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CASE REPORT

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Dermatomyositis and cutaneous necrosis: report of five cases

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Abstract We report on five patients with dermatomyositis (DM) and cutaneous necrosis. Patients presented with classic DM skin eruptions, mild myositis, and a high incidence (4/5) of interstitial pneumonia. Cutaneous necrosis developed independently of steroid therapy, with the majority of lesions being cured following several months of sterilization treatment. In addition, one patient with accompanying cancer presented with multiple necrotic lesions. Topical treatment using gentiana violet against local infection was considered to have been essential in accelerating healing.

Key words Dermatomyositis · Cutaneous necrosis · Cutaneous ulcer

Introduction

Dermatomyositis (DM) is characterized by inflammatory myopathy and typical skin eruptions. In addition to classic eye-lid erythema and Gottron's papula, patients with DM present with a variety of cutaneous manifestations, including erythema on extensor surfaces of knee or elbow joints, V-shaped neck eruptions, bronze-like pigmentation, poikiloderma, diffuse pruritic dermatitis, and cutaneous necrosis. Previous studies have reported an association between diffuse pruritic dermatitis[1] or cutaneous necrosis[2] and malignant disease accompanied by DM. Furthermore, a poor prognosis for DM patients with cutaneous necrosis has been suggested.[3] Although cutaneous necrosis might be relatively common in DM patients,[2,3] clinical descriptions of this lesion, in terms of treatment, prognosis, and any relationship to other organ involvement in DM, are rare. Based on our observations of patients with DM, the present study describes five clinical profiles of cutaneous necrosis and evaluates the treatment process.

Case description

Case 1

Polyarthralgia developed 3 months prior to eye-lid erythema and Gottron's papula, followed by mild muscular weakness. Remittance occurred during prednisolone therapy at an initial daily dose of 40 mg. While undergoing maintenance dosage steroid therapy at our outpatient clinic, the patient showed signs of progressive depression and was readmitted to our institute. Two family members had suffered from depression and/or commited suicide. Delirium was followed by newly developed interstitial pneumonia and a recurrence of muscle and cutaneous symptoms. Following intravenous pulse steroid and oral steroid therapy, no evidence of physical or psychiatric manifestations remained. She was diagnosed by a psychiatrist as having transit syndrome. Systemic lupus erythematosus (SLE) was ruled out given that apart from positive serum antinuclear antibodies, arthralgias, and psychosis, she did not fulfill the criteria for SLE.[5] As these symptoms improved, a cutaneous ulcer on her right elbow developed into deep aseptic necrosis with exposure of the tendon, which was partially improved under sterilization. Six months later, on her third admission, she suffered a fatal recurrence of interstitial pneumonia.

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Case 2

Polyarthralgia, medium-grade fever, Gottron's papula, and eye-lid erythema preceded progressive interstitial pneumonia and mild muscle symptoms. A daily dose of 60 mg prednisolone was effective against the pneumonia. During recovery, the patient developed aseptic cutaneous necrosis on the left elbow, very similar to that of case 1. This was cured by sterilization treatment following discharge.

Case 3

Arthritis of the proximal interphalangial finger joints preceded the onset of polyarthralgia, eye-lid erythema, Gottron's papula, and mild muscle symptoms by 5 years. Remittance occurred with prednisolone therapy at an initial daily dose of 40 mg, and although she developed aseptic cutaneous necrosis bilaterally on her elbows, this was cured after several months of follow-up sterilization treatment at the outpatient clinic.

Case 4

Adenocarcinoma of the left breast was diagnosed by surgeons at another hospital, concomitant with proximal muscle weakness, eye-lid erythema, Gottron's papula, and erythema on the extensor surfaces of the knee and elbow joints. Furthermore, the patient presented with delirium of variable severity throughout almost the entire disease course, diagnosed by a psychiatrist as symptomatic psychosis. In addition, none of the ARA criteria for SLE[5] were evident, except for psychosis. After a radical resection was performed to treat the breast cancer, the muscle symptoms resolved rapidly. However, myogenic abnormalities persisted on electromyography. The patient was referred to our hospital following the introduction of prednisolone therapy at a daily dose of 25 mg to treat the cutaneous lesions. After 4 weeks the steroid dose was reduced, whereupon an aseptic cutaneous ulcer appeared on the right elbow, progressing to deep necrosis. During her 15 months of hospitalization at our institute, multiple skin necroses developed on her fingers, elbows, knees, and buttocks (Fig. 1). These lesions were initially aseptic, but later methicillin-resistant *Staphylococcus aureus* was frequently detected in cultures taken from these lesions. One year postoperatively, neither breast cancer recurrence nor metastasis were detected by radiological examination. Despite the fact that the majority of ulcers closed after repetitive sterilization, systemic infections occasionally occured, ultimately resulting in the patient's death. A biopsy from the border of the ulcer on the elbow revealed mild mononuclear cell infiltration into the dermis, which may have been a nonspecific ulcerative reaction. However, no vasculitis was found (not shown).

Case 5

The main symptom upon admission was rapidly progressive interstitial pneumonia, concomitant with eye-lid erythema, erythema on the extensor surfaces of the knee joints, and cutaneous necrosis on the elbow joints. However, no muscle symptoms or muscular enzyme abnormalities were found. Intravenous pulse steroid and oral cyclosporine therapy was ineffective, and a fatal episode of rapid respiratory failure occurred. An autopsy revealed diffuse alveolar damage in the lung.

Patients

The clinical features of the five patients with DM and cutaneous necrosis who were admitted to the Jichi Medical School Hospital are shown in Table 1. We diagnosed three patients as definite DM (cases 1, 2, and 3), one patient as probable DM (case 4) according to Bohan and Peter's criteria,[5] and one patient as amyopathic DM (case 5). In cases 1–4, despite positive myogenic abnormalities detected

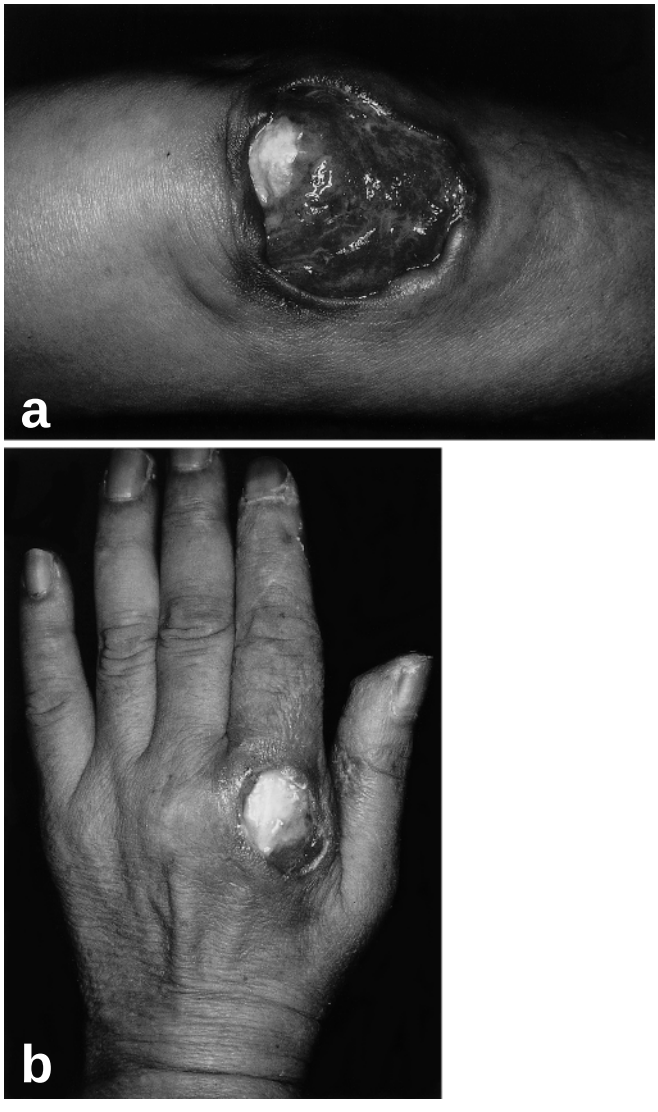

Fig. 1. Multiple cutaneous necrosis on **a** the elbow and **b** the hand in case 4. Similar lesions were also observed on the elbow in cases 1, 2, 3, and 5

Table 1. Clinical features of five patients with dermatomyositis (DM) and cutaneous necrosis

Case	Sex	Age at onset of DM	MMT[a] Enzyme[b] EMG[c] Histology[d]	Interstitial pneumonia[e]	Malignancy	Outcome and disease duration	Antinuclear antibodies	Anti-Jol antibodies
1	F	28	4^+/5 CK 384 Positive Negative	Fatal	–	Respiratory failure[j] 21 months	×80[i] speckled	–
2	F	65	4^+/5 CK 157, Al 13.3 Positive ND	Improved[g]	–	Remission 9 months	–	–
3	F	42	4^+/5 CK 34, Al 8.3 Positive Negative	Mild[h]	–	Remission 9 months	–	–
4	F	57	4^-/5 N/A[f] Positive ND	None	Breast cancer	Sepsis[j] 18 months	–	–
5	M	84	5/5 CK 115 Negative Negative	Fatal	–	Respiratory failure[j] 4 months	×80 speckled	–

F, female; M, male
[a] The lowest score of involved muscles by the manual muscle test (MMT)
[b] Peak level of serum creatine kinase (CK, normal <150 U/ml) or aldolase (Al, normal <5 U/ml)
[c] Myogenic change on electromyogram (EMG)
[d] Inflammatory change on muscle biopsy (cases 1 and 3) or autopsy (case 5)
[e] Defined by bilateral reticular shadows on chest X-ray and interstitial infiltration on computed tomography
[f] Not recorded before referral to us (see text)
[g] %VC from 62 to 60; PaO_2 from 64 mmHg to 98 mmHg
[h] %VC 79; PaO_2 88 mmHg
[i] Anti-DNA antibody (Ab), anti-Sm Ab, anti-RNP Ab, anti-SSA Ab, and anti-SSB Ab were negative
[j] Deceased

by electromyography, muscle symptoms were mild. In case 4, normal serum creatine kinase levels were detected during steroid therapy upon referral to our hospital (Table 1). DM was accompanied by malignancy only in case 4. An extensive radiological survey at the time of DM diagnosis in cases 1, 2, and 3, and during an autopsy for case 5, failed to detect any malignancies in these cases.

In addition to these five patients, we reviewed comprehensive hospital records of 51 patients with DM admitted to our institute at some point between 1978 and 1998, and searched for cases of cutaneous necrosis. No such cases were found. Of these 51 patients with DM, 20 presented with associated malignancy. In addition, interstitial pneumonia was observed in 19 of 31 patients without malignancy, and in 2 of 20 patients with malignancy. The results for the incidence of interstitial pneumonia in DM patients without malignancy revealed that 4/4 (100%) of the patients with cutaneous necrosis developed pneumonia, whereas 19/31 (61%) of patients without cutaneous necrosis did so. This difference was not significant (Fisher's exact probability test).

Cutaneous necrosis

In all of the present cases, painful erythema developed on the elbows prior to ulceration and destruction of whole layers, from the skin to the fat tissue, followed by pocket formation. Multiple necrosis developed only in case 4, where the closure of one lesion and the recurrence of another were observed over a period of 15 months.

Therapy for cutaneous necrosis

The majority of cutaneous lesions were cured during long-term sterilization and debridement. The efficacy of tretinoin tocoferil ointment for skin regeneration was unclear, but it did increase the risk of local infection. Compared with using iodine, topical use of a 0.1% concentration of bactericidal reagent gentiana violet (Pyoktanin blue) reduced the chances of infection, and accelerated the healing of the ulcer in case 4.

Discussion

The incidence of cutaneous necrosis among DM patients at our institute is relatively high (at least 5/56; 9%). Basset-Seguin et al.[2] stated that this rarely reported specific cutaneous disorder in DM patients is one of the prognostic indicators of malignancy. In the present study, only one of five cases presented with a malignancy. In this case, severe

and multiple skin lesions characteristic of a patient with cancer were observed. Although a high incidence of interstitial pneumonia (4/4, 100%) might be characteristic of DM patients with cutaneous necrosis without malignancy, the scale of the present study was small, and no statistically significant differences were observed compared with cases of patients who had neither cutaneous necrosis nor malignancy (19/31, 61%).

The present study offers additional clinical profiles of DM patients with cutaneous necrosis, with an evaluation of the therapeutic course. All five patients had mild or no evidence of myositis, with a high prevalence of interstitial pneumonia (4/5). In all patients, the early stage of cutaneous lesion formation prior to ulceration resembled phlegmon. However, a bacterial culture was taken only after an intractable ulcer had developed. Cutaneous lesion appeared to develop in response to mechanical stimulus, and covering the surface of the knee with a cloth in case 4 prevented ulceration from bullous lesion. Steroid therapy appeared to be ineffective in treating these lesions in cases 1–4, whereas the majority of lesions closed after several months of local treatment. On the other hand, the observed necrotic lesions might not have been caused by the high-dose steroid therapy, given that a similar lesion developed prior to steroid therapy in case 5, and reappeared in case 4 during a 5-mg daily dose of prednisolone. The present study confirmed the previously reported finding that topical bactericidal reagent gentiana violet reduces the risk of local infection with methicillin-resistant *Staphylococcus aureus*,[6] and further found that it accelerated the healing of the skin, as in case 4. Thus, strict sterilization is considered to be essential for treatment of this type of lesion.

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