A Case of Felty’s Syndrome with Delayed Articular Involvement

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Abstract

Felty’s syndrome is mostly characterized by rheumatoid arthritis, neutropenia, and splenomegaly. In rare cases, such as this one, we have patients that present with unexplained neutropenia without any evidence of arthritis. In this case, the patient presented with a history of multiple infections due to his neutropenia. Laboratory investigations revealed low white blood cell count and an elevated anti-cyclic citrullinated peptide antibody and rheumatoid factor. Although the patient had high titers of antibodies for rheumatoid arthritis, the patient denied any signs and symptoms of the active rheumatoid disease. This patient was diagnosed with Felty’s syndrome following laboratory investigations and abdominal imaging studies showing an enlarged spleen. The patient was subsequently started on a triple therapy which is comprised of methotrexate, sulfasalazine, and prednisone. A splenectomy was also done to alleviate the neutropenia. In conclusion, Felty’s syndrome is a medical condition that can be hard to diagnose when it has an atypical presentation such as this case.

Case Presentation

A 69-year-old male presents to his primary care doctor with complaints of redness and pain in his left second toe. A complete review of the system and general examination shows that the patient is in mild distress secondary to pain and discomfort. The patient endorses a low-grade fever. The patient reports decrease in exercise tolerance and slight fatigue and weight gain. Inspection of the upper extremities shows normal metacarpophalangeal and proximal interphalangeal joint movement without nodules or deformities. Evaluation of the lower extremities shows that the second toe on the left has erythema that extends into midfoot and a non-healing wound ulcer. The left lower leg appears to be swollen, and is warm and tender to the touch, suggesting a diagnosis of ascending cellulitis. Previous records show that this is the patient’s fifth visit in the past year for various bacterial and fungal infections, which include a recent hospitalization for acute left foot osteomyelitis and abscess. The patient has a past surgical history of appendectomy and amputation of a second right toe. The patient has a past medical history of sleep apnea, hypothyroidism, hyperlipidemia, morbid obesity and hypertension. All of which have been well controlled with medications and dietary modifications.

Laboratory Investigations

A complete blood count with differential shows the white blood cells (WBC) count of 600 cells/mm² (normal is 3,600 to 11,000) / mm³ down from 3,700 cells/mm² two years ago. With an absolute neutrophils count of 100 cells/mm² and absolute lymphocytes count at 400 cells/mm². The platelet count is 99,000/mm³ (normal is 150-450). The hematocrit is 35.2% (normal is 41-55%) and hemoglobin is slightly decreased at 11.6 g/dl. A complete metabolic panel shows normal glucose level, as well as normal urea and creatinine levels. The Patients also has a normal liver panel with an aspartate aminotransferase (AST) level of 19, and an alanine aminotransferase (ALT) of 20 and Alkaline Phosphatase level of 50.

A hematologic malignancy is suspected due to the extreme decrease in the WBC. The patient is referred to a hematologist oncology specialist for a complete work up. A bone marrow core biopsy demonstrates normocellular bone marrow particles with trilineal hematopoiesis, adequate myelopoiesis and no increase in the blast. The aspirate smears are adequately cellular and they show progressive maturation in all three hematology lineages with no significant dysplasia or maturation. A Fluorescent in situ hybridization (FISH) study is negative for myelodysplasia profile. A flow cytometry report shows no increase in the number of blasts, no B-cell monoclonality and no T-cell Antigenic aberrancy. Peripheral blood smear reveals severe neutropenia, normocytic to macrocytic red cells, mild thrombocytopenia, and mild lymphopenia with heterogeneous morphology. The overall findings of the hematologist specialist are negative for a myeloid or a lymphoid neoplasm, metastatic malignancy or infiltrative processes.

The patient is then referred to a rheumatologist, although he denies any signs and symptoms of rheumatoid arthritis. An Antinuclear antibodies (ANA) panel taken by the rheumatologist shows no specific antibody elevations. However, further investigations show that the patient has a positive rheumatoid factor of 537 (normal is less than 20) and a positive anti-cyclic citrullinated peptide antibody (CCP) greater than 60.

Chest x-ray shows evidence of prior granulomatous disease and is negative for focal consolidation, pneumothorax or pleural effusion. Bilateral X-ray of the hands and wrist is negative for soft tissue swelling and erosions of the joints. A computed tomography (CT) of the abdomen and pelvis with contrast reveals that the spleen is grossly enlarged measuring up to 24 cm × 23 cm × 9.2 cm (Figure 1). The longest dimension of the spleen in a typical adult

Figure 1: Coronal plane of the abdominal CT showing enlarged spleen.
is approximately 11 cm. The CT also shows colonic diverticulosis without evidence of diverticulitis, and a right common iliac artery aneurysm measuring up to 2 cm. The CT is negative for lymphadenopathy. With the positive rheumatoid factor and anti-CCP results along with the severe leukopenia, and the negative hematologic malignancy profile, the patient is diagnosed with Felty’s syndrome.

Management and Outcome

The patient is treated with the triple pharmacotherapy of Methotrexate Sodium 22.5 mg Intramuscular weekly, Sulfasalazine 1,000 mg by mouth twice a day, and Prednisone 10 mg by mouth every night at bedtime. This therapy was chosen as a way to delay the progression of this patient rheumatoid arthritis as much as possible. The patient had his spleen removed in an effort to decrease his abdominal girth and to prevent complications associated with an accidental splenic rupture. The surgical pathology report of the removed spleen shows many hyperplastic germinal centers. The report is negative for intra-splenic lymphoma or malignant neoplasm. A few months after the removal of the spleen the patient showed an increase of WBC from 400 to 1,500 cells/mm³. A five-year follow up after the felty’s syndrome diagnosis, shows that the patient has a successful improvement in his overall clinical picture. The white blood cells levels count range between 3,000 and 4,000 cells/mm³. The patient reported some mild rheumatoid complaints such as early morning stiffness and some intermittent joint discomfort around the fourth year following the felty’s syndrome diagnosis.

Discussion

Felty’s syndrome was first described by Felty himself as a triad of Rheumatoid arthritis (RA), Leucopenia and splenomegaly [1]. This definition had since been revised because of the various initial patient presentations. Felty’s syndrome is now commonly seen as RA plus leucopenia, mostly neutropenia [2]. Neutropenia is the hallmark for the diagnosis. Although an enlarged spleen is often seen in the patients, it is not a requirement for the diagnosis. RA is a chronic inflammatory autoimmune disorder that mostly affects the joints [3]. RA is often treated with disease modifying anti-rheumatic drugs (DMARD). Most of the DMARD agents have leucopenia as a side effect. Another diagnosis challenge is that there is a malignancy of the blood called large granular lymphocyte (LGL) syndrome. The LGL syndrome presents with neutropenia and is associated with RA in 11 to 36% of cases [4]. In order to diagnose Felty’s syndrome, LGL syndrome, as well as DMARD side effects, must be ruled out as potential causes of the neutropenia. The preferred method for ruling those causes out is through negative bone marrow biopsy results. Felty’s syndrome is managed by focusing on treating the RA with the DMARD agents. Patients with severe neutropenia and/or extreme splenomegaly are treated with a splenectomy. Research has shown that removal of the spleen can improve the WBC count by improving the neutrophil count, and can decrease the number of infections [5]. Other reasons to remove the spleen is if the spleen is so large that it starts compressing on nearby organs, causing mechanical obstruction. Also, splenectomy is indicated if it is uncomfortable for the patient and because a large spleen increases the risk of splenic rupture, which can have lethal consequences.

In conclusion, the patient, in this case, is a rare occurrence of Felty’s syndrome presenting with symptomatic neutropenia without any RA symptoms. This presentation is often difficult to diagnose due to the numerous possible etiologies of neutropenia in the general population. The prognosis of Felty’s syndrome is favorable, as current treatment modalities have been able to decrease the risk of infections and increase the quality of life of the patients.

Declarations

Consent for Publication

Written informed consent for publication of their clinical details and/or clinical images was obtained from the patient.

Availability of Data and Material

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Competing Interests

Authors declare no competing interest.

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Authors’ Contributions

LA analyzed and interpreted the patient data, including all laboratory results and imaging studies. LA wrote the manuscript. PP obtained all the laboratory results, made the final diagnosis and followed up with the patient for post diagnosis care. PP edited and made final corrections to the manuscript. All authors read and approved the final manuscript.

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