Annals of Arthritis and Clinical Rheumatology

Case Report
Published: 26 May, 2020

Systemic Lupus Erythematosus Associated Chorea Treated Successfully with Rituximab Case Report

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Abstract

Chorea is a movement disorder well known to be a rare clinical manifestation of Systemic Lupus Erythematosus (SLE) and primary or secondary Antiphospholipid Syndrome (APS). However, there are no clear guidelines in the literature about the treatment of lupus chorea. We are lacking the standard care to approach such presentation. Here we describe a case of a young female patient with systemic lupus erythematosus associated with positive antiphospholipid antibodies, who presented with recurrent episodes of chorea and was initially treated symptomatically. But because of the repeated attacks of chorea, the patient was successfully treated with Rituximab.

Keywords: Systemic lupus erythematosus; Rituximab; APS; IgG; IgM

Introduction

Chorea is an abnormal involuntary movement disorder, belongs to a group of neurological disorders called dyskinesia. Chorea is characterized by brief and irregular contractions that are neither repetitive nor rhythmic, but the contractions flows from one muscle or muscle group to another without any pattern. The incidence of chorea associated with SLE is less than 5 percent of patients [1]. Some data showed strong association between the development of movement disorders in general and presence of antiphospholipid antibodies aPL [1]. Chorea is also seen in approximately 1 percent of patients with primary antiphospholipid syndrome.

Case Presentation

A 15 years old female presented on early April 2016 with symmetrical polyarthritis involving the small joints of the hands and the feet with constitutional symptoms (fever, weight loss and anorexia) for more than 4 months. She had a photosensitive malar rash as well. Other systemic symptoms were unremarkable. The patient had a family history of SLE. She was a student with a good performance at school.

On examination: The patient was fully conscious, alert, and oriented. She was febrile and she had livedo reticularis in her both hands and legs. She had arthritis involving both hands and feet, she also had a malar rash.

The patient was admitted at the time of presentation to the hospital, and her work up revealed: Pancytopenia, ESR of 87, normal renal and liver function tests, and the 24 h urine protein test was: 336 mg/24 h (there was no significant proteinuria).

Her Antinuclear Antibody (ANA) by Immunofluorescence (IF) was positive with a titer of more than 1280. Her Anti-Deoxyribonucleic acid, beta 2 Glycoprotein, IgM, IgG, anti-cardiolipin IgM, and anti-cardiolipin IgG were all highly positive. Anti-smith was negative, C3 and C4 results were low. So based on her clinical presentation and the positive laboratory tests the patient was diagnosed with SLE associated with positive Antiphospholipid antibodies and she was treated with the following: Glucocorticoid 0.5 mg/kg/day, Hydroxychloroquine 200 mg P.O. once daily, Aspirin 81 mg P.O. once daily. Vitamin D and calcium supplements daily 8 month later the patient developed acute onset choreoathetoid involuntary movements of the upper and lower extremities, Physical examination revealed choreic movements of her right hand and foot, They were jerky, purposeless, intermittent, and irregular movement with a positive milking maneuver test. In Magnetic Resonance Imaging (MRI) the findings were: abnormal scattered white matter seen as a high signal in T2, and the periventricular was free of abnormalities such as restricted diffusion or enhancement.

The Electroencephalogram (EGG) results were normal. A repeated antiphospholipid antibody test was strongly positive (triple positive). The Patient was assessed by the neurology department,

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Received Date: *20 Apr 2020*
Accepted Date: *20 May 2020*
Published Date: *26 May 2020*

Citation:
Somaily M, AlMania HI, Ali Buhran L, Alhanash A. Systemic Lupus Erythematosus Associated Chorea Treated Successfully with Rituximab Case Report. Ann Arthritis Clin Rheumatol. 2020; 3(1): 1018.

then she was started on haloperidol 0.5 mg P.O. twice daily for one week then tapered down to once daily, then the treatment was stopped within few weeks. Because she was responding very well 6 months later, the patient developed another episode of chorea, and she was found to have a significant proteinuria. Her 24 h urine protein test was 2548 mg/24 h. Her Renal profile results were normal before, but when she underwent renal biopsy it revealed: Diffuse proliferative glomerulonephritis corresponding to lupus nephritis WHO Class IV. The patient started on pulse steroid therapy, she also started on mycophenolate mofetil 3 g/day and was given another course of haloperidol to control her choreoathetoid movement. After the treatment she had a remission of lupus nephritis, there was no proteinuria during her follow up. But regarding the choreaoathetosis disorder, she did have several attacks of chorea during 2018, and every visit the patient had to the neurologist, she was given a symptomatic treatment (which was haloperidol). At this stage we decided to start her on rituximab 375/m^2/weekly for total of 4 doses. The patient responded very well and there was no more chorea or movement disorders found for almost 6 months.

Discussion

Movement disorders are well known to be rare clinical manifestation in SLE and APS, this has been reviewed in several studies. The mechanism behind that is still not clear, although some theories mentioned three possible mechanisms that may contribute to it; the first mechanism is that the antibodies may generate an inflammatory change in cerebral vessels causing underlying ischemic insult to the basal ganglia which responds for the development of the movement disorder. Second mechanism is that it may result from neuronal dysfunction from the immune complex deposition. And third mechanism is that the immune and non-immune effects of infections or toxins may also be responsible [2]. The second mechanism could have played a role in the development of chorea in our case where the antiphospholipid titer was significantly high where it could lead to venous or arterial thrombosis. In our patient there was no signs found of vascular thrombosis in the radiological studies. Current management for such disorders is using corticosteroid and other immunosuppressive therapy. Some reviewed reported cases were self-limited disorders and therefore may not require therapy. However in our case we kept the patient on haloperidol which was effective to some extent, but the problem was the recurrence of the movement disorders. So recently we started our patient on rituximab. And there was no reported cases discussing use of rituximab in such disorder. Rituximab is a B cell-depleting monoclonal anti CD20 antibody, it was administered to 10 patients with neuropsychiatric SLE, but not on cases which includes choreoathetosis disorders, which were unresponsive to immunosuppressive and plasma exchange therapy.

Result and Conclusion

There isn't many previous studies showing the effect of Rituximab treatment in SLE associated chorea, but some case reports mentioned what treatment was given to these patients with the unusual presentation and what were the results of the treatment, one of the case report was in may 1997, where studies were done in 50 patients with APS related chorea were reviewed. The following antibodies were detected: Lupus anticoagulant (92%), anticardiolipin antibodies (91%), antinuclear antibodies (82%), anti-DNA (59%), anti-Ro (10%), anti-RNP (8%), anti-La (2%), and anti-Sm (2%). Chorea in these patients responded to a variety of medications, for example: steroids, haloperidol, antiaggregants, anticoagulants, or a combination of therapy, these medications are usually prescribed in the presence of other manifestations of APS or SLE [3]. Another case report was in April 1973, one case reported treatment with haloperidol of such a condition in an 18 years old female, resulted in a prompt and complete subsidence of symptoms, in contrast to previous episodes without haloperidol lasting as long as three months. The patient had a recurrence of choreiform movements which happened nine months later and it was responded to treatment similarly [4]. While in 2007, one case reported a 33 years old female with SLE who developed Systemic lupus erythematosus associated chorea with multiple involuntary movements, and was treated with Intravenous Immunoglobulin (IVIg) after which a remarkable improvement of the abnormal movements was achieved [5]. And In 2013, a case report of a 33-year-old female who developed a movement disorder during maintenance therapy for Systemic Lupus Erythematosus (SLE) was treated with a combination of high-dose corticosteroid, immunosuppressive agents (rituximab, cyclophosphamide, mycophenolate mofetil), antithrombotic therapy (heparin, cilostazol), and dopamine antagonists. Her symptoms remitted partially [6].

From this previous case report it was concluded that there is inadequate knowledge of the pathogenetic mechanisms and the lack of randomized-controlled trials, and that the treatment of chorea in SLE is not yet completely codified. The most recent European League against Rheumatism (EULAR) recommendations for the management of SLE with neuropsychiatric manifestations advise to use dopamine antagonist to treat patients with persistent choreic symptoms [7].

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