An elusive case of dermatomyositis

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Abstract

Dermatomyositis is an inflammatory myopathy of unknown aetiology. Muscle involvement may eventuate later in the disease course in some patients, who may present with typical skin disease without clinical signs of myopathy and are referred to as dermatomyositis sine myositis. A 48 year old female presented with intermittent urticaria like rashes, diffuse asymmetrical swelling of proximal limbs, pain in small joints of hands and fatiguability. Initial laboratory work-up for immune markers was negative. Three years later, she developed heliotrope rash and periorbital oedema with no evidence of muscle weakness and was labeled as amyopathic dermatomyositis. After an interval of one year, she developed profound weakness and significantly raised CPK. Patient responded well to steroids and Azathioprine and improved both clinically and biochemically.

Keywords: Dermatomyositis, Amyopathic dermatomyositis, Urticaria Dermatomyositis sine myositis.

Introduction

Dermatomyositis (Dm) is an idiopathic inflammatory myopathy with prototypic skin lesions. The estimated incidence is 9.63 cases per million population.[1] Amyopathic dermatomyositis (ADm), also known as dermatomyositis sine myositis, is a rare variant of Dm. These patients have pathogonomic cutaneous lesions but no clinically evident muscle involvement. We report an unusual case of dermatomyositis, where myositis was a late development in the course of disease, which evolved over a 4 year period. Informed, verbal consent was taken from the patient for publication of this case.

Case Presentation

A 48 year old lady, first presented in 2008 at the Rheumatology clinic, Jinnah Postgraduate Medical Centre, Karachi, with history of intermittent urticarial rashes, diffuse asymetrical swelling of limbs and pain in small joints of hands as well as some large joints, with morning stiffness and fatigue for one year. There was no history of allergy, alopecia, oral or genital ulcers, photosensitivity, any eye symptoms, as well as any history suggestive of myopathy. The only medication in use was an anti-allergic (cetrizine). Patient was obese, general physical and systemic examination was normal, except mildly swollen and tender ankle joints, the rest of musculoskeletal examination was unremarkable. There was no rash or signs of wasting or proximal muscle weakness. Initial impression of Systemic Lupus Erythematosus (SLE) or sarcoidosis was made. Erythrocyte Sedimentation Rate (ESR) and C. Reaction Protein (CRP) were raised, while Anti Nuclear Antibodies (ANA), Rheumatoid Arthritis Factor (RA factor), Lactate Dehydrogenase (LDH), Creatine Phosphokinase (CPK), Angiotensin Converting Enzyme (ACE) levels and thyroid function tests were negative (Table). She was only prescribed Non-Steroidal Anti Inflammatory Drugs (NSAIDs). One year later, i.e. 2009, she presented again with the resurgence of same complaints, having been reasonably well in the intervening period. RA factor was positive (182.7 U/L) on this occasion. NSAIDs and Disease Modifying Antirheumatic Drugs (DMARDs) (Leflunomide 20 mg daily and Hydroxychloroquine 200 mg twice daily) were started. She remained well on this treatment for one year and then stopped treatment on her own. Her next

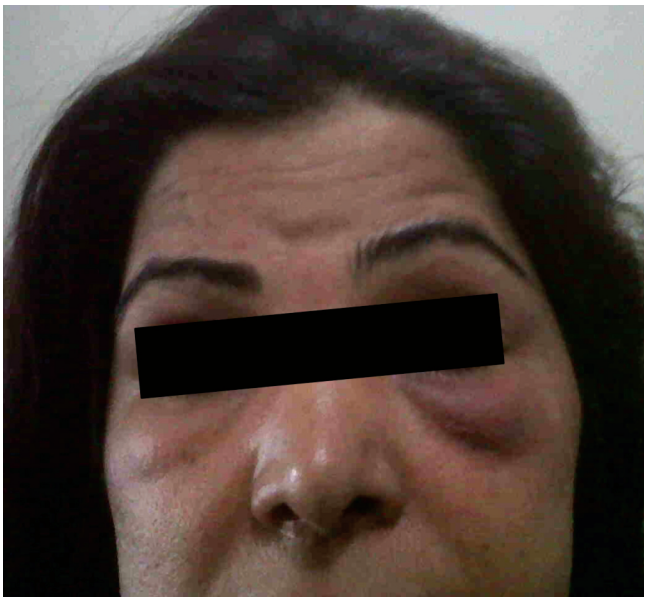

Figure: Heliotrope rash and peri-orbital oedema.

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Table: Laboratory investigations.

	Normal Ranges	Patient's value			
		Apr2008	Feb 2009	Aug 2011	Nov 2011
Hb	12-16g/dl	11.75	12.5	-	12.8
WBC	4000-10000/cmm	8.9	6.4	-	7.2
Platelets	150-400/cmm	441	342	-	302
ESR	0-10 mm/1stHr	64.9	30	18	76
CRP	<5mg/dl	27	-	1.3	23
ALT	15-56 U/L	34	-	-	29
ANA	-	Negative	Negative	Negative	Negative
RA factor	< 14 U/L	Negative	182.4	-	-
TSH	< 30	4.1	-	-	-
Vitamin D	>30 ng/ml	12.6	27.4	-	-
Ca	8.5-10.5 mg/dl	8.9	8.6	9.9	-
CPK	90-140 U/L	43	-	115	2900
Serum ACE level	8-53 µL	41	-	-	-
Cr	0.7-1.1 mg/dl	0.9	0.9	1.0	0.9

Hb: Haemoglobin.
WBC: White Blood Cells.
ESR: Erythrocyte Sedimentation Rate.
CRP: C-Reactive Protein.
ALT: Alanine Aminotransferase.
ANA: Antinuclear Antibodies.
RA factor: Rheumatoid Arthritis Factor.
TSH: Thyroid Stimulating Hormone.
Ca: Calcium.
CPK: Creatine Phosphokinase.
ACE: Angiotensin Converting Enzyme.
Cr: Creatinine.

presentation was with two episodes of asymmetrical periorbital swelling with dusky pigmentation, which lasted for two months each time. Two years later, i.e. August 2011, she again developed a more marked periorbital swelling with purplish discoloration of periorbital area (Figure). Joints and other systemic examination were normal. All immunological markers were repeated and were again normal. She went on to develop oral ulcers, submandibular swelling and circinate papular lesions on both shins. Biopsies of skin, submandibular gland and conjunctiva remained inconclusive. A depot injection of Methylprednisolone was given without a firm diagnosis, which resulted in some relief with regard to the eyes. After an interval of four months, i.e. November 2011, she presented with profound muscle weakness and had also by then developed an erythematous maculo-papular rash of 'shawl' distribution. This was associated with hoarsness, dysphagia and inability to swallow saliva properly, with a need to continuously spit. In addition, muscles of both arms were weak, swollen and stiff. This time she was admitted in hospital. On this occasion, CPK level was markedly elevated at 2900 iu/ml. A diagnosis of dermatomyositis sine myositis was finally made. Intravenous pulse Methylprednisolone 500mg/day was given for 3 days followed by oral steroid (Prednisolone 30mg/day) and Azathioprine (AZA) (100mg/day). Screening for underlying malignancy was negative. Patient improved on treatment, and inflammatory markers i.e. ESR, CRP and CPK normalized. Steroid was tapered off over a six month period, while AZA was continued for 3 years. Patient was last followed in 2015, was well, off drugs with normal CPK and inflammatory markers and without any further episodes of dermal lesions or myositis.

Discussion

Dermatomyositis sine myositis or ADm is characterized by typical cutaneous disease, with no clinical or laboratory evidence of muscle disease. This rare variant of Dm has a reported incidence of 2.08 cases per million population.[1]

The amyopathic nature of disease course in this case posed a difficulty in establishing the diagnosis, which was ultimately clinched when myositis developed late in the course of disease. Joint manifestations are seen in 15-30% of patients of Dm. This patient had non-erosive arthritis and later her Rheumatoid factor (RF) also became positive. A positive RF can be seen in upto 20% of patients

with Dm, however, an overlap syndrome with other connective tissue diseases is not infrequent. Major associations are seen with scleroderma, SLE, rheumatoid arthritis and thyroiditis. This patient was unique, in that she had recurrent urticaria, which is a very rare manifestation of Dm, whereas other autoimmune disorders like SLE, dermatomyositis, polymyositis, Still's disease have an association with chronic urticaria.[3] A case of Dm with urticarial vasculitis has been reported in a patient of nasopharyngeal carcinoma.[4] Eye lid oedema in association with erythema and telangiectasia is a common feature of dermatomyositis. Initially it was overlooked, as this patient had neither signs of myopathy nor the classical cutaneous features of Dm. ADm has a variable prognosis and disease course. This patient remained well for entire two years except she had episodes of periorbital swelling. The average duration of cutaneous disease has been reported as 3.7 years in patients with clinically ADm.[5] Another study has reported 27 cases of ADm, whereby, only 2 out of 27 developed myopathy within 5 years of illness.[6] Sparse data is available to reflect the incidence and disease course of Dm from Pakistan, with only two cases of juvenile Dm having been reported and no reported case of ADm.[7,8] An Italian study reported 13 cases of ADm out of 157 of Dm over a 10 year period.[9] This patient fits into the category of group-2 ADm according to that described by Stonecioher et al, as she had a cutaneous disease at baseline and subsequent evolution into myositis.[10]

Conclusion

This case represents a distinct and rarely reported variant of Dm, i.e dermatomyositis sine myositis or ADm that ultimately evolved into clinical myositis. A high index of suspicion and close follow-up are mandatory to pick-up this rare variant of Dm and subsequently institute correct and potentially life-saving treatment.

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