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Case Report Open Access

Polyglandular Type III C Syndrome Associated with Double-Seronegative Myasthenia Gravis

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Abstract

The neuroimmunologic diseases are a challenge to health services. We described the evolution of MG in the patient presented as proximal weakness, fatigue, chest pain, ptosis, arthralgia, diplopia and dysphonia. Besides that, the patient presented Systemic Lupus Erythematosus (SLE), positivity/reagent for antinuclear antibody (ANA), anti-thyroid peroxidase antibodies (anti-TPO antibodies) and anti-glutamic acid decarboxylase (anti-GAD). Because of the other comorbidities, the hypothesis of the Polyglandular Autoimmune Syndrome (PAS) III was made, represented by the combination of a clinical diagnosis of an autoimmune thyroiditis with Diabetes Mellitus type 1 (DM1).

Keywords: Myasthenia Gravis; Autoimmune disease; Polygrandular syndrome

Introduction

The neuroimmunologic diseases are a challenge to health services, because of their elevated clinical morbitalities [1]. The diagnosis is complex because it involves multiple symptoms, serological exams and image exams or electrical findings. In this case, the evolution of MG in the patient presented as proximal weakness, fatigue, chest pain, ptosis, arthralgia, diplopia and dysphonia. The exams revealed positivity/reagent for antinuclear antibody (ANA), anti-thyroid peroxidase antibodies (anti-TPO antibodies), anti-glutamic acid decarboxylase (anti-GAD) and an electroneuromyography (EMG) with post-synaptic dysfunction at the neuromuscular-junction with a decrement of 10% in ulnar and accessory left nerves. Other comorbidities include: polycystic kidney disease, renal lithiasis and hypoactive bladder.

Case Report

A 50 year-old woman already presented with Systemic Lupus Erythematosus (SLE) on previous treatment in 2008 - date of her first hospital record. At the same year, the patient was diagnosed with polycystic kidney disease associated with renal lithiasis. In 2012, she came to our service due to muscular weakness, ptosis, diplopia and fluctuating dysphonia. Based on that, it was started the neurological investigation. The Ice Pack Test was positive, although the serological exams of autoantibodies anti-acetylcholine and anti-MusK were negative and the electroneuromyography did not have any decrement yet. Based on that, the diagnosis of double-seronegative MG was considered.

Treatment with azathioprine and anticholinesterases resulted in an improvement of the fluctuating neurological symptoms and remission of ptosis, diplopia and dysphonia. However, bulbar symptoms were exacerbated, becoming an OSSERMAN V, and were treated with immunoglobulin, temporarily reducing muscular weakness.

In 2013, this case became to be considered as a type III polyglandular syndrome. In 2014, the patient had aseptic meningitis, believed to have been caused by the use of immunoglobulin. Rituximab was prescribed as an alternative to the azathioprine's failure. In 2016, the department of rheumatology described a pulmonary involvement by her SLE. In May, 2017, she had another complication with bulbar and muscular symptoms, becoming an OSSERMAN V again, and her exam with the vacuum manometer was PINS<50 cmH2O. This clinical condition was treated, another time, with immunoglobulin, successfully. In September 2017, a new electroneuromyography showed more than 15% decrement in two proximal nerves (axillary and musculocutaneous) and facial nerve. By October, the joint accometiment from the SLE became a limitation. In March of 2018, the last description of her hospitalization, she was rated as an OSSERMAN V another time, with bulbar symptoms, dysphagia and dysphonia, bilateral ptosis as diplopia as her current clinical condition.

Discussion

The treatment's success is based on the correct management of the clinical conditions. However, it also involves symptomatic relief, hormonal adjustments and preventing possible complications, depending on each individual case. Therefore, the risks and benefits of using immunomodulatory and immunosuppressive drugs have to be weighted [1,2]. Monoclonal antibodies are a promising new approach to autoimmune disease cases; however, it also brings further challenges to be discovered.

Our study aimed to present an uncommon association of those immune disorders. Systemic Lupus Erythematosus (SLE) is a heterogeneous rheumatic disease with an autoimmune-caused inflammation in multiple organs. The Poly-glandular Autoimmune Syndrome (PAS) is characterized by the coexistence of at least two

immune-mediated endocrinologic disorders. It is classified in I, II, III and IV types [1]. The type III is represented by the combination of a clinical diagnosis of an autoimmune thyroiditis with Diabetes Mellitus type 1 (DM1) [3], without an adrenal cortex disorder that is common to types I and II [4]. Moreover, there is a further classification of PAS type III: A, B and C. As in our case report, the type III-C PAS characterizes itself by an additional organ-specific autoimmune disease, such as MG [5-7].

Conclusion

The epidemiology of Poly-glandular Syndrome is unknown, but more often occurring in females. Its prognosis depends on each individual manifestation. The conditions associated with type III PAS are DM1 and Hashimoto's Thyroiditis in 30%-79% of cases, alopecia or vitiligo in 2%-29% and pulmonary and intestinal abnormalities, rheumatoid arthritis or timoma in 1%-4%. It has already been described as an association between MG and SLE. Therefore, our article presents a report of a rare case of poly-glandular syndrome type III - C, aiming to enlarge the literature acknowledgement about it.

Conflicts of Interest

All authors declared that there are no conflicts of interest.

Ethical Approval and Consent to Participate

Not applicable.

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