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Effective hydroxychloroquine monotherapy for thrombocytopenia in lupus: A case report

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Abstract

A 22-year-old female presented with pruritic papular skin rash and arthralgia in her knees. Lab results showed extremely low platelet count 42000, and antinuclear antibodies (ANA) of 1:320 in a speckled pattern. She was diagnosed with severe immune thrombocytopenia in systemic lupus erythematosus (SLE-ITP). Hydroxychloroquine (HCQ) 300 mg daily was administered, and within 6 months, platelet increased to 76,000 and one year later it was 182000. Her rash and joint pain fatigue completely resolved, and within a year, her platelet count was normal and continued to be normal for 20 years of follow-up. This is a unique case which utilizes HCQ alone as first-line treatment for severe SLE-ITP with long term follow-up.

Keywords

systemic lupus erythematosus; thrombocytopenia; hydroxychloroquine; rheumatology; hematology

Abbreviations

ANA: antinuclear antibodies; SLE-ITP: immune thrombocytopenia in systemic lupus erythematosus; HCQ: Hydroxychloroquine; CSs: corticosteroids

Introduction

SLE is an inflammatory autoimmune disease involving multiple organs, with a wide range of clinical manifestations. Immune thrombocytopenia in SLE (SLE-ITP) has often been associated with other clinical features including (but not limited to) neuropsychiatric and renal involvement, haemolytic anemia, low complement levels, and high titer double-stranded anti-DNA antibodies [1]. Severe SLE-ITP is usually defined as having a platelet count below 50,000, and can be accompanied by potentially life-threatening events resulting from hemorrhage in one or more major organs [1-3]. Standard treatment is corticosteroids, but their adverse effects and chance of relapse in lowering dose is also very challenging.

Case Report

A 22 year old Caucasian female presented with one year of intermittent low grade fever, knee arthritis, headache, nausea, fatigue, facial rash, and photosensitivity. Physical examination was remarkable for punctate red papules over the malar areas of the face plus swelling and pain on motion in both knees. Laboratory tests revealed a platelet count of 42,000 and a positive antinuclear antibody (ANA) in a speckled pattern at a titer of 1:320 (Figure 1). Other testing that was normal or negative

included white blood cells, hemoglobin, chemistryprofile, clotting studies, complement levels, antibodies to double stranded DNA, urinalysis, and bone marrow biopsy. Peripheral smear was not done.There was no history of estrogen use nor over the counter supplements.No obvious bleeding was noted in any organs. A diagnosis of systemic lupus erythematosus (SLE) was made, and treatment was started solely with hydroxychloroquine (HCQ) 300mgdaily. The patient consented to this treatment as she did not wish to institute corticosteroids. In accordance with the policy of our hospital ethical committee, it was not necessary to obtain prior approval. Six months later her platelet count was 76,000, there was no evidence of any bleeding, and all other presenting complaints and physical findings had resolved (Figure 1). One year after initial presentation her platelet count was 182,000, and it remained in the normal range during twenty years of additional follow-up (Figure 1). It was necessary to continue HCQ dosage at 300mg daily to sustain subjective and objective remission of her other SLE pneumonia.

Discussion

Standardized treatment of severe SLE-ITP is heterogeneous, often beginning with high dose corticosteroids (CSs) as first line treatment [3-4]. Long-term responses to CSs alone are quite variable, with relapses in many cases when dosage is lowered. The addition of second-line agents to CSs included azathioprine, cyclophosphamide, danazol, intravenous immunoglobulin, rituximab, and HCQ [1,3-6]. Several studies have indicated that HCQ is effective in treating refractory severe SLE-ITP, with long-term responses reported [3-4]. The required daily dosage of HCQ in this situation (and for SLE in general) is variable, but can be optimized by measuring serum concentrations [7-8]. It is reasonable to assume that our patient could have been treated with a higher initial dosage of HCQ for the first six to twelve months to enhance her response.

We know of no published studies utilizing HCQ alone as first-line treatment of severe SLE-ITP. A recent editorial has commented on the overreliance of CSs in a number of rheumatologic disorders, including SLE [9]. Obviously, each individual case of SLE must be evaluated on its own merits to implement a reasonable and effective treatment regimen. We propose that in selected cases of severe SLE-ITP, uncomplicated by haemorrhage or other life-threatening manifestations, consideration should be given to initial conservative treatment with HCQ alone [10].

Conclusion

In conclusion, there are no previous publications that utilize HCQ alone as first-line treatment of severe SLE-ITP. SLE is an inflammatory autoimmune disease involving multiple organs. Severe SLE-ITP is usually defined as having a platelet count below 50,000. Standard treatment for severe SLE-ITP begins with high dose of corticosteroids (CSs) as first line treatment, followed by HCQ as one of the second-line agents. Try to avoid use of corticosteroids especially in long term because of potential adverse effect and risk outweighs the benefit.

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Figure

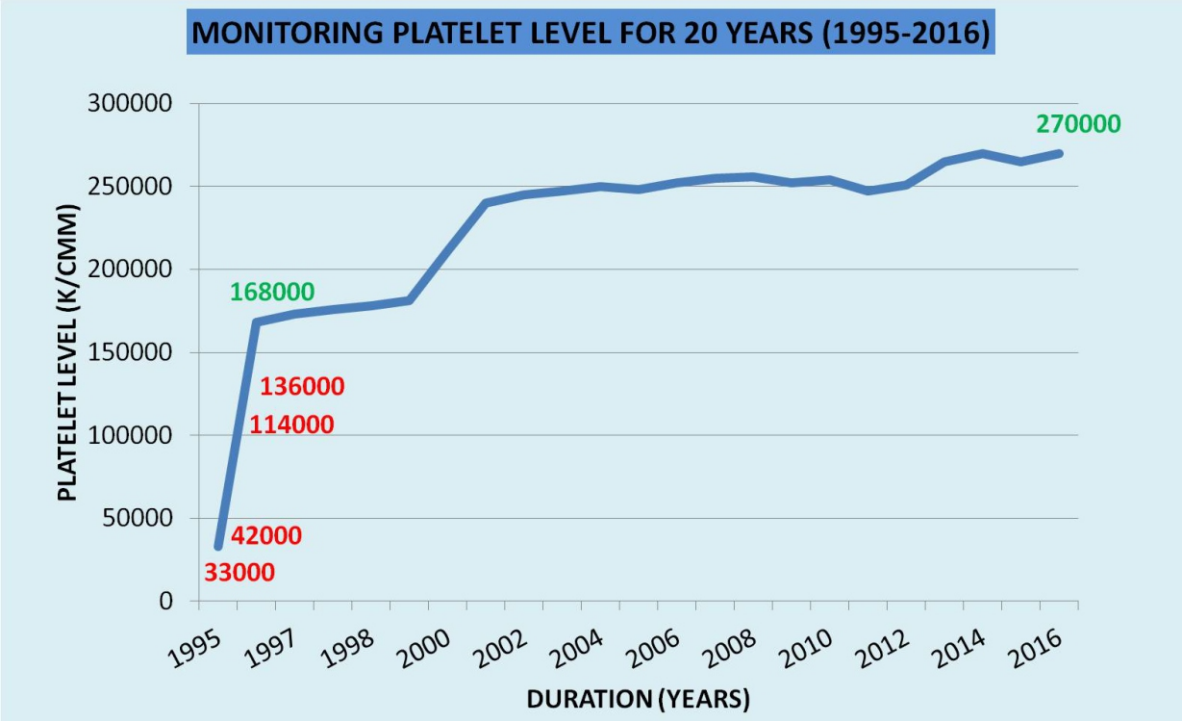

Figure 1: Monitoring Platelet Level for 20 Years [1995-2016]

Initial lab showed platelet count of 42,000. Within 6 months of therapy, platelet increased to 136,000. Within a year, platelet level increased to normal range [above 150,000]. From there, platelet level continued to be normal and gradually increased, reaching as high as 270,000 by 2016.

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