A case of Rheumatoid Arthritis complicated with both Pyoderma Gangrenosum and Felty Syndrome simultaneously.

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Abstract

Background

Felty syndrome (FS) is a rare, severe form of extra-articular manifestation of Rheumatoid Arthritis (RA). It is characterized by the presence of splenomegaly and absolute neutropenia less than 1500/mm^3 in a patient with long standing RA. The incidence of FS in RA patients is around 1-3%. Pyoderma Gangrenosum (PG) is another rare extra articular complication of RA which presents with neutrophilic dermatosis leading to poorly healing skin ulcers. The overall incidence of PG is thought to be around 3-10 cases per million per year. RA increases this risk of having PG more than 3 folds. We present a case report of a patient who developed both these rare complications simultaneously after six years from diagnosis of RA.

Case Report

43 year old mother of two, who had been previously diagnosed to have sero positive RA and had been only on Sulfasalazine as a DMARD, presented with suspected seizure episode. She was transferred to Neurology unit of CSTH from a local hospital. A thorough history and a clinical examination revealed that she had pseudo seizure, PG ulcers on right foot and left inguinal region which was confirmed by skin biopsy and hepatosplenomegaly which was supported by ultrasound scan. Her blood investigations over next few days showed continued bicytopenia, particularly severe neutropenia. The inflammatory markers, ESR and CRP were persistently above 70mm/1^{st} hour and 100mg/dl respectively . She was started on a broad spectrum antibiotic, IV Ticarcillin empirically with the opinion of Microbiologist and granulocyte colony stimulating factor (G-CSF) was also administered upon advice from Haematologist. Two pints of blood were transfused as she had low hemoglobin. After further assessment for

differential diagnosis of septicemia, Hemophagocytic Lymphohistiocytosis (HLH) and Large Granulocytic Leukemia (LGL), the diagnosis of FS seemed to be the likeliest cause. HLH and LGL were excluded with bone marrow aspiration. We initiated treatment with high dose Prednisolone in a tapering off regime. Ciclosporin was also started at a low dose with a plan to increase gradually. We noticed an improvement of her general well-being and PG ulcers when she presented to clinic after one month. We hope to start Methotrexate for FS with gradual improvement of PG which would allow us to tail of both steroids and Ciclosporin.

Conclusion

Even though PG and FS are rare complications in RA, being watchful about these will help diagnose them promptly and will prevent serious outcomes of them.