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Case Report

Ocular Myasthenia Gravis and Alopecia Universalis

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Abstract

Alopecia areata and myasthenia gravis are two autoimmune diseases that are rarely associated with each other. We present a man aged 47 years who was newly diagnosed as having ocular myasthenia gravis but also had alopecia universalis, which started 15 years ago. His findings of negative decrement response, increased jitter values of the orbicularis oculi muscle in single fiber analysis, negative anti-acetylcholine receptor antibody test, and negative thorax computerized tomography scan for thymoma were all compatible with ocular myasthenia gravis. Pyridostigmine 60 mg 4x1 and methylprednisolone 16 mg 1x1 were administered and relief of the symptoms was observed. At the end of the first month, the patient reported improvement related with body hairs including eyebrows and scalp hairs, which were re-growing. Alopecia areata has been defined as a non-motor symptom of thymoma-associated myasthenia gravis, and usually starts after myasthenia gravis. Different from the alopecia areata, the alopecia universalis seen in this case was not related with the thymoma and started long before the ocular myasthenia gravis. The immunosuppressive treatment (steroid) used for ocular myasthenia gravis was also helpful for alopecia universalis in this case.

Keywords: Ocular myasthenia gravis, alopecia universalis, alopecia, thymoma, anti-acetylcholine receptor antibody, treatment

Oküler Myastenia Gravis ve Alopesia Üniversalis

Özet

Alopesi areata ve myastenia gravis iki oto immün hastalıktır ve nadiren birbirleriyle ilişkilidir. 47 yaşındaki erkek olguya yeni olarak myastenia gravis tanısı konmuş iken, hastanın 15 yıl önce başlayan alopesia üniversalis tanısı da bulunmaktadır. Orbikülaris oküli kasındaki normal dekremental yanıt, tek lif EMG incelemesindeki artmış jitter değeri, normal anti-asetil kolin reseptor antikor seviyesi ve normal toraks BT bulguları oküler myastenia gravis ile uyumludur. Pridostigmin 60 mg 4x1 and metilprednizolon 16 mg 1x1 reçete edilmiş ve semptomlarda düzelme gözlenmiştir. Birinci ayın sonunda, kaşlar ve saçlarda iyileşme olduğu bildirilmiş ve bakı sırasında yeniden çıktığı gözlenmiştir. Alopesia areata myastenia gravis ile ilişkili timomada non-motor semptom olarak tanımlanmıştır ve genellikle de myastenia gravis sonrasında başlar. Alopesia areata'dan farklı olarak, bu olguda gözlenen alopesia universalis timoma ile ilişkili değildir ve myastenia gravisten çok uzun zaman önce başlamıştır. Oküler myastenia gravis için kullanılan immünsüpresif tedavi (kortikosteroid) bu vakada alopesia üniversalis için de fayda sağlamıştır.

Anahtar Kelimeler: Oküler myastenia gravis, alopesia üniversalis, alopesia, timoma, anti-asetilkolin reseptör antikoru, tedavi

INTRODUCTION

Alopecia areata and myasthenia gravis (MG) are two autoimmune diseases that are rarely associated with each other. The ratios of alopecia areata in patients with MG were 3% (202 patients) and 3.7% (159 patients) in two different studies, and alopecia areata is strictly related with thymoma (1,2). Myasthenia gravis usually presents earlier than alopecia areata (1-3). Alopecia areata was defined as a non-motor symptom of thymoma-associated myasthenia gravis by Suzuki et al. (2). Alopecia has many forms, and alopecia areata is one form of alopecia that typically defines one or more round or oval patches of hair loss. All body areas can be affected; however, the scalp is the most affected in this form. Alopecia totalis is used to define complete hair loss of the scalp, whereas alopecia universalis is used to define hair loss from the entire body. An autoimmune attack on hair follicles by autoreactive T cells leads to loss of hairs and 5% of patients with alopecia areata could progress to alopecia totalis or alopecia universalis (4). CD8 + T-cell related toxicity was proposed as the mechanism of alopecia areata (2,4). Similar to alopecia, MG can be defined as ocular, bulbar, and generalized myasthenia gravis. Autoimmune attack of the neuromuscular junctions leads to a deficiency of impulse transmission from nerves to muscles and results with weakness of the muscles. Herein, we present a patient who developed ocular myasthenia gravis 15 years after the development of alopecia universalis.

Patient

A man aged 47 years presented with ptosis and diplopia. Fatigable ptosis while looking straight ahead and improvement after some rest were observed on examination Video 1. Otherwise, the neurologic examination was normal. The clinical diagnosis was ocular MG. Findings of negative decrement response, increased jitter values of the orbicularis oculi muscle in single fiber analysis, negative acetylcholine receptor antibody test, and a negative thorax computerized tomography scan for thymoma were all compatible with the diagnosis. Pyridostigmine 60 mg 4x1 and methylprednisolone 16 mg 1x1 were administered and symptom relief was observed in the first week of treatment. At the end of the first month, the patient reported improvement related with his hair (Figure 1 a, b, c); his eyebrows and scalp hairs were re-growing (Figure 1 b, c). In his medical history, he had also had alopecia universalis for fifteen years. Although he had been treated before with topical medications, no improvements were documented.

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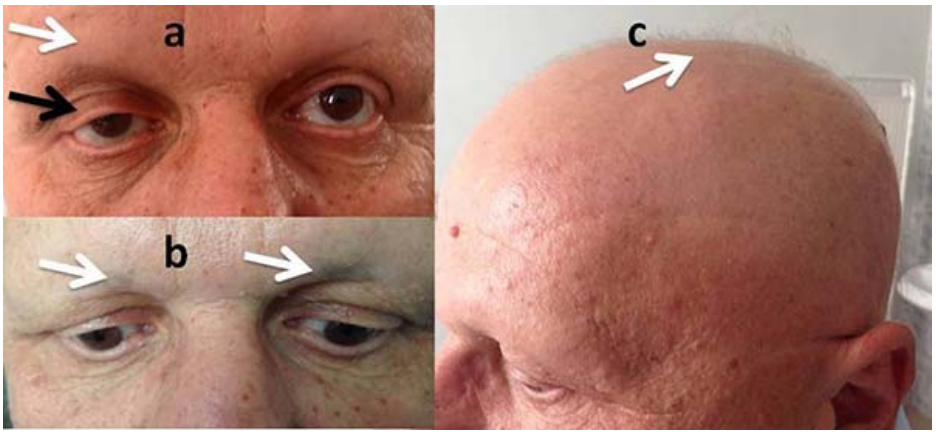

Figure 1: *a. Before treatment, right side ptosis is seen (black arrow) and there is no eyebrow (white arrow) and eyelash. Fig. 1 b and c. After treatment, the eyebrow and scalp hair is seen re-growing (white arrow).*

DISCUSSION

From many aspects, our case was different than previous presented cases. With the exception of the reports by Noguchi et al, most previous cases involved alopecia areata, with high levels of acetylcholine receptor antibody and a strict history of thymoma (1,3,5). Our patient had alopecia universalis and no acetylcholine receptor antibody and a thymoma. The order of appearance of the disease was also different. In two large series MG was defined as the first disease and alopecia areata was the second (1,5). In this case, ocular MG was the second disease, which appeared 15 years after alopecia universalis. Different from alopecia areata, the co-occurrence of alopecia universalis and MG defined previously by Noguchi et al. and the order of diseases were similar to those seen in our case (3). The age of onset of alopecia universalis in Noguchi et al's case was 4 months, whereas it was 32 years in our case, and the time period between the two disorders was 3 years in Noguchi et al's case, and 15 years in ours. Another aspect of our case was the responsiveness of the alopecia universalis to immunosuppressive treatment. Previous case reports/series partially mentioned about the treatment response of alopecia areata and mostly it was unsuccessful, especially for scalp hairs. The prognostic course of alopecia is unpredictable and variable; however, long-duration disease and universalis-type alopecia are known as poor prognostic factors. Despite these factors, our patient's scalp-eyebrow hair was re-growing, as seen in Figure 1.

It seems that co-occurrence of ocular MG and alopecia universalis is not a part of thymoma- related disease, which generally occur in alopecia areata, and despite that, there were negative prognostic factors related with alopecia universalis; immunosuppressive treatment with steroids could be an option for alopecia universalis.

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