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An Interesting Case of Rheumatoid Arthritis

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Abstract:- Felty's syndrome is a rare complication of rheumatoid arthritis, less than 1% of RA people experience this[i]. Felty's syndrome is characterized by triad of rheumatoid arthritis, enlargement of the spleen and low neutrophil count[ii]. Typically, the syndrome only affects people with long-standing disease characterized by RF and ANA seropositivity and the presence of rheumatoid nodules and articular deformities, often in the context of minimal synovitis[iii]. This case report highlights the features of Felty's syndrome.

Keywords:- *Felty's Syndrome, Rheumatoid Arthritis, Large Granular Lymphocyte Syndrome.*

I. INTRODUCTION

A rare autoimmune condition known as Felty's syndrome is defined by the triad of rheumatoid arthritis, splenic enlargement, and low neutrophil counts. Although this triad of symptoms does not have to be present in all patients, Felty's syndrome must be diagnosed with neutropenia, defined as an absolute neutrophil count of less than 100/mm3[iv]. The disorder is more widespread in people between the ages of 50 and 70, and it affects women more frequently than men[v]. More common in Caucasians than in the African descent[vi]. In this case report we discuss in detail about Felty's syndrome.

II. CASE REPORT

The case is of a 52-year-old female patient known case of rheumatoid arthritis on methotrexate and hydroxychloroquine who presented to the emergency department with c/o with fever with chills and rigor, cough with expectoration and chest pain.

On examination, she is poorly built and nourished. Vitals temperature - 101.5 °F BP- 100/60 mmHg, PR- 112 beats/minute sinus rhythm, normal volume character, and condition of the vessel wall, oxygen saturation-92% on room air, respiratory rate – 32 cycles, spleen and cervical lymph nodes palpable, rheumatoid nodules, boutonniere deformity and small joint tenderness are present. The patient was admitted to critical care. Blood investigations showed: HB- 9.8 (13.0 - 18.0 gm/dl), TC-1210, N-22% (4000.0 - 10000.0 / µl), CRP-192 (<6), ESR-81, absolute neutrophil of 28 cells/mm3, and platelet count of 138 × 103/µL. URE- pus cells 4-5, albumin- nil, RBC-nil(foley's catheter was not inserted), sugar 70 mg/dl, urea – 52 (8.0 - 49.0 mg/dl), serum creatinine- 1.1 (0.7 - 1.2 mg/dl) serum sodium- 137 mmol/l (135.0 - 148.0 (mmol/l), serum potassium- 4.8 mmol/l (3.5 - 5.1 mmol/l), S. Total bilirubin- 1.2 (0.0 - 1.2 mg/dL), SGPT- 23 (0.0 - 40.0 IU/L), SGOT -35 (0.0 - 50.0 IU/L), ALP- 124 (35.0 - 129.0 IU/L) Total protein 5.1 (6.4 - 8.3 g/dl), S. albumin - 2.8 (3.5 - 5.2 g/dl) S. globulin - 2.3 (2.0 - 3.5 g/dl), S. calcium - 8.3 (8.5 - 10.1 mg/dL), Peripheral blood smear showed no significant abnormalities with normal reticulocytes. Blood cultures showed growth of Pseudomonas aeruginosa. Chest x-ray reveals areas of opacity (left side). USG abdomen and pelvis -moderate splenomegaly. 2D ECHO-normal LV function, IVC- collapsible. IV fluids were given according to IVC and cardiac status and appropriate antibiotics were started according to the culture and sensitivity. The patient was symptomatically improved and shifted to the ward. The patient was diagnosed with community-acquired pneumonia 6 months back, she also had a history of otitis media 3 months back and recurrent urinary tract infection. The patient was diagnosed with Felty's syndrome. The patient was referred to a rheumatologist for better care.

III. DISCUSSION

- *Felty's Syndrome*

Felty's syndrome comprises an uncommon but severe disease subset, defined by the triad of RA, splenomegaly (present in >90% of cases), and leukopenia[vii]. The syndrome classically occurs in patients with long-standing disease, characterized by RF and ANA seropositivity and the presence of rheumatoid nodules and articular deformities, often in the context of minimal synovitis[viii]. In addition to rheumatoid nodules, these patients often have other extra-articular manifestations and are at an increased risk for bacterial infections and chronic nonhealing ulcers, leading to skin infections. The risk of bacterial infection is greatest in patients with more pronounced neutropenia (e.g., absolute neutrophil count [ANC] <100/mm3)[ix]. Low serum complements may also be seen in Felty's syndrome, with bone marrow biopsy typically yielding either normal or hyperplastic cell lines[x]. In addition to associations with the HLA-DRB1*0401 allele, Felty's syndrome is characterized by the presence of autoantibodies directed against citrullinated histones that bind to activated neutrophils and neutrophil extracellular traps (NETs)[xi].

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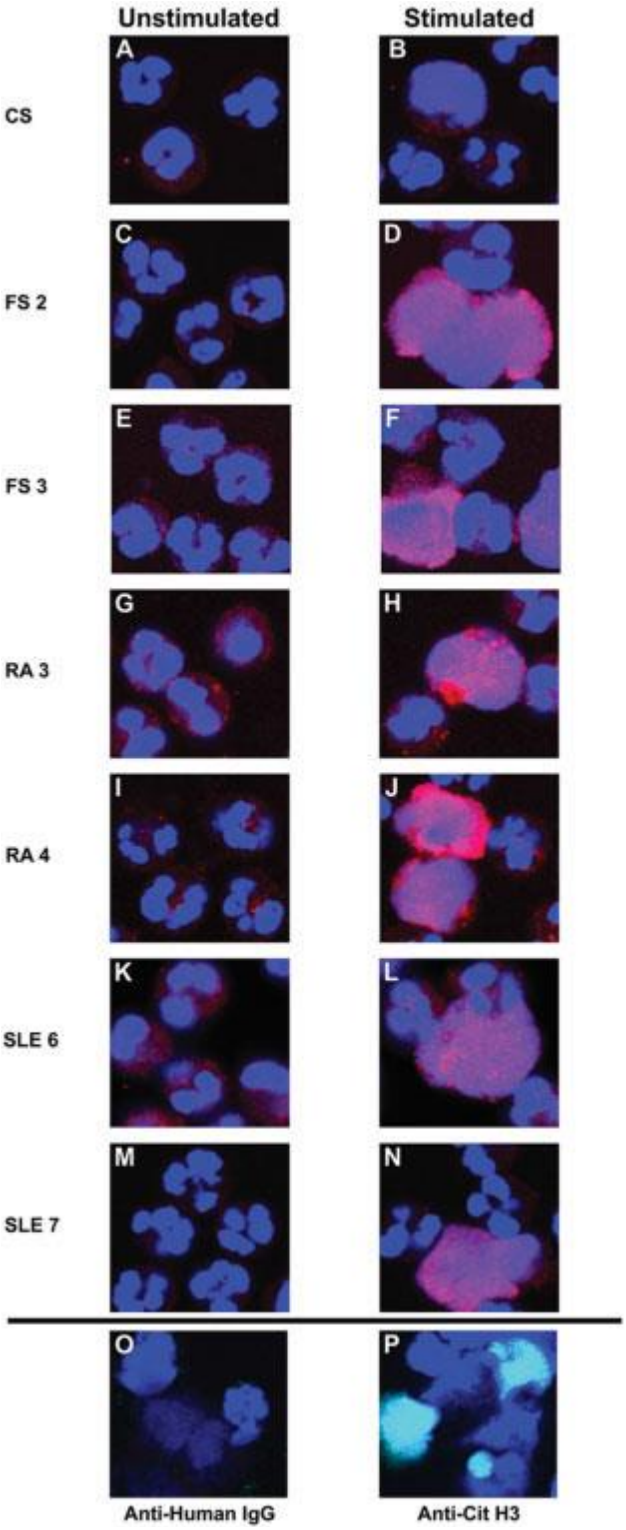

Fig 1. Binding of autoimmune sera to activated neutrophils.

A–N, Unstimulated and lipopolysaccharide (LPS)–stimulated neutrophils were tested for binding by autoimmune and control serum (CS) IgG. Representative sera are shown. **A** and **B,** Control serum. **C** and **D,** Serum from Felty's syndrome (FS) patient 2. **E** and **F,** Serum from FS patient 3. **G** and **H,** Serum from rheumatoid arthritis (RA) patient 3. **I** and **J,** Serum from RA patient 4. **K** and **L,** Serum from systemic lupus erythematosus (SLE) patient 6. **M** and **N,** Serum from SLE patient 7. Stimulated neutrophils, particularly those that had released neutrophil extracellular chromatin traps, reacted more strongly with autoimmune IgG. **O,** as a negative control, anti-human IgG alone was used on LPS-stimulated neutrophils. **P,** LPS-treated neutrophils were used to detect deiminated histone H3. Anti–citrullinated histone H3 (Anti-Cit H3) was detected with green fluorescence. IgG binding is shown in red; DNA is shown in blue. The overlap of green and blue appears as aqua. Original magnification _ 400[xii].

About 30% of Felty's syndromes patients exhibit an increase of large granular lymphocytes (LGL). Large granular lymphocyte syndrome linked to simple Rheumatoid arthritis shares several immunogenetically and phenotypically comparable features with Felty's syndrome but differs clinically. In addition to marked neutropenia, patients typically demonstrate the expansion of LGL cells in both the blood and bone marrow, but splenomegaly is less common than in Felty's. LGL cells represent in vivo–activated cytotoxic T cells (CTC) or natural killer (NK) cells, and they proportionally represent 85% (CTC) and 15% (NK) of LGL syndromes, respectively.73,74 LGL affects as many as 1% of patients with RA but is also seen in other inflammatory diseases (SLE, Sjögren's syndrome), viral infections, and hematologic malignancies (myelodysplastic syndrome). As opposed to aggressive and fatal subsets of LGL leukaemia, the lymphocytosis associated with RA tends to follow an indolent course[xiii]. In addition to rare malignant transformation (characterized by monoclonality), LGL places patients with RA at risk of infection. Although seemingly counterintuitive in the context of marked neutropenia, methotrexate remains the treatment of choice for both and LGL syndromes complicating Rheumatoid arthritis[xiv]. Other second-line therapies include cyclosporine, cyclophosphamide, glucocorticoids, and granulocyte colony-stimulating factors[xv]. Splenectomy is reserved for those patients with Felty's syndrome and documented splenomegaly who are recalcitrant to medical management[xvi].

IV. CONCLUSION

Felty's syndrome is an uncommon form of rheumatoid arthritis that includes a triad of splenomegaly, neutropenia, and persistent rheumatoid arthritis. Even though this illness was first identified more than 80 years ago, there is no established management and therapy plan for Felty's syndrome.

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