A case of dermatomyositis with underlying unusual malignancy

Sir,

Dermatomyositis is a rare autoimmune idiopathic inflammatory myositis with an incidence of about 1/100,000 population.[1] Peri-orbital edema is commonly seen in patients with dermatomyositis, while with severe inflammatory involvement, edema can also be seen at other sites such as limbs and trunk. Rarely, anasarca may be the presenting manifestation of dermatomyositis. Various theories proposed for the pathogenesis of subcutaneous edema include the immune complex-mediated vasculitis, complement activation and vascular endothelial damage, leading to increased vascular permeability and edema in the tissues and muscles.[2] Subcutaneous edema is also considered by some to be a marker of very active inflammation suggesting a poor prognosis. A search of the literature yielded about twenty cases reported previously where patients

presenting with anasarca rapidly developed other features of dermatomyositis.[3] Out of these, only five cases had an underlying carcinoma (endometrial, prostate, colon, gastric and cervical).

We report the case of a 55-year-old woman, who presented to the casualty with generalized edema for 2 months. This was followed by marked peri-orbital edema and itchy red lesions on the face, neck, chest, shoulders, thighs and buttocks. Though initially there was no muscle weakness, within 2 weeks, she developed rapidly worsening proximal muscle weakness of both the upper and lower extremities. On physical examination, there was peri-orbital edema along with pedal edema [Figure 1]. Cutaneous examination showed the presence of macular violaceous erythema over the peri-orbital area (heliotrope rash), forehead, nasolabial folds, pinnae, V area of the chest (shawl sign), left shoulder, buttocks, thighs (holster sign), elbows and knees (Gottron's sign) [Figure 2]. There were no Gottron papules. Serum creatine phosphokinase (634 U/L [50–200]) and lactate dehydrogenase (1954 U/L [220–600]) were elevated. Muscle biopsy from the triceps showed chronic inflammatory infiltrate between the muscle fibers. Skin biopsy from erythematous plaque showed interface dermatitis with perivascular and peri-follicular inflammatory infiltrate and abundant mucin deposition in the dermis. Lesional direct immunofluorescence showed linear positivity of immunoglobulin G (2+) and C3 (1+) along the dermoepidermal junction. On indirect immunofluorescence, antinuclear antibody was positive in a diffuse pattern (3+), anti-double-stranded DNA antibody (dsDNA) was positive (2+, 100 IU/ml) and anti-Jo-1 antibodies were negative. Ultrasound abdomen revealed cholelithiasis with a polypoidal iso to hypo-echoic lesion adherent to the posterior wall of the gall bladder of suspected malignant etiology. Further evaluation by a contrast-enhanced computed tomography thorax, abdomen and pelvis showed cholelithiasis with enhancing soft tissue lesion (size 20x11mm) at fundo-posterior wall of gallbladder likely to be of malignant etiology. Hematological and biochemical investigations revealed raised erythrocyte sedimentation rate (70 mm in the 1st hour) and positive C-reactive protein with hypoalbuminemia (2.8 mg/dl, [normal 3.5–5 mg/dl]). Based on the above findings, a presumptive diagnosis of dermatomyositis with gallbladder malignancy was made.

Paraneoplastic dermatomyositis accounts for 15%–30% of all the cases of dermatomyositis, and it may precede (40%), develop concomitantly (26%) or follow (34%) the diagnosis of the malignancy.[4] Most commonly associated malignancies are ovarian, bronchogenic, colorectal, gastric, non-Hodgkins lymphoma, breast, cervical, pancreatic, esophageal, bladder and renal malignancies. It has been hypothesized that the malignant cells express certain cryptic antigens on their cell surface, generating autoantibodies which cross react with the muscle cells to cause myositis.[5] The risk factors for associated malignancy are old age, male sex, smoking, cutaneous leukocytoclastic vasculitis, cutaneous necrosis, lesions resistant to therapy, a rapid onset of myositis, dysphagia, raised erythrocyte sedimentation rate, C-reactive protein, raised creatine kinase, positive anti-transcription intermediary factor 1γ, positive human leukocyte antigen-A28 and negative anti-Jo-1. Gallbladder malignancy is extremely rare with dermatomyositis. To the best of our knowledge, only eight such cases have been reported till date which are illustrated in Table 1. In all the previously reported cases, the malignancy was found after the diagnosis of dermatomyositis was made, as seen in our case.

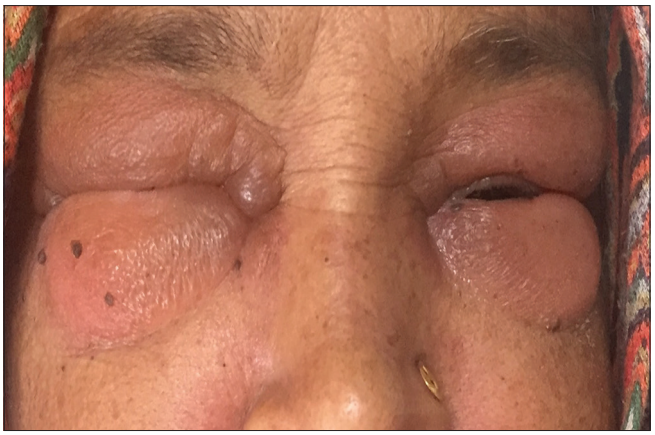

Figure 1: Severe periorbital edema

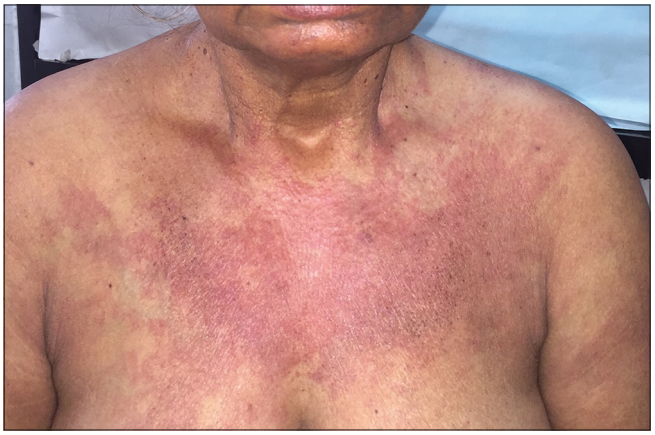

Figure 2: Violaceous erythema over front of chest (V sign)

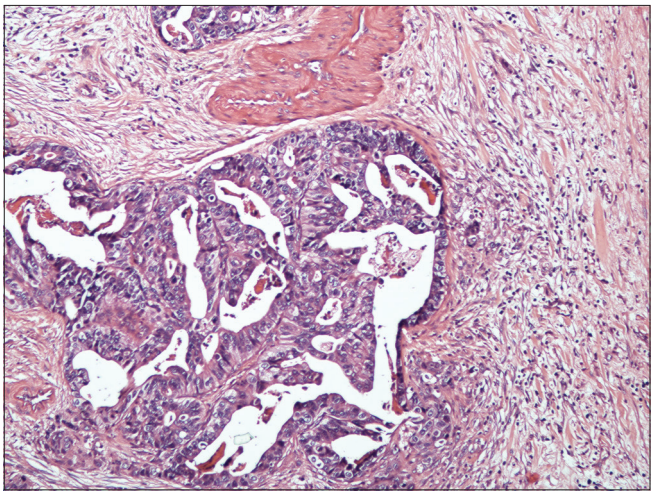

Figure 3: Microphotograph revealing well differentiated adenocarcinoma of gallbladder (H and E, ×400)

Our patient was treated with oral prednisolone (40 mg daily), pulsed intravenous methylprednisolone, hydroxy-chloroquine (200 mg twice daily), antihistamines and topical mometasone furoate 0.1%. She underwent an extended cholecystectomy which on histopathological examination revealed a well-differentiated

Table 1: Previously reported cases of diabetes mellitus with gallbladder malignancy

Authors	Year	Age/sex	Place	Clinical sites	Muscle involved	Treatment	Follow-up
Yiannopoulos *et al*.[6]	2002	75/female	The Netherlands	Heliotrope rash, Gottron's papules	Proximal muscles, neck	Steroids, inoperable at exploratory laparotomy	Metastasis, lower lobe pneumonia, death
Kundu *et al*.[7]	2005	44/male	India	Face, limbs	Proximal muscles	Steroids	Lost to follow-up
Narasimhaiah *et al*.[8]	2011	65/female	India	Heliotrope rash, V sign, shawl sign	Proximal muscles	Death before operation	Death
Ni *et al*.[9]	2013	67/female	China	Heliotrope rash, Gottron's sign	No muscle weakness	Resection of gallbladder	Resolution
Sawada *et al*.[10]	2014	90/female	Japan	Heliotrope rash, Gottron's papules	Proximal muscles	Steroids	Palliative care
Park *et al*.[11]	2014	71/male	Korea	Heliotrope rash, Gottron's papules/sign, V and shawl sign	Proximal	Steroids, chemotherapy, inoperable cancer	Acute renal failure, death
Premkumar *et al*.[12]	2014	47/female	India	Heliotrope rash, Gottron's papules/sign	Proximal muscles	Steroid, gemcitabine, inoperable	Regular follow-up
Jurcic[13]	2015	48/female	Croatia	Face, neck, ear	Stiffness, dysphagia	Steroid, gemcitabine, cisplatin	Palliative care, status same

adenocarcinoma gallbladder stage T1b Nx [Figure 3]. With this treatment, the patient's muscle weakness improved clinically and skin lesions started regressing. The patient was also started on oral methotrexate 7.5 mg weekly.

This case illustrates an unusual malignant association with dermatomyositis and highlights the importance of increased suspicion and heightened screening for malignant disease in elderly patients with dermatomyositis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given her consent for her images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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Quick Response Code:	Website: www.ijdvl.com
	DOI: 10.4103/ijdvl.IJDVL_365_16

How to cite this article: Garg T, Sanke S, Chander R, Agarwal M, Agarwal K, Kumar A. A case of dermatomyositis with underlying unusual malignancy. Indian J Dermatol Venereol Leprol 2017;83:473-6.

Received: August, 2016. **Accepted:** September, 2016.