Idiopathic Hypereosinophilic Syndrome with Rheumatoid Arthritis

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Abstract: Idiopathic Hypereosinophilic Syndrome is defined as 1) sustained eosinophilia more than 1500/mm³, 2) absence of other causes of eosinophilia, 3) multiorgan involvement. Very few case reports have been reported in the past of rheumatoid arthritis and Idiopathic Hypereosinophilic Syndrome. Here we report a rare case of 59 year old patient, known case of rheumatoid arthritis since the last ten years on DMARDs (Disease Modifying Anti Rheumatoid Drugs) who developed marked eosinophilia and multi-organ involvement in the form of stroke, urticarial skin rashes and left sided pleural effusion. She was treated with steroids along with methotrexate initially. Once she started developing cushingoid features, steroids and methotrexate was replaced by cyclosporine and sulfasalazine. This case highlights importance of eosinophilia in rheumatoid arthritis and also possibility of common pathogenesis of Idiopathic Hypereosinophilic Syndrome and rheumatoid arthritis.

Keywords: eosinophilia, stroke, pleural effusion, urticarial

I. Case Report:

We describe a 59 year old female patient, known case of seropositive Rheumatoid Arthritis since the last ten years. She was started on methotrexate at the dose of 15mg once a week which was continued for three months initially. As the patient did not show improvement in DAS-28 score and clinically, her treatment was escalated to triple drug therapy in the form of methotrexate (15mg once a week), sulfasalazine (1000mg twice a day) and hydroxychloroquine (200mg twice a day). She was also started on low dose steroids (10mg once a day). This treatment was continued for six months and patient showed improvement in joint pain and swelling. In July 2015, she developed urticarial rash all over the body which was associated with severe pruritus. At this time she also complained of dry cough associated with shortness of breath on exertion. Examination of respiratory system was suggestive of reduced air entry in left axillary and infrascapular region. On investigations her total leucocyte count (TLC) was 30,000/mm³ and eosinophil percentage was 34% (count-10,200/mm³). Her ESR was 67mm and CRP was 11.03mg/dl and positive Rheumatoid Factor (RF). Chest x-ray was suggestive of left sided pleural effusion. Pleural fluid aspiration was done.

It was exudative in nature as per Lights criteria with total leucocyte count of 200 cells with 5% eosinophils. Parasitological investigations in the form of stool routine and microscopy were normal. She did not complain of joint pain and swelling at this time. We increased the dose of steroid to oral prednisolone of 40mg/day and antihistaminics. Her rash improved over three weeks. Two months after developing rash, she presented in emergency with left sided hemiparesis. On examination power of left upper and lower limb was 1/5 and that of right upper and lower limb was normal. Left plantar was extensor. NCCT head was showed deep white matter ischaemic changes. Complete hemogram showed persistent eosinophilia (TLC-18870/mm³) with eosinophil percentage of 25%. We continued her on oral prednisolone (40mg daily) along with methotrexate, sulfasalazine, hydroxychloroquine and antiplatelet agent (ecosprin-75mg). Other blood investigations—liver and kidney function tests were normal. Bone marrow test was done to rule out any malignancy which also showed increased eosinophilic counts. Bone marrow smear was hypercellular with increased eosinophilic precursors suggestive of Idiopathic Hypereosinophilic Syndrome.

ANA (anti-nuclear antibody) and ANCA (anti-nuclear cytoplasmic antibody) antibodies were absent. Serum IgE levels were normal. 2-D echocardiography was normal. Done to rule out evidence of eosinophilic myocarditis. Repeat complete hemogram was done after two months which showed persistent eosinophilia, although it had improved (TLC-11000, with 20% eosinophils). Since patient had started developing cushingoid facies we tapered the dose of steroids and added cyclosporine (50mg twice daily) along with sulfasalazine. Complete hemogram after two months of cyclosporine therapy showed marked improvement with TLC count of 7000/mm³ and eosinophil percentage of 15%. At present patient is stable on cyclosporine therapy with tapering dose of steroid therapy. A diagnosis of Hypereosinophilic Syndrome was made on the basis of...
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criteria – persistent eosinophilia, absence of other causes of eosinophilia and multiorgan involvement in the form of lungs, skin and central nervous system in our case.

II. Discussion

Idiopathic Hypereosinophilic System is a disease of exclusion. Eosinophilia in rheumatoid arthritis patients has been directly related to increased disease activity and various other extra-articular manifestations like rheumatoid nodules, vasculitis, neuritis, pulmonary fibrosis and cutaneous infarctions (1,2,3). Our patient did not report any deterioration in joint pains and swelling at the time of development of eosinophilia, hence it cannot be linked to increased disease activity. Persistent eosinophilia can be found in 10 to 40% cases of rheumatoid arthritis (4). Eosinophilia has been known to occur in patients receiving gold therapy in rheumatoid arthritis (5). Gold therapy is no longer given these days. Although rare, articular manifestations of HES (Hypereosinophilic Syndrome) have been described in the past, mimicking features of rheumatoid arthritis (6-10). Tay described ten patients of acute onset polyarthritis with hypereosinophilia as “eosinophilic arthritis” (11). Our patient already had long standing history of rheumatoid arthritis and so co-existence of the two diseases could be a possibility. Eosinophilia could also be due to drug hypersensitivity reactions like by drugs like adalimumab (12).

III. Conclusion

This case holds importance as it is not only a rare case of hypereosinophilic syndrome with rheumatoid arthritis. It also highlights the importance of eosinophilia in rheumatoid arthritis. Both steroids and cyclosporine proved to be useful in treating eosinophilia in our patient.
References


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[17] HypereosinophiliaandSeroconversionofRheumatoidArthritis

[18] Casebasedreview

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