PICTORIAL CME

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Amyopathic Dermatomyositis

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A 48-year-old lady presented in rheumatology clinic with complaints of periorbital rash, swelling of face, rashes over body, polyarthralgia and extreme fatigue for last seven months. There was no history of any motor weakness, dyspnoea or any drug intake. On examination: heliotrope rash, Shawl sign, V sign and Gottron's papules were present (Fig. 1 - 4). There was no muscle weakness and chest examination was normal. Blood investigation showed normal haemogram and biochemical parameters. Anti-nuclear antibody (ANA) test was positive with a titre of 1:320 and homogenous pattern. An extractable nuclear antigen (ENA) panel, CPK and LDH levels were normal. However, since patient had strong clinical features of dermatomyositis, we got a muscle biopsy done which was also normal. Subsequently, a diagnosis of amyopathic dermatomyositis was made and screening tests for occult malignancy was done, which came negative. We initially gave her prednisolone at dose of 1 mg/kg/day for 6 weeks and was gradually tapered off. Azathioprine and hydroxychloroquine was also started simultaneously with steroids and patient was followed up monthly. After 1 month of therapy patient started showing considerable improvement and her skin lesions started resolving. After

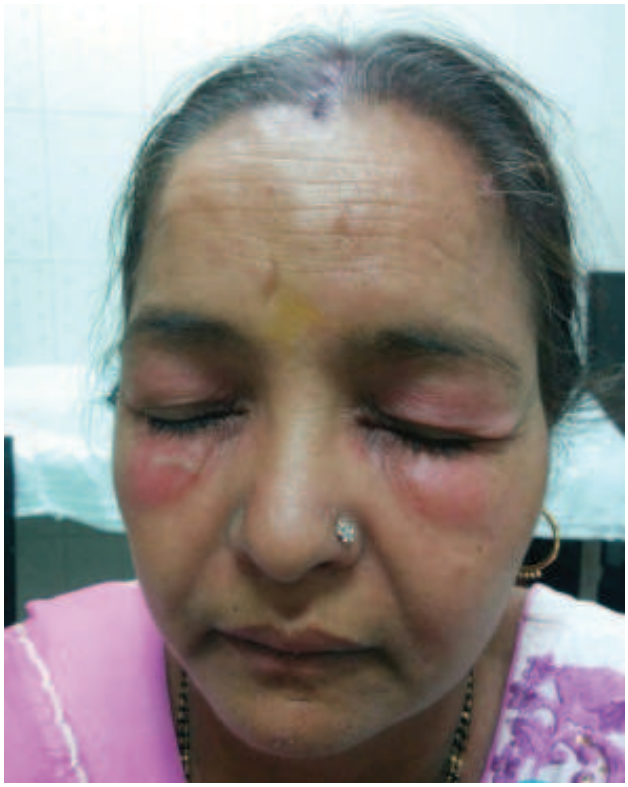

Fig. 1: *Heliotrope rash.*

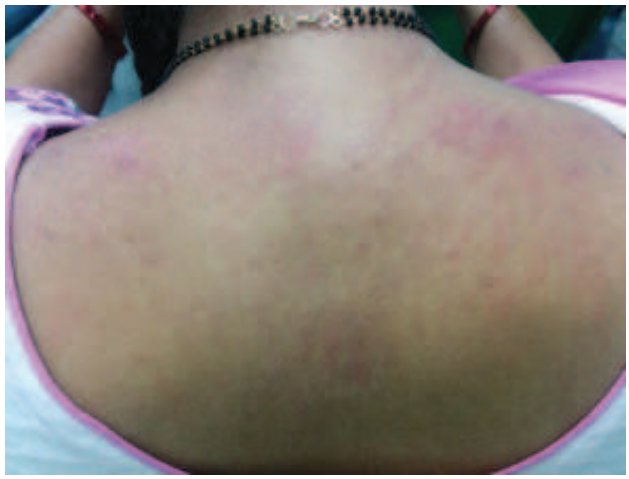

Fig. 2: *Shawl sign.*

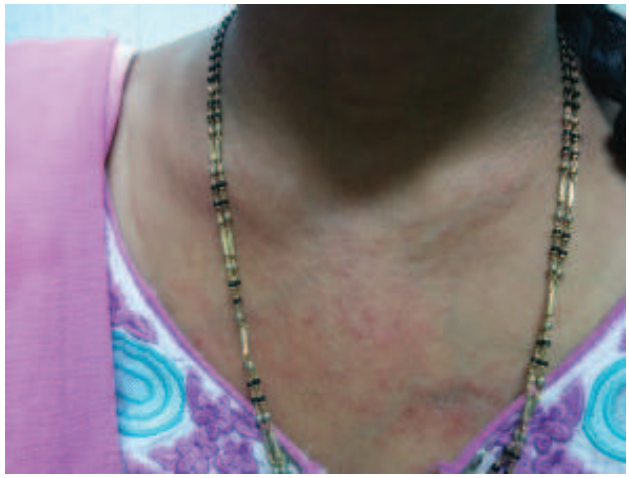

Fig. 3: *V sign.*

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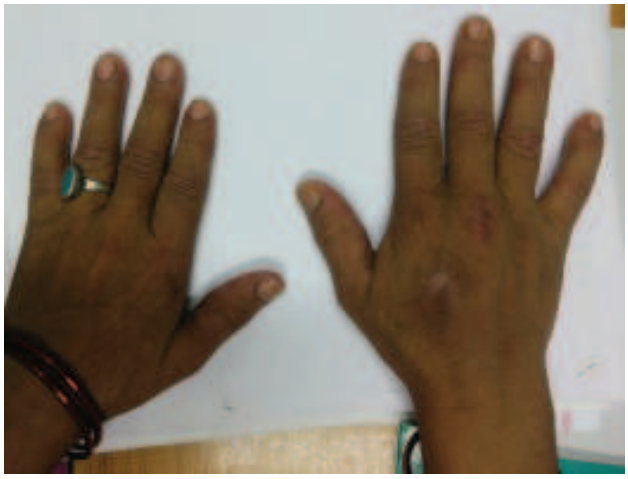

Fig. 4: *Gottron's papule.s*

three months her polyarthralgia and skin lesions completely resolved (Fig. 5).

Dermatomyositis (DM) is an idiopathic inflammatory myopathy characterised by classical skin manifestations with clinical features of muscular weakness. Amyopathic dermatomyositis (ADM) is a subset of DM when classical skin manifestations are present without any myositis[1].The term hypomyopathic DM is used when patients have skin

Fig. 5: *Resolution after 3 months of treatment.*

involvement with subclinical myositis which can be demonstrated by raised muscle enzymes, muscle biopsy or magnetic resonance imaging. Recently, clinically amyopathic dermatomyositis (CADM) term has been proposed to include patients of ADM and hypomyopathic dermatomyositis[2]. 10 - 30 per cent patients of DM have CADM. The classical skin manifestations in DM include: Gottron's papules – erythematous papule over metacarpophalangeal and proximal interphalangeal joints; V and Shawl sign – violaceous rash over upper chest and back respectively; Heliotrope rash – erythematous rash over eyelids with periorbital oedema; Holster sign – poikiloderma on lateral side of thigh; Mechanic's hand – hyperkeratosis over lateral aspect of palm and Calcinosis cutis – calcium deposition in skin. For diagnosis of ADM these skin manifestations should be present for at least six months in absence of any clinical and/or laboratory evidence of myositis. Patients of DM have 5.5 times increased risk of malignancies but risk is comparatively lower in ADM.There are multiple reports of interstitial lung disease (ILD) in ADM and risk of rapidly progressing ILD increases when anti-MDA5 (melanoma differentiation-associated protein) antibodies are present[3].

There is no consensus regarding treatment of ADM. Hydroxychloroquine and topical steroids are initially used for cutaneous manifestations; however, aggressive treatment with immunosuppressants are recommended when there is associated ILD, excessive fatigability or if classical DM develops[4]. A close follow-up of ADM patients are needed as half of them develop classical DM over a period of time.

References

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