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Contents

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World Journal of Clinical Cases

Thrice Monthly Volume 10 Number 4 February 6, 2022

Contents

CASE REPORT

World Journal of Clinical Cases

Thrice Monthly Volume 10 Number 4 February 6, 2022

Contents

World Journal of Clinical Cases

Thrice Monthly Volume 10 Number 4 February 6, 2022

Contents

ABOUT COVER

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LETTER TO THE EDITOR

Rituximab as a treatment for human immunodeficiency virus-associated nemaline myopathy: What does the literature have to tell us?

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Abstract

We presented a letter about a case of a 37-year-old Black female with a history of human immunodeficiency virus and an undetectable viral load. She was evaluated with weakness in the scapular (grade III) and pelvic girdles (grade II), elevation of creatine phosphokinase levels and muscle biopsy compatible with nemaline myopathy. She was treated with rituximab showing improvement of the condition.

Key Words: Case report; Human immunodeficiency virus; Nemaline myopathy; Rituximab; Rheumatology; Therapy

Core Tip: Rituximab may be a therapeutic possibility for the treatment of nemaline myopathies (*e.g.*, human immunodeficiency virus-associated and monoclonal gammopathy of undetermined significance-associated) because it is less aggressive and has fewer side effects compared to current therapies. It may be especially helpful in cases of severe visceral involvement. However, the cost and unavailability of therapy can be a limiting factor.

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TO THE EDITOR

Dear Journal Editor,

We read the paper of Profs. Wang and Hu[1] who presented a case report about nemaline myopathy (NM) with dilated cardiomyopathy. The authors aimed to describe a rare myopathy with severe cardiovascular impairment and whose outcome was positive.

We would like to present a case of human immunodeficiency virus (HIV)-associated NM (HIV-NM), and it responded to rituximab treatment.

The case involves a 37-year-old Black female with a history of HIV and an undetectable viral load with regular use of dolutegravir, darunavir and ritonavir. Six years ago, the patient started to present objective muscle weakness in all four limbs, in addition to increased muscle enzymes and electroneuromyography with evidence of a myopathic pattern. With the initial hypothesis of polymyositis, the patient received prednisone (1 mg/kg/d) with partial improvement of clinical and laboratory status. The patient was admitted to our service for a clinical reassessment and follow-up 3 years ago. The patient had an undetectable viral load, normal protein electrophoresis and serum levels of creatine phosphokinase at 2550 U/L using methotrexate 25 mg/wk and prednisone 5 mg/d.

She had weakness in the scapular (grade III) and pelvic girdles (grade II), required a wheelchair for locomotion and showed muscle magnetic resonance with evidence of symmetrical and bilateral muscle edema in the muscular bellies of the pelvic girdle and thighs. A muscle biopsy showed a myopathic and dystrophic patterns with the presence of marginal vacuoles with nemaline rods. Regarding the possibility of HIV-NM, methylprednisolone pulse therapy 3 g was started in one dose, and immunosuppressive drugs (azathioprine 300 mg/d, methotrexate 20 mg/wk and prednisone 20 mg/d) of previous use were maintained without significant improvement. Due to refractory disease and despite off-label, the patient consented to introduce rituximab 2 g/cycle as a possible option to promote disease induction.

The patient had progressive improvement. After two rituximab cycles, there was an important improvement in muscle weakness (lower limbs grade III and upper limbs grade IV) and independence for basic activities of daily living and a drop in creatine phosphokinase (275 U/L).

According to the literature, HIV-NM usually has a good response to immunosuppressive therapy[2-4]. A case report of a 65-year-old woman with severe cardiomyopathy and NM was recently described, in which the treatment was clinical compensation for cardiomyopathy associated with autologous stem cell transplantation[4]. A German cohort demonstrated that the most effective treatment strategy in NM was autologous bone marrow transplantation, but the one performed was immunosuppression with glucocorticoids (62%)[3]. Thus, in severe cases such as the one presented by the authors[1], we ask ourselves if the use of rituximab could not be an option as a way to delay the evolution of the NM.

Among the postulated theories, two stand out: (1) HIV should result in the formation of rods and/or serve as a trigger for the immune system to destroy muscle fibers; and (2) Genetic disorders caused by HIV cause rod formation[3]. In this context, therapy with rituximab may be an interesting treatment option in NM because: (1) Recent studies have shown improvement in the weakness of rituximab with no side effects obtained; (2) Lymphocytic infiltrates in muscles of NM patients are commonly confused with polymyositis; and (3) Limited effects with treatment[5].

Another interesting point to remember is that NM is often associated with monoclonal gammopathy of undetermined significance in case series[6], retrospective studies[7] and cohorts[3], denoting exacerbated lymphocyte activity. The cause for this association, as well as for the association of NM with HIV, is still unknown. However, the good response of this pathology to immunosuppressive therapies[2-4] and bone marrow transplantation[4] denote that options, such as rituximab, with fewer side effects, better dosage comorbidity and lower risks may be a real therapeutic possibility. Even more aggressive treatment regimens such as associations with dexamethasone, thalidomide and cyclophosphamide have already been proposed[8].

Finally, we emphasize that these treatment modalities might be used as an optional treatment to the autologous stem cell transplantation or before that. However, despite the rarity of the disease, further studies with a higher number of patients and adequate follow-up are required.

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