

Case Report

Dermatomyositis and Myasthenia Gravis: An Uncommon Association With Therapeutic Implications[☆]



Clara Sangüesa Gómez, Bryan Josué Flores Robles, Clara Méndez Perles, Carmen Barbadillo, Hildegarda Godoy, José Luis Andréu*

Servicio de Reumatología, Hospital Puerta de Hierro-Majadahonda, Majadahonda, Madrid, Spain

ARTICLE INFO

Article history:

Received 23 July 2014

Accepted 10 October 2014

Available online 21 January 2015

Keywords:

Dermatomyositis

Myasthenia gravis

Inflammatory myopathies

ABSTRACT

The association of dermatomyositis with myasthenia gravis (MG) is uncommon, having been reported so far in only 26 cases. We report the case of a 69 year-old man diagnosed with MG two years ago and currently treated with pyridostigmine. The patient developed acute proximal weakness, shoulder pain and elevated creatine-kinase (CK). He also developed generalized facial erythema and Gottron's papules. Laboratory tests showed positive antinuclear and anti-Mi2 antibodies. Further analysis confirmed CK levels above 1000 U/l. The clinical management of the patient and the therapeutic implications derived from the coexistence of both entities are discussed.

© 2014 Elsevier España, S.L.U. All rights reserved.

Dermatomiositis y miastenia gravis: una asociación infrecuente con implicaciones terapéuticas

RESUMEN

La asociación de dermatomiositis y miastenia gravis (MG) es infrecuente, habiéndose comunicado hasta la actualidad únicamente 26 casos. Se presenta el caso de un varón de 69 años diagnosticado de MG 2 años atrás, en tratamiento con piridostigmina, que inicia cuadro agudo de debilidad muscular proximal, artralgias en hombros y elevación de creatinasa (CK); así como aparición de eritema facial generalizado y pápulas de Gottron. En el estudio de laboratorio se evidenció positividad de anticuerpos antinucleares y anti-Mi2. Ulteriores determinaciones de CK mostraron niveles por encima de 1.000 U/l. Se discute el manejo clínico de este paciente y las implicaciones terapéuticas que plantea la coexistencia de ambas entidades.

© 2014 Elsevier España, S.L.U. Todos los derechos reservados.

Introduction

Idiopathic inflammatory myopathies (IIM), including dermatomyositis (DM), are a heterogeneous group of systemic autoimmune diseases whose main clinical feature is predominantly proximal muscle weakness.¹ They are rare diseases, with an estimated annual incidence of 6 cases/10⁵ individuals² and may be associated with other autoimmune diseases such as lupus or Sjogren's syndrome.

Another autoimmune disease that causes muscle weakness, myasthenia gravis (MG), can also be associated with the IIM.^{3–9} MG is rare, with an estimated prevalence of 140 cases/10⁶ inhabitants.¹⁰ Its pathogenesis derives from the compromise resulting from the binding of autoantibodies to proteins involved in the neuromuscular transmission signaling. Recognizing the association between the two entities is important due to its therapeutic implications. We present a case of DM in a patient previously diagnosed with MG.

Clinical Case

The patient was a 69 year-old-male Caucasian, diagnosed two years prior with MG, positive to anti-AChR antibodies and treated with pyridostigmine (60 mg/8 h). He was referred to our center due to the appearance of weakness which appeared a month

[☆] Please cite this article as: Sangüesa Gómez C, Flores Robles BJ, Méndez Perles C, Barbadillo C, Godoy H, Andréu JL. Dermatomiositis y miastenia gravis: una asociación infrecuente con implicaciones terapéuticas. Reumatol Clin. 2015;11:244–246.

* Corresponding author.

E-mail address: jlandreu@arrakis.es (J.L. Andréu).



Fig. 1. Gottron's papules.

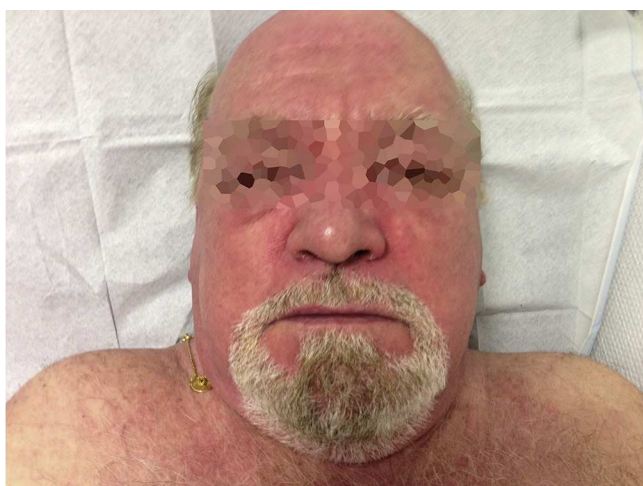


Fig. 2. Facial and neckline erythema.

before, manifested by difficulty dressing or getting out of bed, shoulder joint pain and elevated creatine kinase (CK). He had presented diffuse facial erythema for 6 months. The physical examination showed cuticular thickening and bleeding, Gottron's papules (Fig. 1) and facial erythema and on the neckline (Fig. 2). He had proximal muscle weakness in the upper and lower limbs (4/5). The rest of the examination was normal.

The laboratory exams highlighted CK 1254 U/l. Antinuclear antibodies were positive (>1/160, with a granular pattern, and anti-ENA and anti-DNA were negative) as well as the anti-Mi2. Nailfold capillaroscopy showed a pattern suggestive of DM (Fig. 3).

Electromyography showed a myopathic pattern with proximal predominance in the upper limbs. A muscle biopsy was not considered necessary. Given the high sensitivity of the test for occult neoplasia, a PET-CT was performed and was normal.

With the diagnosis of DM, prednisone (1 mg/kg/day) was started. After three weeks, significant improvement of skin lesions was observed, but muscle weakness persisted, with added dysphagia. CK levels increased to 3800 μ /l. Weekly methotrexate (h 20 mg/week SC) and a course of high-dose intravenous immunoglobulins was added to treatment. The patient showed improvement in muscle strength and swallowing, as well as disappearance of Gottron's papules, with gradual decrease in CK.

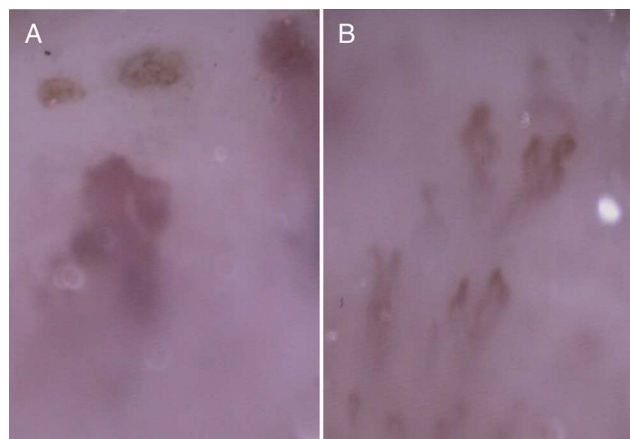


Fig. 3. Capillaroscopy showing bleeding in multiple areas, capillary thrombosis and two large megacapillaries (A) and moderate-severe expansion alternating with single minimal dilations and simple tortuosity (B) without avascular areas.

Discussion

The association of MG to MII is very rare, with 26 cases^{3–9} having been communicated until the present. It is important to know of this association because it has therapeutic implications. Unlike patients with IIM, MG presents with fluctuating weakness that worsens with activity and during the course of the day. In addition, a real muscle fatigability in IIM is not characteristic, while it is a typical clinical characteristic of MG. In most patients with MG, the eye muscles are initially affected, causing intermittent diplopia and ptosis, symptoms that are not observed in the IIM. Proximal muscle weakness is a common symptom of MG and in DM, appearing with bulbar symptoms such as dysphagia or dysarthria.

Treatment of MG includes anticholinesterase although not all patients respond. As in IIM, glucocorticoids, immunosuppressants and immunomodulators–intravenous immunoglobulin–are frequently used in MG. The usual pattern of steroid use in DM is high prednisone doses (1 mg /kg/day) with subsequent gradual tapering. However, in MG it is recommended to start with a low dose and gradually increase due to risk of exacerbation of weakness.¹⁰

In summary, MG may precede or complicate the course of IIM. Knowing this association is important because it requires different handling and treatment, with a gradual intensification of therapy with glucocorticoids.

Ethical Responsibilities

Protection of people and animals. The authors declare this research did not perform experiments on humans or animals.

Data confidentiality. The authors declare that they have followed the protocols of their workplace regarding the publication of patient data.

Right to privacy and informed consent. The authors have obtained the informed consent of patients and/or subjects referred to in the article. This document is in the possession of the corresponding author.

Conflict of Interest

The authors have no conflict of interest to state.

References

1. Dimachkie MM, Barohn RJ. Idiopathic inflammatory myopathies. *Semin Neurol.* 2012;32:227–36.
2. Furst DE, Amato AA, Iorga SR, Gajria K, Fernandes AW. Epidemiology of adult idiopathic inflammatory myopathies in a U.S. managed care plan. *Muscle Nerve.* 2012;45:676–83.
3. Vasilescu C, Bucur G, Petrovici A, Florescu A. Myasthenia in patients with dermatomyositis: clinical, electrophysiological and ultrastructural studies. *J Neurol Sci.* 1978;38:129–44.
4. Hassel B, Gilhus NE, Aarli JA, Skogen OR. Fulminant myasthenia gravis and polymyositis after thymectomy for thymoma. *Acta Neurol Scand.* 1992;85:63–5.
5. Raschilas F, Mouthon L, Andre MH, Azorin J, Couvelard A, Guillevin L. Concomitant polymyositis and myasthenia gravis reveal malignant thymoma. A case report and review of the literature. *Ann Med Interne (Paris).* 1999;150:370–3.
6. Diaco M, Ancarini F, Montalto M, Verrechia E, Evoli A, Servidei S, et al. Association of myasthenia gravis and antisynthetase syndrome: a case report. *Int J Immunopathol Pharmacol.* 2004;17:395–9.
7. Van de Warrenburg BP, Hengstman GJ, Vos PE, Boerman RH, ter Laak HJ, van Engelen BG. Concomitant dermatomyositis and myasthenia gravis presenting with respiratory insufficiency. *Muscle Nerve.* 2002;25:293–6.
8. De Reuck J, Thiery E, de Coster W, van Der Eecken H. Myasthenic syndrome in polymyositis. *Eur Neurol.* 1976;14:275–84.
9. Paik JJ, Corse AM, Mammen AL. The co-existence of myasthenia gravis in patients with myositis: a case series. *Semin Arthritis Rheum.* 2014;43:792–6.
10. Gilhus NE. Myasthenia and the neuromuscular junction. *Curr Opin Neurol.* 2012;25:523–9.