Lagophthalmos as a presenting sign in dermatomyositis with muscle involvement limited to the ocular muscles

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INTRODUCTION

Dermatomyositis (DM) is an autoimmune inflammatory disorder characterized by cutaneous findings and muscle inflammation.[1] Adult patients with DM without clinically significant muscle involvement are classified as having amyopathic DM (DM sine myositis).[2] Ophthalmologic findings associated with DM commonly include a heliotrope rash, periorbital redness, and periorbital edema.[3,4] Other findings can include exophthalmos, chemosis, ptosis, retinopathy, internuclear ophthalmoplegia, and nystagmus.[5,6] Lagophthalmos refers to the inability to fully close the eyelids and can cause increased evaporation of the tear film, corneal exposure, corneal ulceration, and exposure keratopathy.[7] There are many etiologies of lagophthalmos, and the most common causes include facial nerve palsy, trauma, surgery, or infection.[7] We present a unique case of bilateral lagophthalmos in DM with muscle involvement limited to the ocular muscles, which later resolved with medical treatment.

Abbreviation used:

DM: Dermatomyositis

CASE REPORT

A 68-year-old woman with lagophthalmos was referred by her ophthalmologist for a 7-month history of a persistent skin eruption that acutely developed over her face and upper chest.

At an outside clinic, the diagnosis of polymorphous light eruption was suggested, and she was treated unsuccessfully with adapalene (Differin; Galderma Laboratories, L.P.) cream. She denied any history of aphthous ulceration, acral ulceration, difficulty swallowing, Raynaud's phenomena, alopecia, or photosensitivity reactions. She also denied a history of sunbathing, living in the tropics, or having worked outdoors extensively. There were no systemic findings or relevant family history. The patient did not report any swollen joints or muscle weakness and primarily complained of itchiness and redness around the eyes and anterior chest. She had periocular erythema and erythema on the anterior chest without any Gottron's sign, papules, or shell sign and a normal musculoskeletal examination. She had a history of hypothyroidism and rheumatoid arthritis that she reported resolved with a raw food diet. She also had a history of 2 melanomas, one on her back approximately 17-18 years ago and one on her left leg approximately 2-3 years ago. She used levothyroxine (Synthroid; AbbVie, Inc.), vitamin B12, and fish oil and at the time of presentation, artificial tears and Lacri-Lube (petrolatum 56.8% and mineral oil 42.5%, AbbVie, Inc.) for dry eyes. She had no known allergies to medications. Physical examination found poikilodermatous changes on her face, neck, and upper torso and subtle skin induration of the face and cape area (Fig 1). She had significant dermal atrophy of the eyelids, which were virtually transparent with some sclerosis, and she was unable to close her eyes completely (Fig 2). She had no periorbital edema or exophthalmos. There was no apparent decrease in the size of the oral opening or sclerodermoid changes. No abnormalities were found upon dermoscopy of the nail folds

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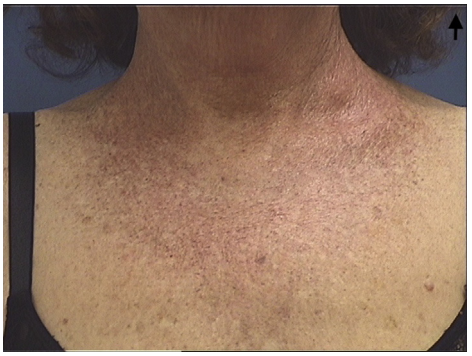

Fig 1. Dermatomyositis with muscle involvement limited to the ocular muscles. Clinical picture showing poikilodermatous changes on the neck and upper torso, and subtle induration of the skin.

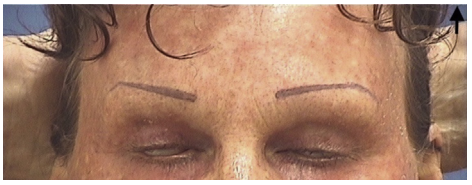

Fig 2. Dermatomyositis with muscle involvement limited to the ocular muscles. Clinical picture showing lagophthalmos.

and there was no alopecia or active arthritis detected. The remainder of the physical examination was unremarkable. A previous pathology report showed an interface dermatitis, lichenoid type with vacuolar changes on skin biopsy of the sternum and marker studies were negative for the lymphoproliferative disorder (Figs 3 and 4). The following investigations were within normal limits: complete blood count, electrolyte panel, creatinine, ferritin, liver function tests, creatine kinase, lipid panel, thyroid-stimulating hormone, complements C3 and C4, and an unremarkable fasting glucose and hemoglobin A1c. She had a negative extractable nuclear antigen, rheumatoid factor, Anti-Jo-1 antibody, antineutrophil cytoplasmic antibodies, and anti-cyclic citrullinated peptide antibodies. Her antinuclear antibody results were deemed clinically insignificant at the titer of 1:80. Computed tomography scan of the chest, abdomen, and pelvis was normal and her bilateral mammogram was unremarkable. Adapalene was discontinued and she was examined by a neurologist a month later. She did not have head drop, dysphagia, ptosis, or diplopia. CN I-XII were normal, and she had equal and bilateral 5/5 upper and lower extremity strength. Reflexes were 2+ throughout

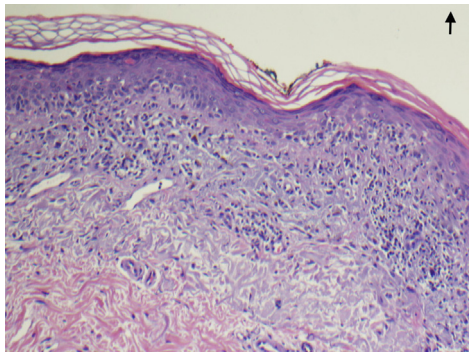

Fig 3. Dermatomyositis with muscle involvement limited to the ocular muscles. Histologic picture depicting interface dermatitis, lichenoid type with vacuolar changes. (Hematoxylin-eosin stain.)

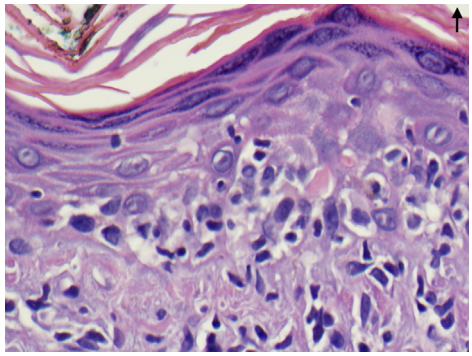

Fig 4. Dermatomyositis with muscle involvement limited to the ocular muscles. Histologic picture depicting interface dermatitis, lichenoid type with vacuolar changes. (Hematoxylin-eosin stain.)

and her electromyography nerve conduction studies were unremarkable. The resulting diagnosis of DM with muscle involvement limited to the ocular muscles was provided. The patient then was started on hydroxychloroquine 200 mg twice daily and responded positively. She was assessed by a rheumatologist 3 months later and hydroxychloroquine was reduced to a maintenance dose of 200 mg daily and the patient's symptoms remained under control and without relapse. Approximately 2 years later, she was weaned to hydroxychloroquine 100 mg on alternating day doses, which led to a slight exacerbation of the rash on her anterior chest and she reported mild weakness. Repeat electromyography nerve conduction was again normal, and hydroxychloroquine was increased to 200 mg daily. Four years after her initial presentation, her symptoms remain under control with oral hydroxychloroquine 200 mg twice daily. While she continues to suffer from dry eyes managed by an eye lubricant, she has no evidence of active DM or lagophthalmos.

DISCUSSION

Ocular involvement in cutaneous connective tissue diseases, such as DM, while rare, may be the first presentation of systemic disease and warrants prompt evaluation.[8] To our knowledge, we report the first case of medically resolved lagophthalmos in DM with muscle involvement limited to the ocular muscles. Typically, the ocular muscles are considered to be spared in DM, even in advanced and/or untreated cases.[5] While extraocular muscle imbalances may occur, to our knowledge, there have been no reports of lagophthalmos as a direct ocular manifestation of DM. Furthermore, lagophthalmos is found uncommonly in inflammatory myopathies and there is virtually no literature surrounding medically resolved lagophthalmos in DM sine myositis. One case described what the investigators claim as the first report of left-sided lagophthalmos, right-sided ptosis, and complaints of epiphora and eye irritation in a patient later diagnosed with inclusion body myositis, an inflammatory myopathy.[9] Our patient clearly differs from the above finding as she does not have inclusion body myositis. Another case mentioned lagophthalmos in a patient with DM with muscle inflammation. However, the affected eye had a lower lid ectropion presumably secondary to DM, which likely contributed to the lagophthalmos.[10] As well, the lagophthalmos was not described to have fully resolved with medical treatment alone. In conclusion, this case demonstrates the importance of a thorough investigation, evaluation, and management in a patient presenting with lagophthalmos. It also describes a unique case of DM that presented with lagophthalmos that resolved without surgery.

Conflicts of interest

None disclosed.

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