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Marked Efficacy Of A Therapeutic Strategy In A Patient With Necrotizing Myopathy Associated With Anti-SRP Autoantibodies

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Abstract

Background and Rational:

Necrotizing autoimmune myopathy (NAM) (also known as immune-mediated necrotizing myopathy [IMNM]) is a subtype of idiopathic inflammatory myopathy that is characterized by symmetrical, proximal, subacute muscle weakness, high creatine kinase level (CK) (1), and also characterized by its muscle biopsy findings of pauci-immune necrosis and the absence of extra muscular manifestations (2,3). NAM is known to be strongly associated with autoantibodies against signal recognition particle (SRP)(4) and 3-hydroxy-3- methylglutaryl-coenzyme A reductase (HMGCR) (5). While other autoimmune myopathies such as dermatomyositis are strongly associated with other known autoantibodies including Mi-2, melanoma differentiation-associated gene 5 (MDA5), transcriptional intermediary factor 1 γ (TIF1 γ), and nuclear matrix protein 2 (NXP-2) (6). In NAM, SRP antibody levels correlate with CK levels to determine the severity and prognosis of the patient's illness (4).

Inflammatory myopathies including NAM are treated with a variety of treatments, including steroids, intravenous immunoglobulin (IVIG), and other immunosuppressants including methotrexate. Nevertheless, numerous evidence reported treatment-resistant among those patients, and almost all patients require two or more immunosuppressants (7). Given the correlation between pathogenic anti[1]bodies and disease severity in NAM, therapeutic plasma exchange (TPE) could aid in therapy by eliminating these antibodies. Additionally, the effectiveness of rituximab in treating SRP and HMGCR-associated NAM lends credence to the idea that the illness is primarily antibody-mediated (8,9). In the current study, we present the first case report in Saudi Arabia of a 26-year-old female patient who was diagnosed with NAM and had marked improvement after an effective therapeutic strategy.

Introduction

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I. Objectives:

Primary Objective

To present the first case of NAM in Saudi Arabia and explore the differences between the case and other evidence worldwide.

Secondary Objectives

- To provide a view of an unusual treatment plan for necrotizing autoimmune myopathy
- To investigate antibodies SRP and MI2 association with necrotizing autoimmune myopathy
- To explore the malignancy association and its variety among the cases

II. Study design:

This is a case report study.

III. Study Population:

The study will include a full description of the clinical characteristics, investigations, and treatment plan of a 1-case presented to our center in KAMC.

IV. Study procedures:

- The patient was identified and diagnosed with the rare illness; necrotizing autoimmune myopathy and is still being followed up in our neurology OPD in KAMC.
- The authors will prospectively review the system to collect information regarding the history of the present illness, associated symptoms, and past medical & surgical history. As well as the laboratory profile and radiological imaging. The treatment plan will also be reported in the case report. Finally, the follow-up outcome of each OPD will be reviewed and reported.
- No imaging of any site of the body will be taken from the patient.
- Informed consent will be obtained from the patient after gaining ethical approval from KAMC IRP.
- For any additional information needed from the patient, we will contact the patient by phone.
- The study will then publish in a scientific journal only.

- **Study Duration/ Study Timeline:**

Task	**MONTHS**

	1	2	3	4	5	6	7	8	9
Data collection	√	√							
Review of Database and Data Entry in report			√						
Writing Research Paper				√	√	√			
Publishing the Research							√	√	√

V. Ethical part & confidentiality:

The patient will be included in the study after signing the informed consent form (the consent form was provided along with all uploaded files). Patient will be aware that confidentiality will be maintained as possible. And the material will only be published in a scientific journal. Ethical approval will be sought from KAMC IRB. No study activities will be started until the IRB approval is obtained.

VI. Publication:

The main credit in the publication will go to the principal investigator and co-investigators. Those who will contribute less substantially to data collection and analysis will have an acknowledgment in the manuscript.

"The ICMJE recommends that authorship be based on the following 4 criteria:

a. Substantial contributions to the conception or design of the work; or the acquisition, analysis, or interpretation of data for the work.

b. Drafting the work or revising it critically for important intellectual content.

c. Final approval of the version to be published.

d. Agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and

resolved.

VII. Attachments:

Informed consent form.

VIII. References:

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