Case Report

ANNALS OF MEDICINE & SURGERY

OPEN

A 30-year-old female with dermatomyositis without high elevation of muscle enzymes: a rare case report from Syria

Suaad Hamsho, MD[a], Bilal Sleiay, MD[b], Mouhammed Sleiay, MD[b,*], Obeda Batrash, MD[b], Hadi Alabdullah, MD[b], Majed Aboud, PhD[a]

Introduction and importance: Dermatomyositis (DM), sometimes referred to as inflammatory and degenerative changes in the skin and muscles, is a rare autoimmune disorder. DM is distinguished by myopathic disease, symmetrical proximal muscle weakness, and increased creatine kinase (CK).

Case presentation: A 30-year-old-female presented to the department of dermatology with a history of chronic right hand pain spreading to the shoulder, severe tachycardia, and dyspenia that increased during routine tasks like using the bathroom. What makes this case unique is that the CPK developed without doubling, and the final concentration was 207 ng/ml. Other common clinical symptoms include amyopathic/hypomyopathic muscle involvement and DM-specific rash (Gottron's papules, heliotrope rash), and these manifestations were in our patients. Sun protection, topical treatment with corticosteroids and/or calcineurin inhibitors, and systemic medication should be utilized for all individuals with nonvasculopathic disease. In our case, the patient stopped using azathioprine and began taking methotrexate.

Clinical discussion: Sun protection, topical therapy with corticosteroids and/or calcineurin inhibitors, and systemic medication should be utilized in layers for all individuals with nonvasculopathic illnesses. Mycophenolat Mofetil is beneficial in treating refractory illnesses as well as individuals with interstitial lung disease or substantial skin disease.

Conclusion: Even if test findings are not conclusive, dermatomyositis should always be considered when muscular weakness manifests. It's important to distinguish the disorder from connective tissue diseases like lupus erythematosus. In fact, to correctly diagnose DM, if there are any doubts, a muscle biopsy is required.

Keywords: dermatomyositis, ischaemia, mycophenolat mofetil, polymyositis

Introduction

Dermatomyositis (DM) is an uncommon autoimmune condition causes inflammatory and degenerative changes in the skin and muscles. Beyond this, DM can have serious systemic side effects and can be paraneoplastic in up to 30% of individuals. Women experience DM two to three times as frequently as men do. Particularly, individuals with newly diagnosed DM had considerably higher gene expression levels of the adipokine visfatin, which is linked to female sex as opposed to male sex[1]. The muscle enzyme used most often to diagnose polymyositis and evaluate treatment outcomes is creatine kinase. Despite the fact that most polymyositis patients appear with elevated creatine kinase levels, some individuals in documented series have normal creatine kinase levels upon presentation[2]. The relevance of having normal creatine kinase levels in polymyositis or DM is still unclear, despite claims that they seem to be associated with a bad prognosis due to either an accompanying malignancy or severe interstitial lung disease[3]. New criteria for IIM and its major subgroups were developed and published in 2017 by the European League Against Rheumatism (EULAR) and the American College of Rheumatology (ACR). These criteria took into account a number of factors, including age at disease onset, proximal muscle weakness, skin manifestations, and laboratory tests, with or without a muscle biopsy. When the patient's information is entered, these criteria can be used to determine the

HIGHLIGHTS

- Dermatomyositis (DM), sometimes referred to as inflammatory and degenerative changes in the skin and muscles are brought on by a rare autoimmune disorder.
- DM is distinguished by myopathic disease, symmetrical proximal muscle weakness, and increased creatine kinase.
- Even if test findings are not conclusive, DM should always be considered when muscular weakness manifests. It's important to distinguish the disorder from connective tissue diseases like lupus erythematosus.

[a]Department of Rheumatology, Faculty of Medicine, Damascus University, Damascus and [b]Faculty of Medicine, Hama University, Hama, Syria

Sponsorships or competing interests that may be relevant to content are disclosed at the end of this article

*Corresponding author. Address: Faculty of medicine, Hama University, Hama, Syrian Arab Republic. Tel.: +963 932 624 542. E-mail: abdmouh1234mouhmouh@gmail.com (M. Sleiay).

Annals of Medicine & Surgery (2024) 86:2167–2171

Received 6 October 2023; Accepted 24 December 2023

Published online 11 January 2024

http://dx.doi.org/10.1097/MS9.0000000000001682

likelihood that the patient has the condition. Additionally, they permitted subgrouping based on the age of onset and the existence of cutaneous symptoms[2]. Here we report a case of a female with severe DM without a high elevation of muscle enzymes. And it may be very rare in the medical literature.

Presentation of case

A 30-year-old woman presented to the rheumatology department with a history of progressive muscle weakness for about a year that increases during routine work, with chronic pain in the right hand that spreads to the shoulder, in addition to painful aphtus and arthralgia. The patient then experienced swelling in her middle finger, which led to dysfunction.These symptoms started when she was 28 years old. As a result, she was asked to go to the orthopaedic department, where she was given a cortisone injection into the joint, and the swelling subsided. One month later, the patient felt fatigued and exhausted, in addition to having a purplish maculopapular rash on the extensor surfaces of her fingers. In addition to oedema around the eyes, the patient had sensitivity to light and erythema around her eyes and nose (Fig. 1), which prompted her to go to the immunology department. On examination, all her vital observations were normal. There was no dyspnoea, cough, fever, or difficulty swallowing. There was no Raynaud's phenomenon. There was a history of miscarriage due to hydrops fetalis. There was no known statin use. She did not smoke or drink alcohol. There was proximal nail fold erythema but no sclerodactyly. There was diffuse tenderness over most of her joints, but no joint effusions or synovial thickening. Complete blood count tests were normal. The C-reactive protein level was normal, but the erythrocyte sedimentation rate (ESR) was 34 mm at 1 h and 85 mm at 2 h. Although aspartate transferase (AST) was normal and lactate dehydrogenase (LDH) was 402 U/l (within the normal range), alanine transaminase (ALT) was increased to 88 U/l. However, her hepatitis C profile ruled out liver disease because her Hepatitis B surface antigen and Hepatitis B core antibody results were normal. Creatinine and urinalysis results were normal. Thyroid stimulating hormone (TSH) and gamma-glutamyltransferase (GGT) were within the normal range. Rheumatoid factor (RF), anti-cyclic peptide antibody (anti-CCP), anti-nuclear HEP-2 antibody, HEP-2 cell immunofluorescence test (ANA-HEP-2 IF), adolase, and complement C3 and C4 levels were all within the normal range(Table 1).The results of the double antibody DNA (DsDNA) and anti-Sm tests were negative, ruling out the diagnosis of mixed connective tissue disease(Table 1). But what makes this case special is that the CPK developed without doubling, and the final result was 207 ng/mL (total CPK normal values: 10–120 micrograms per liter).Because DM may be associated with Cancers in females, especially in the ovaries and uterine , ovarian and uterine ultrasounds were performed, which showed normal findings. Because DM may be associated with ILD, we performed a chest X-ray, which also showed normal findings (Fig. 2). No computed tomography (CT) scan was requested. The electrocardiogram (ECG) was normal. X-rays of the hand also showed no abnormalities (Fig. 3). Electroneurography in the upper extremities was examined, and the results were normal with no signs of peripheral neuropathy (Fig. 4). Electromyography: Results from a muscle-focused needle electrode showed myositis. The biopsy revealed muscle fibre necrosis, degeneration, regeneration, and inflammatory cell infiltration, which confirmed the diagnosis of DM (Fig. 5). The patient was then treated with cortisone and azathioprine for 1 year following an electromyography. The muscle weakness improved on azathioprine, but the skin signs did not completely subside. Due to the lack of complete absence of skin signs on Azathioprine, we changed it to Methotrexate 2.5–4 pills per week, and we noticed that the skin signs disappeared after several months. The patient did not complain of any chest or heart symptoms, and no malignant tumours were discovered during this period.

Discussion

DM is an uncommonly acquired immune-mediated muscle illness. It is characterized by muscular weakening and a rash on the skin. It is categorized as an IIM (idiopathic inflammatory myopathy). DM often manifests in children between the ages of 5 and 15 years old. More females than males suffer from DM[4]. Our patient was 30 years old, which makes this case very rare.

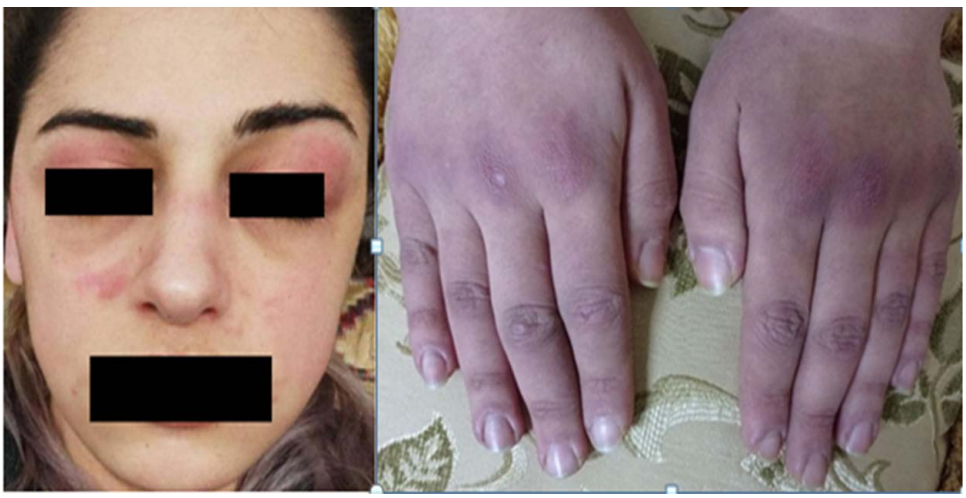

Figure 1. Showing dermatological changes like erythema around the nose and eyes and purple rash on hands (Gottron's papules).

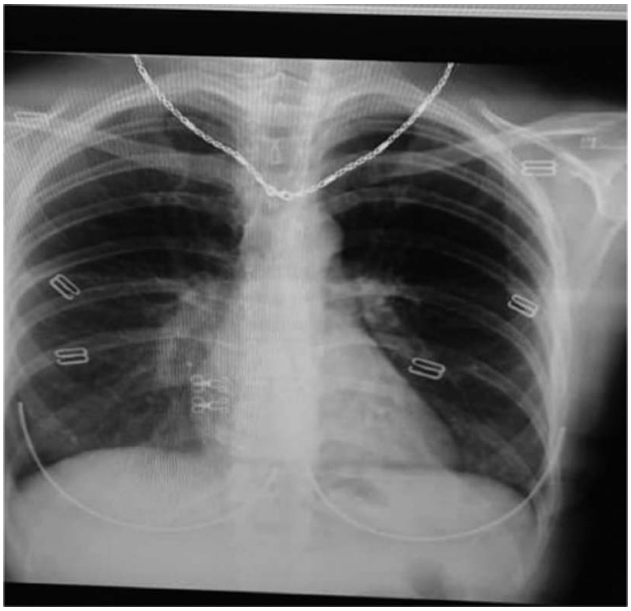

Figure 2. The chest X-ray were normal.

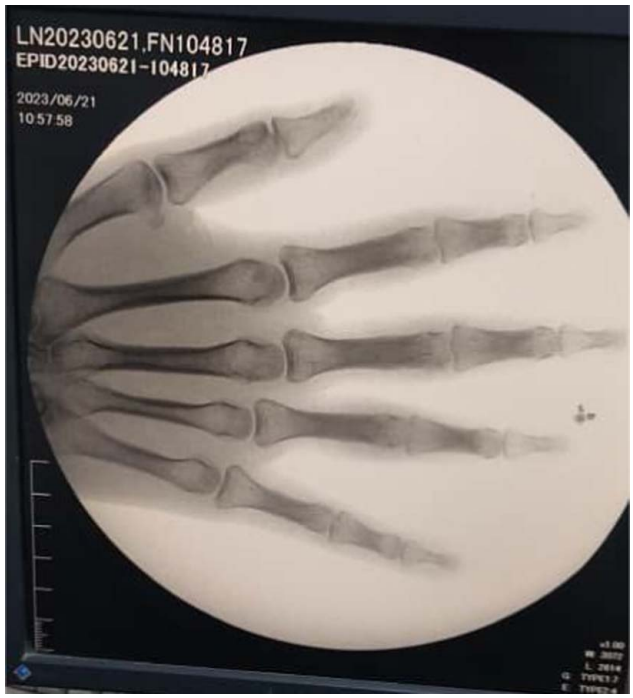

Figure 3. The hand X-ray wasn't showed abnormalities.

Despite the fact that all idiopathic inflammatory myopathies show similar muscular weakness, they vary clinically in terms of the muscle groups affected and histological findings[4]. Typical symptoms of DM include symmetrical proximal skeletal muscle weakening. Additionally, it may have an impact on several organ systems, including the gastrointestinal, cardiovascular, and respiratory systems[5]. Other common clinical symptoms include a heliotrope rash with periorbital swelling, in addition to an erythematous rash with violaceous patches with an overlying scale and hyperpigmentation in Metacarpo-phalangeal joint (MCP), Proximal Interphalangeal Joint (PIP) or Distal Interphalangeal Joint (DIP) (Known as Gottron's papules), and these manifestations were noticed in our patients. Women are more likely than men to have the disorder, with incidence rates of 3.98 and 4.68 per 1 000 000, respectively[6]. These studies suggest that environmental factors may act as triggers for the condition, as evidenced by the higher prevalence of DM and clusters of clinically amyopathic DM (CADM) in areas with high airborne pollution in a cohort study from Pennsylvania. Without appropriate DM-specific antibody screening, it is easy to overlook the correct diagnosis in cases that do not demonstrate elevated CK levels. Amyopathic DM is now frequently misdiagnosed as systemic lupus erythematosus or an undifferentiated connective tissue disorder[7]. More than 90% of cases of DM are characterized by myopathic disease, symmetrical proximal muscle weakness, and elevated CK[8]. But what makes this case special is that the CPK developed without doubling, and the end result was 207 ng/ml (total CPK normal values: 10–120 micrograms per liter). DM is frequently associated with cardiac issues such as myocarditis, ischaemia, arrhythmias, and cardiomyopathies[8]. Interstitial lung disease (ILD) develops in atypical DM patients more than typical DM patients. But in our case, no abnormalities were found on the chest X-ray or ECG. DM patients also seem to have an increased risk for internal malignancy; however, it is still unclear how this risk compares to that of classic DM patients[7].

In screening for occult malignancies, and since the chest images and chest examination were normal, a chest CT was not performed. The ultrasound of the ovaries and uterus was normal. A colonoscopy was not performed due to the fact that she was female and young, and there was no constipation, bloody stool, or anything suggestive of cancer. Three layers of therapy should be used for all patients with nonvasculopathic disease: sun protection, topical therapy with corticosteroids and/or calcineurin inhibitors, and systemic therapy[9]. With an 83% improvement in skin characteristics, a decline in CK levels (when raised), and better muscular strength after 22 months, Mycophenolat Mofetil (MMF) is effective in refractory illness as well as in patients with ILD or significant skin disease.[8] Intravenous immunoglobulin (IVIG) is an effective treatment for many autoimmune disorders, including DM. Patients who are resistant or intolerant to at least antimalarials and methotrexate (MTX), and often MMF as well, are given IVIG as a substitute. For muscle involvement in DM, corticosteroids are a staple of treatment and are regarded as first-line therapy. However, only between 60 and 75 percent of people benefit from this treatment, with 25–50% of individuals producing side effects such as avascular vertebral fractures and skeletal necrosis[10,11]. The differential diagnosis that we suggested was lupus erythematosus, rheumatoid arthritis or Mixed connective tissue disease (MCTD), but testing ruled these out. By using Electromyography and biopsy, she was confirmed to have DM and was treated for a year with cortisone and azathioprine (100 mg per day). After this, the patient started taking methotrexate) 10 mg per week)instead of azathioprine and kept taking cortisone (50 mg per day with gradual decreasing dose for 6 months). After the treatment there was a good improvement in the daily activities in our patient with less complaints from her skin lesions.

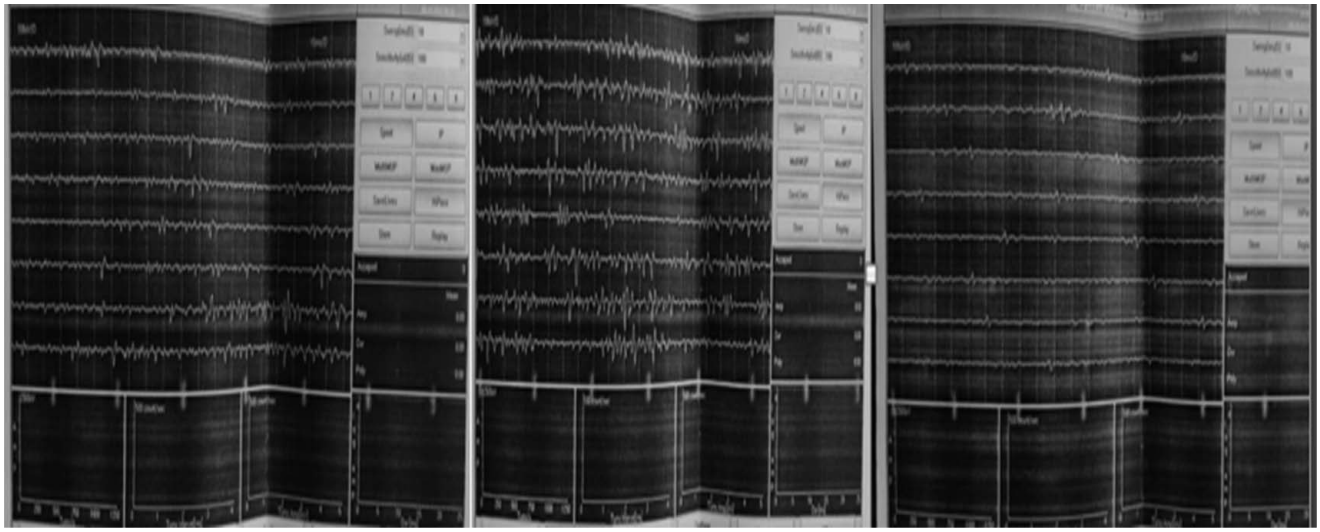

Figure 4. Findings from a concentrated needle electrode of the muscles showed myositis.

Conclusion

This case study highlights the unique diagnostic and therapeutic challenges posed by atypical presentations of DM. Over 90% of DM cases present with myopathic disease with symmetrical proximal muscle weakness and raised CK. When muscle weakness appears, DM should always be taken into consideration, even if tests of muscle enzymes such as CK, LDH, and aldolase are

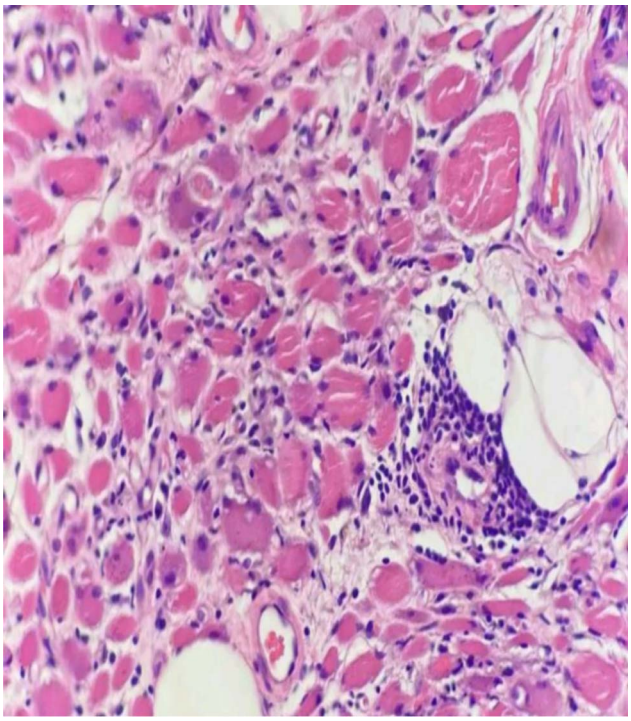

Figure 5. The biopsy revealed muscle fibre necrosis, degeneration, regeneration, and an inflammatory cell infiltrate.

Table 1

Laboratory tests and their values

test	Result	Normal range
WBCs	4200/mm^3	4500–10 500
Haemoglobin	13.10 g/dl	12–16
MCV	89.3 fl	78–96
Platelets	185 10^3 /mm^3	150–450
Lymphocytes/neutrophils	17/80%	
CRP	1.7 mg/l	0–6
ESR	34 mm/1h 85 mm/2h	0–20
AST	31 U/l	5–45
ALT	88 U/l	5–45
HBS Ag	0.521	Negative, less than: 1 Positive, more than: 1
HBC Ab	0.231	Negative, less than: 1.0 Positive, more than: 1.0
Creatinine	0.69 mg/dl	0.57–1.13
Urinalysis	Normal	
RF	5.6 IU/ml	0–30
Anti-CCP	7.1 U/ml	Up to 17
ANA-HEP-2 IF	Negative	Negative up to 1/80
Aldolase	1.12	1.0–7.5 U/l
Complement C3	133	C3: 80–178 mg/dl
Complement C4	36	C4: 12–42 mg/dl
DNA(DsDNA)	Negative	Less than 30.0: IU/ml More than 75.0: IU/ml
Anti-Sm	Negative	0–7 U/ml
CPK	207 ng/ml	Up to 140
GGT	29 U/l	0–39
LDH	402 U/l	210–480
T-bilirubin	0.61 mg/dl	0–1
D-bilirubin	0.14 mg/dl	0–0.25

ALT, alanine transaminase; AST, aspartate transferase; ANA-HEP-2 IF, anti-nuclear antibodies HEP-2 cells immunofluorescence test; Anti-CCP, cyclic citrullinated peptide antibodies; Anti-Sm, Anti-Smith antibody; ESR, erythrocyte sedimentation rate; CPK, creatinephosphokinase; CRP, C-reactive protein; GGT, gamma-glutamyltransferase; HBC Ab, Hepatitis B core antibody; HBS Ag, Hepatitis B surface antigen; LDH, lactate dehydrogenase; MCV, Mean corpuscular volume; RF, rheumatoid factor; WBC, white blood cell.

inconclusive or normal. The disease must be distinguished from connective tissue diseases such as lupus erythematosus. Clinical history and the characteristic rash are most important in the diagnosis.

Methods

The work has been reported in line with the SCARE criteria[12].

Ethical approval

It is not applicable because all data belong to the authors of this article.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Source of funding

None.

Author contribution

S.H. wrote most of the manuscript and performed data analysis or interpretation and designed the study. B.S. wrote a part of the manuscript and performed data analysis or interpretation and designed the study. M.S. wrote a part of the manuscript and designed the study. H.A. wrote a part of the manuscript. O.B. wrote a part of the manuscript. M.A. wrote a part of the manuscript. All authors reviewed the final manuscript.

Conflicts of interest disclosure

None.

Research registration unique identifying number (UIN)

None.

Guarantor

Not applicable . All data belong to the authors. The guarantor author is Mouhammed Sleiay.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Acknowledgements

The authors appreciate the efforts and the help of SMSR lab. and its role in binging our team together.

References

[1] Kassamali B, Mazori DR, LaChance AH, *et al*. Exploring dermatomyositis through an interdisciplinary lens: pearls from dermatology and rheumatology. Int J Womens Dermatol 2021;7(5Part A):576–82.
[2] Udman EJ, Schnitzer TJ. Dermatomyositis without creatine kinase elevation. A poor prognostic sign. Am J Med 1986;80:329–32.
[3] Lee YH, Choi SJ, Ji JD, *et al*. Dermatomyositis without elevation of creatine kinase presented as bronchiolitis obliterans organizing pneumonia. Korean J Intern Med 2000;15:85–8.
[4] Briani C, Doria A, Sarzi-Puttini P, *et al*. Update on idiopathic inflammatory myopathies. Autoimmunity 2006;39:161–70.
[5] Gerami P, Schope JM, McDonald L, *et al*. A systematic review of adult-onset clinically amyopathic dermatomyositis (dermatomyositis siné myositis): a missing link within the spectrum of the idiopathic inflammatory myopathies. J Am Acad Dermatol 2006;54:597–613.
[6] Kuo CF, See LC, Yu KH, *et al*. Incidence, cancer risk and mortality of dermatomyositis and polymyositis in Taiwan: a nationwide population study. Br J Dermatol 2011;165:1273–9.
[7] Bailey EE, Fiorentino DF. Amyopathic dermatomyositis: definitions, diagnosis, and management. Curr Rheumatol Rep 2014;16:465.
[8] Kwan C, Milosevic S, Benham H, *et al*. A rare form of dermatomyositis associated with muscle weakness and normal creatine kinase level. BMJ Case Rep 2020;13:e232260.
[9] Waldman R, DeWane ME, Lu J. Dermatomyositis: diagnosis and treatment. J Am Acad Dermatol 2020;82:283–96.
[10] Femia AN, Vleugels RA, Callen JP. Cutaneous dermatomyositis: an updated review of treatment options and internal associations. Am J Clin Dermatol 2013;14:291–313.
[11] Wu W, Guo L, Fu Y, *et al*. Interstitial lung disease in anti-MDA5 positive dermatomyositis. Clin Rev Allergy Immunol 2021;60:293–304.
[12] Sohrabi C, Mathew G, Maria N, *et al*. The SCARE 2023 guideline: updating consensus Surgical CAse REport (SCARE) guidelines. Int J Surg Lond Engl 2023;109:1136.