that rituximab would not be a preferred agent, but is still an option. Splenectomy which is surgical removal of the spleen may be considered by the patient's physicians. This procedure results in high rates of hematological response (80%), however it is not first line in most patients [1].

Finally, the cellulitis presentation to probable Felty's syndrome was challenged by limited treatment options with antibiotic choices. This case study may serve as atypical situation were infections complicated with other co-morbidities may need further pharmacy intervention and assessment.

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