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A Case Report on the Differential Diagnosis of Progressive Pulmonary Lesions During Treatment for Anti-Mi-2α and Anti-NXP2 Double-Positive Dermatomyositis

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Case Report

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Abstract

Background:

Progressive pulmonary lesions in patients with dermatomyositis receiving glucocorticoids and immunosuppressive therapy require careful differential diagnosis. Possible causes include dermatomyositis-associated pulmonary involvement, opportunistic infection, bacterial or viral pneumonia, and drug-induced lung injury. Pneumocystis jirovecii pneumonia may manifest with fever, cough, dyspnea, hypoxemia, and ground-glass opacities on chest imaging, features that can closely resemble worsening dermatomyositis-associated interstitial lung disease or increased disease activity. We report a case of anti-Mi-2α and anti-NXP2 double-positive dermatomyositis complicated by progressive pulmonary lesions during treatment. After systematic evaluation, the pulmonary lesions were ultimately attributed to Pneumocystis jirovecii pneumonia. The lung abnormalities improved after antimicrobial therapy, whereas the underlying dermatomyositis subsequently relapsed.

Case Presentation:

A 46-year-old man presented with rash, proximal myalgia, and muscle weakness. Physical examination showed heliotrope rash, V-sign, periungual erythema, and Gottron's papules. Laboratory testing revealed a markedly elevated creatine kinase level, peaking at 10,082.90 U/L. A myositis-specific antibody panel was positive for anti-Mi-2α++ and anti-NXP2+, and electromyography indicated myopathic changes. Based on these findings, dermatomyositis was diagnosed. The patient received high-dose methylprednisolone pulse therapy, intravenous immunoglobulin, methotrexate, and tofacitinib, after which the rash, myalgia, and muscle weakness improved transiently. During treatment, he developed high fever, cough, dyspnea, and hypoxemia. Chest computed tomography demonstrated pulmonary inflammatory changes with ground-glass opacities. At that stage, dermatomyositis-related pulmonary involvement, increased disease activity, and uncomplicated infectious pneumonia were all considered. Further microbiological testing detected Pneumocystis jirovecii nucleic acid, along with positive anti-cytomegalovirus IgM. After partial withdrawal of immunosuppressive agents, the patient was treated with co-trimoxazole, ganciclovir, and supportive measures. His body temperature normalized, respiratory symptoms resolved, and follow-up chest computed tomography showed interval improvement. After the pulmonary infection was controlled, however, the patient again developed worsening rash over the face, neck, and anterior chest, accompanied by increased fatigue, proximal muscle weakness, and dysphagia. Muscle enzyme levels remained elevated, raising concern for reactivation of dermatomyositis. Methylprednisolone was continued, and intravenous immunoglobulin and methotrexate were reintroduced. Thereafter, the rash and fatigue improved compared with the previous disease state.

Conclusion:

This case highlights that progressive pulmonary lesions during treatment for dermatomyositis should not be automatically attributed to primary disease-related pulmonary involvement or increased disease activity. Opportunistic infections, particularly Pneumocystis jirovecii pneumonia, should be actively considered in the differential diagnosis. When rash, muscle weakness, dysphagia, or elevated muscle enzymes recur after improvement of pulmonary lesions, reactivation of dermatomyositis should be reassessed. In patients with active dermatomyositis complicated by opportunistic infection, treatment requires ongoing adjustment between infection control and the intensity of immunosuppression, guided by clinical manifestations, imaging findings, microbiological evidence, and markers of myositis activity.

Key Summary Points

1. Why report this case?

When pulmonary lesions progress in a patient with dermatomyositis during immunosuppressive therapy with corticosteroids, methotrexate, and Janus kinase inhibitors, several possibilities must be considered, including dermatomyositis-related pulmonary involvement, increased disease activity, common infectious pneumonia, and opportunistic infection.

The present patient had anti-Mi-2α and anti-NXP2 double-positive dermatomyositis. At disease onset, he presented with characteristic rash, proximal myalgia, muscle weakness, markedly elevated muscle enzyme levels, and electromyographic evidence of myopathic damage. During intensive immunosuppressive treatment, he developed high fever, cough, dyspnea, hypoxemia, and progressive abnormalities on chest imaging. Microbiological testing ultimately confirmed Pneumocystis jirovecii pneumonia.

A notable feature of this case was the occurrence of two clinically distinct episodes of “progression.” The first was characterized by worsening pulmonary lesions, which were mainly attributable to Pneumocystis jirovecii pneumonia. The second occurred after antimicrobial therapy and presented with aggravation of rash, muscle weakness, and dysphagia, findings that were more consistent with reactivation of the underlying dermatomyositis.

2. What insights does this case provide?

Progressive pulmonary lesions during the treatment of dermatomyositis should not be automatically ascribed to dermatomyositis-associated interstitial lung disease or relapse of the primary disease. A structured differential diagnosis is needed, with attention to the intensity of immunosuppressive therapy, clinical manifestations, hypoxemia, chest computed tomography findings, inflammatory markers, and microbiological evidence.

In patients with dermatomyositis who are receiving moderate- to high-dose glucocorticoids combined with immunosuppressants or Janus kinase inhibitors, opportunistic infections such as Pneumocystis

jirovecii pneumonia should be considered early when unexplained fever, dry cough, dyspnea, hypoxemia, or ground-glass opacities appear on chest imaging.

Resolution of pulmonary abnormalities after antimicrobial therapy does not necessarily indicate remission of dermatomyositis. If rash worsens, proximal muscle weakness progresses, dysphagia develops or deteriorates, or muscle enzyme levels remain elevated, disease activity should be reassessed.

For patients with active dermatomyositis complicated by opportunistic infection, treatment requires continual adjustment between infection control and suppression of autoimmune activity. Clinical decisions should be guided by serial evaluation of symptoms, muscle strength, swallowing function, muscle enzyme levels, pulmonary imaging, and microbiological test results.

Introduction

Dermatomyositis is a highly heterogeneous subgroup of idiopathic inflammatory myopathies. Its typical manifestations include characteristic cutaneous eruptions, proximal muscle weakness, elevated muscle enzyme levels, and myopathic changes, and the disease may affect multiple organ systems, including the lungs, heart, and gastrointestinal tract. In recent years, increasing attention has been paid to the role of myositis-specific antibodies in the diagnosis, classification, and prognostic assessment of dermatomyositis. Different antibody subtypes are often associated with distinct clinical phenotypes and patterns of organ involvement(1). Anti-Mi-2 antibodies are commonly linked to classic dermatomyositis skin lesions and prominent myositis, whereas anti-NXP2 positivity may be associated with more severe muscle involvement, variable cutaneous manifestations, and a potential risk of malignancy(2). Even in patients with relatively well-characterized antibody profiles, newly developed pulmonary lesions during treatment may arise from different mechanisms, and identifying their cause is often critical for subsequent therapeutic decisions.

Pulmonary involvement is one of the major extramuscular manifestations of dermatomyositis, and interstitial lung disease is closely related to poor prognosis(3). When patients with dermatomyositis develop fever, cough, dyspnea, hypoxemia, or ground-glass opacities on chest computed tomography during treatment, dermatomyositis-related pulmonary involvement or progression of the underlying disease is often considered first. Recent studies have shown that interstitial lung disease occurs more frequently in dermatomyositis than in polymyositis and may appear early in the disease course(4). Patients with active dermatomyositis often require moderate- to high-dose glucocorticoids, methotrexate, calcineurin inhibitors, Janus kinase inhibitors, or other immunosuppressive therapies. At present, there is no universally accepted international standard treatment regimen for myositis, and clinical management is usually individualized according to disease activity and organ involvement(5). In this clinical context, infectious pneumonia, especially opportunistic infection, may closely mimic primary pulmonary involvement in both symptoms and imaging findings, making differential diagnosis challenging.

Pneumocystis jirovecii pneumonia is a potentially fatal opportunistic infection in immunocompromised patients. It typically presents with fever, dry cough, progressive dyspnea, hypoxemia, and bilateral ground-glass opacities. Patients with idiopathic inflammatory myopathies have been reported to be at increased risk of Pneumocystis jirovecii pneumonia, particularly during the early phase of intensive immunosuppressive therapy(6). In patients with anti-MDA5-positive dermatomyositis, Pneumocystis jirovecii pneumonia has been associated with higher short-term mortality, while prophylactic co-trimoxazole may confer protection(7). Reports focusing on the differential diagnosis of progressive pulmonary lesions during treatment and subsequent reactivation of the underlying disease in non-anti-MDA5 subtypes remain limited, particularly in patients with anti-Mi-2α and anti-NXP2 double-positive dermatomyositis. Here, we describe a patient with anti-Mi-2α and anti-NXP2 double-positive dermatomyositis who developed progressive pulmonary lesions during intensive immunosuppressive therapy. The lesions were ultimately confirmed to be Pneumocystis jirovecii pneumonia. After anti-infective treatment, the pulmonary abnormalities improved; however, worsening rash, muscle weakness, and dysphagia subsequently developed, suggesting reactivation of the underlying dermatomyositis. This case underscores the need for continuous differential diagnosis when pulmonary lesions progress in patients with dermatomyositis and highlights the importance of balancing infection control with appropriate immunosuppressive treatment.

Case Presentation

A 46-year-old man was admitted to the hospital with a rash persisting for more than 6 months, myalgia for more than 3 months, and fever for 24 days.

Initial Clinical Presentation and Diagnosis of Dermatomyositis

In October 2025, the patient developed multifocal erythematous eruptions without an obvious trigger. The rash mainly involved the nasal bridge, periorbital region, forehead, chin, anterior neck, proximal upper limbs, and both thighs. It was accompanied by pruritus, desquamation, and localized warmth of the skin. At disease onset, he had no fever, arthralgia, dry mouth, dry eyes, alopecia, recurrent oral or genital ulcers, Raynaud’s phenomenon, chest tightness, dyspnea, abdominal pain, or diarrhea. He initially sought care at a local hospital, where “dermatitis” was considered. Symptomatic and topical treatment provided little benefit, and the rash persisted.

In December 2025, the patient gradually developed proximal myalgia and weakness of the limbs. These symptoms manifested as difficulty raising both upper limbs, squatting and standing up, and lifting the head while lying supine. Mild limitation of mouth opening and localized swelling of the upper arms were also noted. There was no obvious coughing when drinking, dysphagia, or dyspnea. Laboratory testing showed a creatine kinase (CK) level of 10,082.90 U/L, creatine kinase-MB (CK-MB) of 1,071.08 U/L, and an erythrocyte sedimentation rate (ESR) of 48 mm/h. Myositis antibody profiling revealed anti-Mi-2α antibodies at + + and positivity for anti-NXP2 antibodies. Electromyography demonstrated myopathic

changes. On the basis of the characteristic rash, proximal myalgia and muscle weakness, markedly elevated muscle enzyme levels, myopathic changes on electromyography, and positive myositis-specific antibodies, the patient was diagnosed with anti-Mi-2α and anti-NXP2 double-positive dermatomyositis.

From January 7 to January 9, 2026, he received induction therapy with intravenous methylprednisolone sodium succinate 1.0 g once daily for 3 consecutive days, together with intravenous immunoglobulin (IVIG) 20 g once daily for 3 days. Beginning on January 10, 2026, treatment was adjusted to oral methylprednisolone 48 mg once daily. At follow-up on January 21, 2026, CK had decreased to 7,081 U/L, and serum ferritin was 630 ng/mL. Methotrexate was therefore added at a dose of 10 mg orally once weekly. At the follow-up visit on February 11, 2026, the methotrexate dose was increased to 15 mg orally once weekly, and oral tofacitinib citrate 5 mg twice daily was introduced. Approximately 2 months after initiation of this regimen, the rash, myalgia, and muscle weakness had improved compared with baseline, and the methylprednisolone dose was gradually tapered to 24 mg orally once daily.

Fever and Progression of Pulmonary Lesions Following Immunosuppressive Therapy

On March 16, 2026, the patient developed recurrent fever without an apparent cause, with a peak temperature of 40.0°C. The fever was sometimes preceded by chills and was accompanied by cough with white sputum, followed by gradually worsening dyspnea. He self-administered oral cefixime and later received intravenous ceftriaxone at a local hospital, but his symptoms did not improve significantly.

On March 23, 2026, the patient was readmitted. On admission, his temperature was 38.1°C, pulse rate 122 beats/min, respiratory rate 20 breaths/min, and blood pressure 133/81 mmHg. He was alert and oriented, with fair general condition. Physical examination showed pharyngeal congestion and patchy, dark-red, chronic-appearing rashes on the back and both thighs. Breath sounds were clear bilaterally, without obvious dry or moist rales. Cardiac rhythm was regular, and no definite pathological murmurs were heard over the valvular areas. The abdomen was soft, without tenderness or rebound tenderness. No significant lower-limb edema was observed.

Laboratory tests on admission showed a white blood cell count of 11.67 × 10^9/L, neutrophils 87.30%, lymphocytes 6.00%, C-reactive protein (CRP) 51.28 mg/L, ESR 70 mm/h, and procalcitonin (PCT) 0.080 ng/mL. Muscle enzyme levels had decreased compared with the initial presentation, with CK 2,183.50 U/L and lactate dehydrogenase (LDH) 786.00 U/L. Cytomegalovirus (CMV) IgM was 49.545 AU/mL. Chest computed tomography showed pulmonary inflammatory changes, mild ventilation-perfusion abnormality, multiple small bilateral pulmonary nodules, and intrapulmonary inflammatory streaks (Fig. 1A). Given his recent exposure to glucocorticoids, methotrexate, and tofacitinib, as well as his immunosuppressed status, opportunistic infection was considered. Bacterial pneumonia, viral pneumonia, drug-induced lung injury, and progression of dermatomyositis-associated pulmonary involvement also required differentiation.

After admission, methotrexate and tofacitinib were discontinued. Treatment for dermatomyositis was maintained with oral methylprednisolone 24 mg once daily, and empirical antimicrobial therapy was initiated with intravenous cefoperazone-sulbactam 3 g every 12 hours. Because anti-CMV IgM was positive, CMV infection was suspected, and intravenous ganciclovir 0.25 g every 12 hours was added. Despite treatment, the patient continued to have recurrent high fever, cough, dyspnea, and signs of hypoxia.

On March 26, 2026, arterial blood gas analysis showed pH 7.52, $PaCO_2$ 32 mmHg, PaO_2 65 mmHg, and oxygen saturation 94.8%, indicating hypoxemia. Oxygen therapy was administered, and intravenous moxifloxacin 0.4 g once daily was added for antimicrobial coverage.

Against the background of intensive immunosuppressive therapy, the combination of persistent high fever, progressive dyspnea, hypoxemia, and abnormal chest computed tomography findings raised concern for an opportunistic pulmonary infection. Polymerase chain reaction testing of an oral swab was positive for Pneumocystis jirovecii nucleic acid. Based on the clinical presentation and microbiological findings, Pneumocystis jirovecii pneumonia (PJP) was considered. The regimen was adjusted to include oral co-trimoxazole, 3 tablets four times daily, for anti-PJP treatment. Ganciclovir was increased to 0.35 g intravenously twice daily, and methylprednisolone sodium succinate 40 mg was administered intravenously twice daily.

Repeat arterial blood gas analysis on March 31, 2026, still showed a PaO_2 of 65 mmHg. Concurrent laboratory tests revealed CK 1,380.25 U/L and LDH 881.18 U/L. Anti-PJP and supportive therapy were continued. Follow-up chest computed tomography on April 1, 2026, showed partial resolution of the pulmonary inflammation compared with the scan from March 23, 2026, although mild ventilation-perfusion abnormality persisted (Fig. 1B). After treatment, the fever resolved, and cough, chest tightness, and dyspnea improved. As muscle enzyme levels had decreased, the methylprednisolone dose was gradually tapered to 40 mg once daily. On April 7, 2026, the patient was discharged in stable condition. After discharge, he continued oral co-trimoxazole for PJP and methylprednisolone for dermatomyositis; methotrexate and tofacitinib had not yet been resumed.

Dermatomyositis Relapse Following Resolution of Pulmonary Infection

After improvement of the pulmonary infection, the patient gradually developed increased erythematous rashes on the face, neck, and anterior chest beginning on April 8, 2026, accompanied by pruritus. Proximal muscle weakness persisted, and dysphagia developed, although there was no obvious coughing while drinking. He had no fever, cough, sputum production, or dyspnea. Considering the improvement in pulmonary symptoms and imaging findings, together with worsening rash, fatigue, proximal muscle weakness, and dysphagia, this clinical change was considered more consistent with relapse of the underlying dermatomyositis than with ongoing progression of pulmonary infection.

On April 9, 2026, the patient was readmitted for further treatment. At admission, his temperature was stable, and he had no fever, cough, sputum production, or dyspnea. Physical examination revealed a Cushingoid appearance, patchy erythematous rashes on the face, neck, both ears, and anterior chest, and patchy dark-red chronic rashes on the back (Fig. 2). Breath sounds were clear bilaterally, without dry or moist rales. Cardiac rhythm was regular, with no obvious pathological murmurs over the valvular areas. The abdomen was soft, without tenderness or rebound tenderness. Proximal muscle strength of the limbs was approximately grade 4^-, with normal muscle tone. No significant lower-limb edema was noted. Laboratory tests showed CRP < 0.50 mg/L, PCT 0.106 ng/mL, and ESR 70 mm/h. Infection-related markers had improved compared with previous results, whereas rash, proximal muscle weakness, and dysphagia had worsened, further supporting increased dermatomyositis activity.

After readmission, the patient received intravenous methylprednisolone sodium succinate 40 mg once daily and IVIG 20 g once daily for 5 consecutive days. Anti-PJP therapy was continued with oral trimethoprim-sulfamethoxazole tablets, 3 tablets every 6 hours. On April 12, 2026, while anti-infective treatment was maintained and the risk of recurrent infection was closely monitored, methotrexate 15 mg orally once weekly was reintroduced. Tofacitinib was not resumed.

Following treatment, the rash gradually faded, and fatigue and proximal muscle weakness improved. Dysphagia did not progress further, and no fever, cough, sputum production, chest tightness, or dyspnea occurred during hospitalization. Follow-up testing on April 13, 2026, showed CK 1,520.80 U/L, CK-MB 240.02 U/L, LDH 532.00 U/L, alanine aminotransferase 94.50 U/L, aspartate aminotransferase 93.10 U/L, albumin 27.00 g/L, and ESR 80 mm/h. Although muscle enzyme levels remained above the normal range, they had decreased substantially compared with the initial presentation. Continued management of dermatomyositis activity was required, together with ongoing monitoring of liver function, nutritional status, muscle enzyme trends, and infection risk.

On April 14, 2026, the patient was discharged after stabilization. The discharge medications were as follows: methylprednisolone tablets 40 mg orally once daily; methotrexate tablets 15 mg orally once weekly; folic acid tablets 10 mg orally once weekly, taken on the second day after methotrexate administration; and compound sulfamethoxazole tablets, 3 tablets orally every 6 hours, for continued anti-PJP treatment. The patient was instructed to return to the Rheumatology and Immunology outpatient clinic after 2 weeks for follow-up, including repeat complete blood count, CRP, ESR, liver and kidney function tests, electrolytes, cardiac enzymes, and ferritin. Regular reassessment with immunoglobulin testing, chest computed tomography, and echocardiography was also recommended to evaluate dermatomyositis activity, the outcome of pulmonary infection, and the safety of immunosuppressive therapy.

The patient's overall clinical course, diagnostic evaluation, treatment adjustments, and outcomes are summarized in Table 1.

Table 1. Timeline of the patient's clinical course, diagnostic assessment, treatment, and outcomes

Date	Clinical event	Diagnostic assessment	Treatment/intervention	Outcome
October 2025	Initial skin manifestations	Multiple erythematous rashes developed on the nasal bridge, periorbital area, forehead, jaw, anterior neck, proximal upper limbs, and thighs, with pruritus, desquamation, and local warmth. The patient initially had no fever, arthralgia, sicca symptoms, alopecia, oral/genital ulcers, Raynaud's phenomenon, dyspnea, abdominal pain, or diarrhea.	Diagnosed as dermatitis at a local hospital; symptomatic and topical treatment was given.	Skin lesions persisted and recurred.
December 2025	Development of myositis symptoms and diagnosis of dermatomyositis	The patient developed proximal myalgia and weakness, including difficulty raising the upper limbs, squatting, and lifting the head in the supine position. CK 10,082.90 U/L, CK-MB 1071.08 U/L, ESR 48 mm/h. Myositis antibody panel showed anti-Mi-2α +++ and anti-NXP2 positivity. Electromyography showed myogenic damage.	Anti-Mi-2α and anti-NXP2 double-positive dermatomyositis was diagnosed based on typical rash, proximal muscle symptoms, markedly elevated muscle enzymes, myogenic electromyographic changes, and myositis-specific antibodies.	Diagnosis established.
January 7–9, 2026	Initial intensive treatment	Active dermatomyositis with severe muscle enzyme elevation.	Intravenous methylprednisolone sodium succinate 1.0 g once daily for 3 days and intravenous immunoglobulin 20 g once daily for 3 days.	Initial intensive immunotherapy completed.
January 10, 2026	Transition to oral glucocorticoid therapy	—	Oral methylprednisolone 48 mg once daily.	Continued treatment for dermatomyositis.
January 21, 2026	Follow-up after initial treatment	CK decreased to 7081 U/L; serum ferritin 630 ng/mL.	Methotrexate 10 mg once weekly was added.	Muscle enzymes decreased but remained elevated.
February 11, 2026	Further immunosuppressive treatment adjustment	Persistent disease activity was considered.	Methotrexate was increased to 15 mg once weekly, and tofacitinib citrate 5 mg twice daily was added. Methylprednisolone was later tapered to 24 mg once daily.	After approximately 2 months of treatment, rash, myalgia, and muscle weakness improved.
March 16, 2026	Onset of fever and respiratory symptoms	Recurrent fever developed, with a maximum temperature of 40.0 °C, sometimes preceded by chills, followed by cough with white sputum and progressive dyspnea.	The patient self-administered cefixime and later received ceftriaxone at a local hospital.	Symptoms did not improve significantly.
March 23, 2026	Readmission for pulmonary lesion progression during immunosuppressive therapy	WBC 11.67 × 10⁹/L, neutrophils 87.30%, lymphocytes 6.00%, CRP 51.28 mg/L, ESR 70 mm/h, PCT 0.080 ng/mL, CK 2183.50 U/L, LDH 786.00 U/L, anti-CMV IgM 49.545 AU/mL. Chest CT showed pulmonary inflammation, mild ventilation-perfusion abnormality, multiple small nodules, and inflammatory fibrotic streaks.	Methotrexate and tofacitinib were discontinued. Oral methylprednisolone 24 mg once daily was continued. Empirical cefoperazone/sulbactam 3 g every 12 h was started. Ganciclovir 0.25 g every 12 h was added because of positive anti-CMV IgM.	Persistent high fever continued. Differential diagnoses included opportunistic infection, bacterial or viral pneumonia, dermatomyositis-associated pulmonary involvement, and drug-related lung injury.
March 26, 2026	Development of dyspnea and hypoxemia	Arterial blood gas analysis showed pH 7.52, $PaCO_2$ 32 mmHg, PaO_2 65 mmHg, and oxygen saturation 94.8%.	Oxygen therapy was given, and moxifloxacin 0.4 g once daily was added.	Hypoxemia supported clinically significant pulmonary involvement.
Late March 2026	Diagnosis of Pneumocystis jirovecii pneumonia	Pneumocystis jirovecii nucleic acid test was positive in the setting of immunosuppression, persistent fever, dyspnea, hypoxemia, and abnormal chest imaging.	Treatment was adjusted to trimethoprim-sulfamethoxazole, 3 tablets four times daily; ganciclovir was adjusted to 0.35 g twice daily; intravenous methylprednisolone sodium succinate 40 mg twice daily was administered.	PJP was considered the main cause of pulmonary lesion progression.
March 31, 2026	Reassessment during anti-infective therapy	Arterial blood gas still showed PaO_2 65 mmHg. CK was 1380.25 U/L; LDH was 881.18 U/L.	Anti-PJP and supportive treatment were continued.	Muscle enzymes were lower than at initial dermatomyositis diagnosis, while pulmonary manifestations remained the dominant clinical issue.
April 1, 2026	Radiological and clinical response after anti-PJP treatment	Chest CT showed absorption of pulmonary inflammation compared with March 23, 2026, while mild ventilation-perfusion abnormality remained.	Anti-infective therapy was continued. Methylprednisolone was gradually reduced to 40 mg once daily because of decreasing muscle enzymes.	Fever resolved, and cough, chest tightness, and dyspnea improved, supporting infection-driven pulmonary progression.
April 7, 2026	Discharge after stabilization of pulmonary infection	—	Oral trimethoprim-sulfamethoxazole was continued for PJP. Methylprednisolone was continued for dermatomyositis. Methotrexate and tofacitinib were not resumed at discharge.	Pulmonary symptoms improved, and the patient was discharged in a relatively stable condition.
April 8, 2026	Dermatomyositis flare after infection control	Increased erythematous rash on the face, neck, and anterior chest, with pruritus; persistent proximal muscle weakness; and new dysphagia without obvious choking. No fever, cough, sputum production, or dyspnea was present.	Rheumatology evaluation recommended continued methylprednisolone 40 mg daily, methotrexate 15 mg weekly, and IVIG 20 g daily for 3–5 days, with continued anti-infective management.	The clinical pattern suggested dermatomyositis flare after infection control rather than ongoing pulmonary infection.

Abbreviations: CK, creatine kinase; CK-MB, creatine kinase-MB; CMV, cytomegalovirus; CRP, C-reactive protein; ESR, erythrocyte sedimentation rate; IVIG, intravenous immunoglobulin; LDH, lactate dehydrogenase; PCT, procalcitonin; PJP, Pneumocystis jirovecii pneumonia; WBC, white blood cell count.

Discussion

This case illustrates the considerable complexity of diagnosing and managing progressive pulmonary disease in patients with dermatomyositis receiving multiple immunosuppressive therapies. The patient experienced two clinically distinct episodes of deterioration within a relatively short disease course. The first episode was characterized mainly by fever, cough, dyspnea, hypoxemia, and worsening chest imaging findings. Microbiological testing detected Pneumocystis jirovecii, and both respiratory symptoms and radiological abnormalities improved after antimicrobial therapy, primarily with co-trimoxazole. These findings supported Pneumocystis jirovecii pneumonia (PJP) as the main cause of the initial pulmonary deterioration. The second episode occurred after the pulmonary infection had been controlled. At that time, the patient had no fever, cough, sputum production, or dyspnea, and infection-related indicators had improved compared with the previous episode; however, rash, proximal muscle weakness, and dysphagia worsened, suggesting reactivation of the underlying dermatomyositis. This

case therefore emphasizes the need for dynamic reassessment of several competing or overlapping possibilities, including infection, disease activity, drug-related lung injury, and dermatomyositis-associated pulmonary involvement, particularly in patients receiving combined treatment with glucocorticoids, conventional immunosuppressants, and targeted agents.

The first lesson from this case is that progressive pulmonary lesions in dermatomyositis should not be automatically attributed to dermatomyositis-associated interstitial lung disease. In clinical practice, cough, dyspnea, hypoxemia, and new or progressive pulmonary infiltrates on chest computed tomography may suggest inflammatory myopathy-related pulmonary involvement. These manifestations, however, are nonspecific. During the first episode of deterioration in this patient, several findings favored opportunistic infection: recent exposure to glucocorticoids, methotrexate, and tofacitinib; fever and hypoxemia; a reduced lymphocyte percentage; a positive test for Pneumocystis jirovecii; and clinical and radiological improvement after anti-PJP therapy. Previous studies have reported that PJP in non-HIV immunocompromised patients often has a more acute onset, progresses more rapidly, is harder to diagnose, and carries a higher mortality risk(8, 9). For patients with dermatomyositis receiving potent immunosuppressive therapy, progressive respiratory symptoms should therefore prompt early and comprehensive microbiological evaluation rather than being attributed solely to autoimmune pulmonary involvement.

A second notable feature of this case was the relatively clear temporal distinction between infection-related deterioration and subsequent flare of dermatomyositis. During the first episode, muscle enzyme levels had already decreased compared with the initial diagnosis, and the rash and muscle weakness had temporarily improved, whereas fever, hypoxemia, and pulmonary infection-related findings predominated. After anti-PJP therapy, the fever resolved, cough, sputum production, chest tightness, and dyspnea improved, and chest computed tomography showed partial resolution of pulmonary inflammation. By contrast, rash, proximal muscle weakness, and dysphagia worsened. This shift suggests that the dominant pathological process changed from opportunistic infection to active dermatomyositis during the disease course. Such dynamic assessment is clinically important because the treatment strategies differ substantially. Premature intensification of immunosuppression before infection control may aggravate the infection, whereas excessive delay in resuming or strengthening immunosuppressive therapy after infection control may allow further progression of myositis, muscle weakness, and dysphagia.

The development of PJP in this patient should be interpreted in the context of combined immunosuppressive therapy rather than attributed to a single agent. Glucocorticoids are a major risk factor for opportunistic infection, and this risk increases when they are used together with other immunosuppressants. Studies have shown that, in autoimmune inflammatory diseases, glucocorticoids combined with additional immunosuppressive agents are more strongly associated with PJP than glucocorticoids alone(10). In the present case, the patient received methotrexate and tofacitinib in addition to glucocorticoids. Janus kinase inhibitors exert anti-inflammatory effects by modulating the JAK-STAT signaling pathway, which also participates in host defense against infection. Recent studies

have indicated that oral Janus kinase inhibitor therapy is associated with an increased overall risk of infection(11). Although this case cannot establish a direct causal relationship between tofacitinib and PJP, it supports heightened vigilance for opportunistic infections when glucocorticoids, conventional immunosuppressants, and Janus kinase inhibitors are used in combination.

This case also draws attention to the prevention of PJP in patients with rheumatic and autoimmune diseases. At present, fully standardized guidelines for PJP prophylaxis in non-HIV populations are lacking, and clinical practice varies across diseases, immunosuppressive regimens, and individual risk profiles. Glucocorticoid dose and duration, combination immunosuppressive therapy, lymphocytopenia, and pre-existing pulmonary disease may all increase the risk of PJP. Recent studies suggest that risk assessment and prophylactic treatment for PJP remain insufficient in adults receiving systemic glucocorticoid therapy(12). In this case, PJP developed while the patient was receiving methotrexate and tofacitinib in addition to moderate- to high-dose glucocorticoids. For similar high-risk patients with dermatomyositis, the need for prophylactic trimethoprim-sulfamethoxazole should be evaluated early in the treatment course, with individualized decisions based on complete blood count, liver and renal function, drug allergy history, and potential drug interactions.

From a therapeutic perspective, this case demonstrates the need to balance infection control with treatment of the underlying autoimmune disease. During the active phase of PJP, management focused on discontinuing methotrexate and tofacitinib, maintaining a necessary dose of glucocorticoids, and intensifying antimicrobial therapy, including co-trimoxazole for PJP. After the infection improved, the patient developed worsening rash, proximal muscle weakness, and dysphagia, indicating the need to strengthen treatment for dermatomyositis. At this stage, methylprednisolone and intravenous immunoglobulin were administered, methotrexate was resumed, anti-PJP therapy was continued, and the risk of recurrent infection was closely monitored. Intravenous immunoglobulin may be particularly useful in this setting. The ProDERM study showed that intravenous immunoglobulin improved the overall clinical condition of adults with active dermatomyositis(13), and subsequent safety analyses further supported its clinical value, although adverse events such as thromboembolism still require attention(14). Tofacitinib was not resumed in this patient, reflecting a cautious approach to the risk of recurrent opportunistic infection during recovery.

The central clinical message of this case is that dermatomyositis activity, opportunistic infection, and drug-induced pulmonary injury should not be regarded as mutually exclusive diagnoses. These processes may overlap or predominate at different stages of the disease. A single cross-sectional assessment may therefore be misleading. Clinical interpretation should integrate changes in symptom patterns, muscle enzyme trends, infection markers, oxygenation status, microbiological results, serial chest computed tomography findings, and treatment response. In this patient, the first episode of pulmonary deterioration was primarily attributed to PJP, whereas the second episode after infection control was more consistent with active dermatomyositis. Distinguishing infection-driven from disease activity-driven deterioration is essential for determining the timing of antimicrobial therapy, PJP prophylaxis, and the resumption or intensification of immunosuppressive treatment.

This case report has several limitations. First, as a single case, it cannot establish a definitive causal relationship among anti-Mi-2α and anti-NXP2 double positivity, tofacitinib exposure, and the occurrence of PJP. Second, because continuous CD4 + T-cell counts, serum β-D-glucan levels, quantitative pathogen-load testing, and standardized bronchoalveolar lavage analysis were not available in the case records, evaluation of mixed infection, PJP burden, and the activity of interstitial lung disease was limited. Third, long-term follow-up is needed to monitor dermatomyositis activity, the risk of PJP recurrence, pulmonary imaging outcomes, the safety of resumed immunosuppressive therapy, and potential malignancy risk.

In summary, this patient with anti-Mi-2α and anti-NXP2 double-positive dermatomyositis developed PJP-related progressive pulmonary lesions during multiple immunosuppressive therapies, followed by exacerbation of dermatomyositis activity after infection control. The case suggests that opportunistic infections such as PJP should be considered early when pulmonary lesions progress in patients with dermatomyositis receiving immunosuppressive therapy. A dynamic differential diagnosis among infection, disease activity, and drug-related toxicity is required. After infection control, recurrence or worsening of rash, muscle weakness, dysphagia, or elevated muscle enzyme levels should prompt reassessment for dermatomyositis reactivation, and immunosuppressive therapy should be resumed or intensified cautiously while anti-infective treatment and close monitoring are continued.

Declarations

Conflict of interest

The authors declare that they have no competing interests.

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Author Contribution

Minghao Wang: Conceptualization, data curation, literature retrieval, case data collection and sorting, manuscript drafting, preliminary revision of the manuscript.Zhiqiang Liang: Project supervision, clinical diagnosis and treatment guidance, manuscript design and critical revision, final approval of the manuscript, correspondence work.

Ethics approval

This case report was conducted in accordance with the Declaration of Helsinki. Institutional ethics approval was waived because this article reports a single case and does not involve experimental

intervention.Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Data availability

The data supporting the findings of this case report are available from the corresponding author upon reasonable request.

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Figures

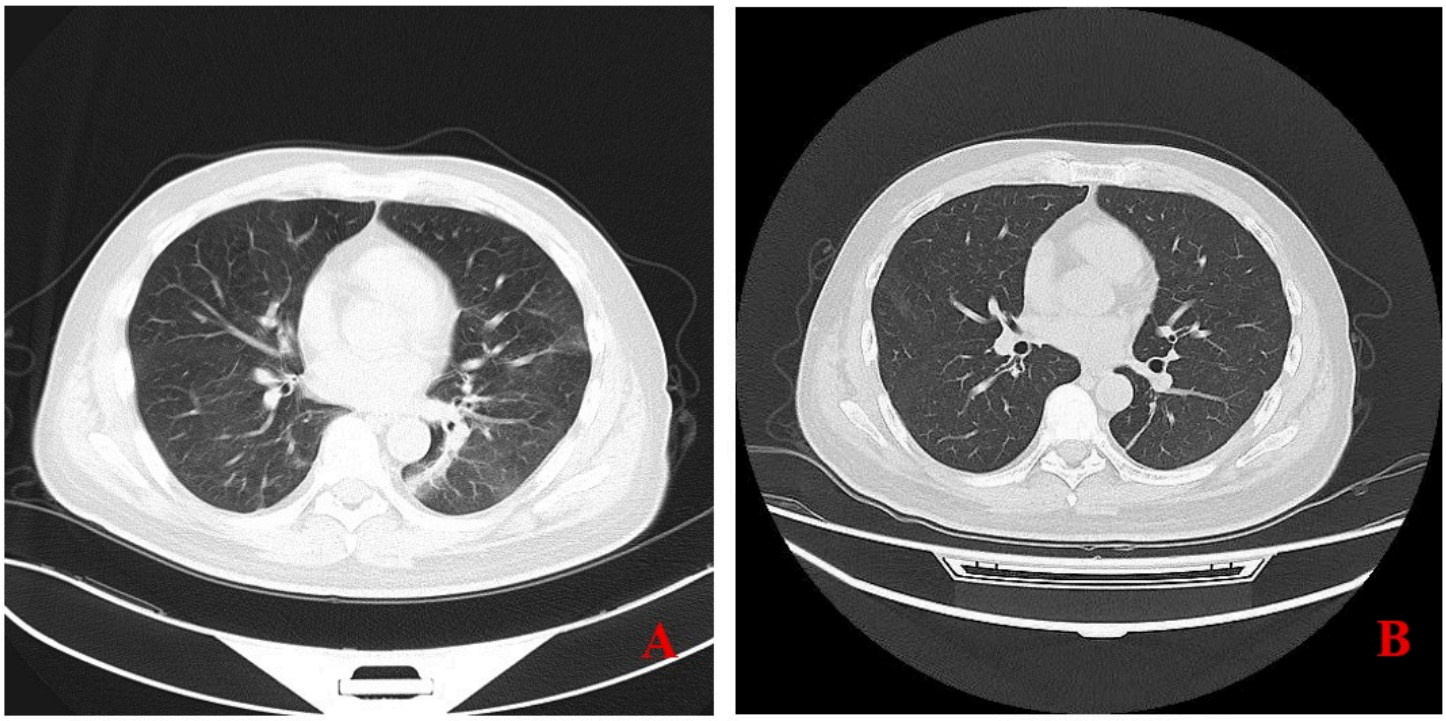


Figure 1

Comparison of post-treatment chest CT scans showing lung progression following immunosuppressive therapy

1A: Chest CT scan of the patient on March 23, 2026: multiple patchy, ground-glass opacities; pulmonary inflammation; and poor perfusion-ventilation. Multiple small nodules in both lungs. Inflammatory streaks within the lungs. Figure 1B: Follow-up chest CT scan on March 31, 2026, following aggressive antimicrobial therapy: pulmonary inflammation has resolved compared to the previous scan.



Figure 2

The patient developed a rash again after infection control measures were

After the patient’s lung infection was brought under control, on April 8, 2026, the patient gradually developed an increasing number of red rashes on the face, neck, and anterior chest, with old rashes visible on the back.