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Pneumomediastinum: A severe complication of dermatomyositis

Shawn Zhenhui Lee, Mohammed Tousif Syed, Pranav Kumar

ABSTRACT

Dermatomyositis is an autoimmune disease that is considered a subset of idiopathic inflammatory myopathy. It is characterized by a skin rash with progressive muscle weakness. Pneumomediastinum is a rapidly progressive complication of dermatomyositis, which is hypothesized to be a result of ruptured subpleural cysts, pulmonary vasculopathy, and steroid-mediated weakening of alveolar walls. This complication is associated with a high mortality rate of 30%. Hence, it should be rapidly investigated and treated when there is a high clinical suspicion. In this article, we report a rare case of patient with dermatomyositis (DM) who developed pneumomediastinum and discussed a literature review of this occurrence.

Keywords: Dermatomyositis, Dyspnoea, Pneumomediastinum

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INTRODUCTION

Dermatomyositis (DM) is an autoimmune inflammatory myopathy, characterized by muscle weakness in combination with a pruritic or burning rash predominantly present in sun-exposed areas of the body [1]. This report evaluates the presentation of pneumomediastinum (PnM) in a patient with DM. An extensive review of the literature reveals that this finding is not as uncommon as previously thought.

CASE REPORT

A 48-year-old Filipino female was diagnosed with DM, after presenting with Gottron papules, a heliotrope rash, and muscle weakness. Laboratory studies and muscle biopsy were used to confirm her diagnosis. She was treated in the Philippines and was discharged on a drug regime which included prednisolone, hydroxychloroquine (HCQ) 200 mg daily, and mycophenolate mofetil (MMF) 500 mg daily.

On her return to Australia, her general practitioner (GP) referred her to the outpatient department due to a poor clinical response to treatment after eight months. On outpatient review, she was found to have ongoing cutaneous manifestations and shortness of breath (SOB) (Figure 1A–C). She also had per rectum bleeding, a weight loss of 12 kg in the past one year, severe proximal muscle weakness limiting her mobility, and ongoing arthralgia primarily affecting her wrists, interphalangeal joints, and left knee. On physical examination, she had a blood pressure of 118/78 mmHg, a heart rate of 110 bpm, a respiratory rate of 16 bpm, oxygen saturation of 96% on room air, and was afebrile with a temperature of 37.1°C. She was cachectic with a typical heliotrope rash over her eyelids, Gottron papules localized to her hands and feet, and swollen metacarpophalangeal and carpometacarpal joints. Respiratory examination revealed bibasal, end inspiratory crepitations and coarse crackles. She had proximal weakness with muscle power graded 4/5 in the upper and lower extremities bilaterally.

Initial investigations included a comprehensive laboratory panel, computed tomography (CT) of her chest, abdomen and pelvis, a positron emission tomography (PET)-CT scan, gastroscopy, and colonoscopy. Her creatine kinase (CK) was normal (84 U/L). Her gamma-glutamyltransferase (GGT: 257 U/L), alanine transaminase (ALT: 127 U/L), and aspartate transaminase (AST: 108 U/L) were all elevated. Her autoimmune panel which consisted of rheumatoid factor, antinuclear antibodies, anti-dsDNA, anti-neutrophil cytoplasmic antibodies (ANCA) and extractable nuclear antigens (including anti-soluble liver antigen/liver-pancreas (SLA/LP), anti-LC-1, anti-gp210, anti-PML, anti-sp100, anti-3E, and anti-Mi-2) each returned negative results. Tests for hepatitis, HIV, tuberculosis, syphilis, G6PD deficiency and thyroid disease were also negative.

Imaging of the chest, abdomen, and pelvis revealed extensive PnM and multiple bilateral peripheral patches of consolidation (Figure 2A–C). In her previous chest radiograph five years prior, she was noted to have bilateral apical fibrosis so extensive progression was evident. Following CT, a PET-CT scan was arranged to exclude malignancy and it revealed mild fluorodeoxyglucose (FDG) uptake (SUV_{max}: 4) within the multifocal, predominantly peripheral, and subpleural pulmonary consolidation. Other findings on imaging included subcutaneous emphysema (SE), peribronchial thickening, and ground glass opacities with mild avidity. There was no dominant nodule or mass visible. There was also mildly increased multifocal FDG uptake throughout the proximal upper and lower limbs and larger muscle groups of the shoulder and pelvic girdles. There was no associated abdominal or pelvic mass or lymphadenopathy. Finally, the colonoscopy and gastroscopy were performed and did not reveal any concerning pathologies.

The patient was commenced on Prednisolone 50 mg once daily. A bronchoscopy was then performed, revealing a posterior tracheal wall disruption (Figure 2D) with bronchoalveolar lavage (BAL) showing bronchial epithelial cells, fungal elements, and alveolar macrophages. She was then prescribed Methylprednisolone IV 500 mg daily and antibiotics consisting of ceftriaxone and clarithromycin. Following clinical improvement on day four of admission, she was discharged from the hospital on Prednisolone 50 mg daily and MMF 1g BD, with outpatient follow-up arranged. Few days later, the patient was seen by her GP and was given antibiotics, which was not effective. She re-presented to the hospital one month later following a two-week history of increasing swelling in her neck and face associated with difficulty swallowing solids; she denied shortness of breath. The patient was arranged for an inter-hospital transfer to a tertiary center for persisting PnM confirmed on repeat chest X-ray (Figure 2E). A repeat bronchoscopy did show same anatomical defect in the tracheobronchial tree. A barium contrast swallow study revealed no concerning features and importantly, no sign of esophageal tear. As the patient showed clinical improvement, she was discharged home. At a one-month review, her chest radiograph (CXR) showed ongoing blunting of the left costophrenic angle, similar to her previous study, and a more prominent extensive SE overlying the neck and chest when compared to previous CXR, suggesting further deterioration. She was admitted to the hospital for observation and was managed conservatively with a constant input from cardio-thoracic team. One stable, she was discharged for outpatient follow-up.

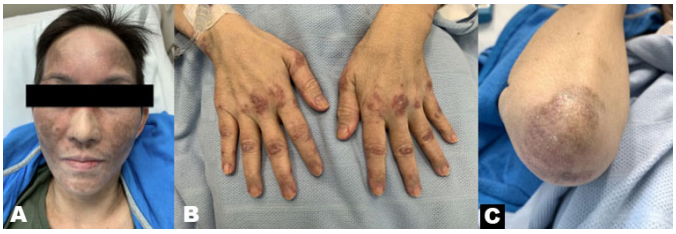

Figure 1: (A) Heliotrope rash with associated mid-facial edema involving the nasolabial folds; (B) Multiple Gottron papules in bilateral metacarpal and interphalangeal joints; (C) Gottron sign in right elbow.

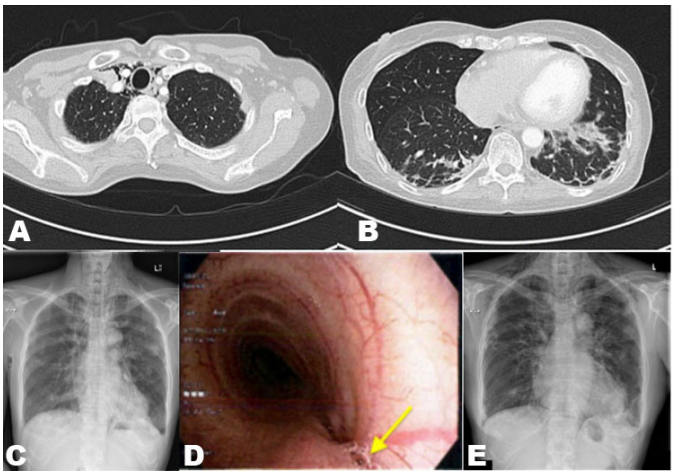

Figure 2: (A) CT chest: Multiple peripheral patches of consolidation are seen in both lungs. Small area of consolidation is also seen involving the inferior aspect of the posterior segment of left upper lobe of lung. Subsegmental atelectasis is seen involving both lower lobes basal segments; (B) CT chest: There is significant pneumomediastinum; (C) CXR: New extensive pneumomediastinum, subcutaneous surgical emphysema in the lower neck and left supraclavicular fossa, and new left lower lobe pulmonary infiltrate; (D) Bronchoscope: Tracheal wall defect at 5 o'clock position likely causing pneumomediastinum and subcutaneous emphysema in patient (yellow arrow); (E) Repeat CXR one month later: There is surgical emphysema over the chest wall extending to the neck with pneumomediastinum and no pneumothorax. There are some increased peribronchial lung markings peripherally in the right mid and at both lung bases particularly the left.

DISCUSSION

Dermatomyositis is an autoimmune condition that involves rash and muscle weakness with an estimated annual incidence of 1.9–7.7 cases per million people and a prevalence of 20 cases per million people with a predilection toward females (2:1) [2]. A five-point criterion was first conceptualized in 1975 by Bohan and Peter, who combined clinical findings, investigation results, and pathological features in order to diagnose this. The requirements include the presence of typical DM rashes, symmetrical proximal muscle weakness, elevated serum levels of muscle-associated enzymes, muscle biopsy showing evidence of myositis and myopathic changes on electromyography. Definite DM was defined as the presence of a rash with three other features; probable DM was defined as the presence of a rash with two other features; and possible DM was defined as the presence of rash with one other feature (Table 1) [3].

Dermatomyositis is sometimes associated with respiratory disease, such as bronchiolitis obliterans, organizing pneumonia, interstitial pneumonitis, and diffuse alveolar damage, in up to 50% of patients [4]. The authors would like to further add that spontaneous PnM is a rare but distinctive complication of DM. The first reported case of PnM in a patient with DM was in 1986 [5]. To date, there are 63 reported cases of PnM in the context of DM, presenting at a mean age of 40.7 years old, with an age range between 10 and 74 years (Table 2) [1, 4–13]. Dermatomyositis can happen in any age groups, however, previous case reports suggest that are more common in the adult population with only 3 children. In these 63 cases, 36 were male and 27 were female. Pneumomediastinum in DM has been associated with a poor prognosis, especially when the patient has a normal CK level. Previous research reveals that the mortality rate of PnM in DM patients may reach as high as 50%, a rate which is directly related to the severity of the patient's interstitial lung disease (ILD) [6, 7]. In our literature review of 63 patients, 20 patients (31.7%) had died, with the deaths mostly related to rapidly progressive ILD.

There are three hypotheses surrounding the pathogenesis of PnM in DM. The first of these is the rupture of subpleural cysts secondary to raised intra-alveolar pressure in ILD patients with previously damaged alveoli. Secondly, pulmonary vasculopathy causing disruption of the mucosal barrier and rupture of airway lesions has been proposed. Finally, the use of glucocorticoids in the treatment of ILD resulting in alveolar wall weakening and thus increasing the risk of rupture has also been suggested [4, 8, 9]. Known risk factors for the development of PnM in DM include the presence of ILD, cutaneous vasculopathy, hoarseness of the voice, laryngeal lesions, previous steroid use, younger age, and normal CK levels [8, 10].

Previously reported clinical features of DM-specific patients developing PnM include SOB, DM-specific dermatological features, no or little evidence of muscular involvement, the presence of cutaneous vasculopathy, normal or slightly elevated levels of CK, ILD and a history of systemic glucocorticoid use [13]. The differential diagnoses for dyspnoea in patients with DM should include: pulmonary infections, aspiration pneumonia from esophageal dysfunction, concomitant ILD or drug-mediated ILD, pneumothorax, and hypoventilation from weakened respiratory muscle activity [8].

There are four categories of investigations that can be performed to assess this. These include laboratory studies, imaging, electromyography, and histology. In laboratory testing, the two foci are the assessment of muscle-associated enzyme levels, which include CK, aldolase, lactate dehydrogenase (LDH) and AST, as well as an autoimmune screen [3]. Of the muscle enzyme levels, the most sensitive and specific is CK [3, 14]. Autoimmune assessment of DM is complex and fraught with low sensitivities [15]. Such tests should be subclassified into DM-specific autoantibodies and DM-associated

Table 1: Clinical features of DM

Cutaneous manifestations	Characteristic	Face: Heliotrope rash Neck: Shawl sign, V sign Upper extremities: Gottron papules, Gottron sign, Mechanic's hands, Nail fold telangiectasia Lower extremities: Holster sign
	More commonly in Juvenile DM	Cutaneous calcinosis
	Rare	Non-scarring alopecia, Erythroderma, Veisculobullous lesions, Leukocytoclastic vasculitis, Livedo reticularis
Muscular manifestations	Progressive symmetrical truncal and upper and lower limb proximal muscle weakness.	
Extramuscular manifestations	Joint	Athralgia and arthritis of wrists, knees and small joints of hands
	Cardiac	Arrhythmia, conduction abnormalities, cardiac arrest, congestive heart failure, myocarditis, pericarditis, angina
	Pulmonary	Interstitial lung disease, aspiration pneumonia, hypoventilation
	Gastrointestinal	Nasal speech, hoarseness, nasal regurgitation

Table 2: Previous cases of pneumomediastinum in patients with DM categorized by age

Age/ Gender	Symptoms/Signs			Investigations				Initial management prior to pneumothorax, pneumomediastinum or subcutaneous emphysema	Progression	References
	Dermatological	Muscular	Respiratory	Bloods	Autoimmune panel	Imaging	Biopsy			
10/F	• Gottron's papules, photosensitivity, alopecia	• Proximal upper and lower muscle weakness	• Subcutaneous emphysema	CK: 104 U/L, LDH: 1980 U/L, AST: Not done	Unknown	CT Chest: Pneumomediastinum, subcutaneous emphysema, basal opacities suggestive of interstitial lung disease (ILD) and epidural emphysema	Muscle biopsy: Consistent with dermatomyositis	Steroid	Initial management of chloroquine and methotrexate → Diagnosis →Steroid and Cloxacillin management → Development of subcutaneous emphysema → Continued on steroid management → Resolution of subcutaneous emphysema	[31]
16/M	• Periungual erythema • Gottron's papules • Erythema on both elbows, knees and shoulders	• Bilateral arthralgia • Slight proximal muscle weakness (4/5)	• Acute chest pain	CRP: 0.6mg/dL, CK: 43 U/L, Aldolase: 3 U/L, Myoglobin: 85 ng/mL, AST: 333 IU/L, ALT 35 IU/L, LDH: 339 U/L	ANA, Anti-Jo-1: –ve	CXR: Pneumomediastinum, pneumothorax, and subcutaneous emphysema CT: Pneumomediastinum around the aorta and in the pericardial space, left pneumothorax and subcutaneous emphysema expanding from the supraclavicular fossa to the cervix. Slight interstitial changes in left lower lobe.	Skin biopsy: lymphocytic infiltration around the capillaries, and mucin deposits between the collagen bundles of mid dermis Muscle biopsy: Infiltration of the lymphocytes around interstitial vessels though degeneration, and atrophy of muscle fibers was not observed	Strict bed rest	Initial diagnosis → Strict bed rest for 2 months → Ongoing skin ulcers and interstitial lung disease requiring steroid and CY A → Symptom resolution and nil recurrence on 4 years follow-up	[22]
18/F	• Scarring lesions in the finger pads, periungual erythema, hypopigmented macules, • Previous Gottron's papules in MCPs and PIPs • Indurated subcutaneous nodules in forearms and thighs	• NIL	• SOB, ILD	ESR: 84 mm/h, CRP: 17 mg/L, CK: 40 U/L (N), Aldolase: 2 IU/L (N),	ANA: 1/80, nucleolar pattern, Anti-Ro52: +ve, Anti-MDA5: +ve, RF, Anti-CCP, Anti-DNA, Anti-ENA, Anti-Scl70, Anti-centromere, APLA, Anti-Mi2, Anti-Jo1, Anti-PL12, Anti-PL7, Anti-OJ, Anti-EJ, Anti-SRP, Anti-Ju, an Anti-PM/ Scl: –ve	CXR: Bibasilar opacities and spontaneous pneumothorax, CT scan: Patchy peribronchial opacities of basilar predominance, suggestive of organizing pneumonia and an extensive pneumomediastinum	Muscle biopsy: Nonspecific myopathic changes without inflammatory infiltrate	Steroid & Antibiotics	Initial presentation →Steroid and IV Antibiotics management → Partial improvement of SOB → Addition of Methotrexate and folic acid management →Interhospital transfer → Diagnosis → Steroid and AZA management → Radiological improvement of spontaneous pneumothorax → Lost to follow-up	[33]
20/F	• Heliotrope rash • Gottron's signs • Shawl sign • Periungual hypertrophy	• Proximal upper and lower limbs weakness	• SOB, Interstitial pneumopathy	CK: 3031 U/L	Anti-Mi2 and Anti-MDA-5: –ve	Unknown	Unknown	Steroid, AZA, Methotrexate	Clinical remission	[7]

Table 2: *(Continued)*

Age/ Gender	Symptoms/Signs			Investigations				Initial management prior to pneumothorax, pneumomediastinum or subcutaneous emphysema	Progression	References
	Dermatological	Muscular	Respiratory	Bloods	Autoimmune panel	Imaging	Biopsy			
20/F	• Heliotrope rash, Malar rash • Gottron's papules	• Polyarthralgia, No weakness	• SOB, Neck pain	CK: 293 IU/L, Aldolase: normal	ANA, Anti-dsDNA, Anti-Jo-1: –ve	CXR & CT Chest: Progression of subcutaneous emphysema, pneumomediastinum, thickening of interlobular septa, and a reticulonodular pattern	Muscle biopsy: No abnormal finding	Steroid, AZA	Initial diagnosis → Steroid & AZA therapy → Ongoing cutaneous manifestation → Addition of Hydroxychloroquine→Dyspnoea→ Increased steroid and commenced CYC → Steroid pulse and Rituximab therapy → ICU admission for severe hypoxemia → Chest tube insertion into both pleural spaces → Intubated → Percutaneous tracheostomy 1 week later → Deceased secondary to severe hypoxemia despite intensive therapy	[36]
23/M	• Heliotrope rash • Gottron's sign	• No muscle weakness	• Subcutaneous emphysema	CK: 24 U/L	Anti-MDA5: +ve, Anti-ARS, Anti-SRP, Anti-TIFγ, Anti-Mi2, Anti-SAE, Anti-NXP2: –ve	Unknown	Unknown	Unknown	Lost to follow-up	[12]
23/M	• Cutaneous vasculopathy	• No muscle weakness	• Interstitial pneumonitis	CK 219 IU/L	Unknown	Unknown	Unknown	Steroid	Initial diagnosis → Steroid management → Alive	[30]
25/M	• Cutaneous vasculopathy	• No muscle weakness	• Interstitial pneumonitis	CK: 377 IU/L	Unknown	Unknown	Unknown	Steroid, AZA, CSA	Initial diagnosis → Steroid, AZA and CY management → Deceased	[30]
25/M	• Heliotrope rash, digital tip ulceration, alopecia	• Proximal upper and lower limb weakness	• Radiological evidence of pneumomediastinum	CK: 56 U/L, LDH: Not done, AST: 65 U/L	Anti-RNP 28 U/mL (raised), Anti-SSA 55 Ru/mL (raised)	HRCT Chest: pneumomediastinum, irregular reticular lines and ground glass opacities with basal and subpleural predominance seen bilaterally	Muscle biopsy: Consistent with dermatomyositis	NIL	Initial presentation and diagnosis → Steroid and MMF management → Symptomatic improvement	[31]
27/M	• Heliotrope rash • Gottron's signs • Periungual hypertrophy	• Proximal upper and lower limbs weakness	• SOB, Interstitial pneumopathy, Pulmonary fibrosis	CK: 190 U/L	Anti-Mi2 and Anti-MDA-5: –ve	Unknown	Unknown	Steroid, AZA, Methotrexate, CY	Clinical remission	[7]
28/M	• Heliotrope rash • Gottron's signs • Shawl sign • Periungual hypertrophy	• Proximal upper and lower limbs weakness	• SOB, Interstitial pneumopathy	CK: 65 U/L	Anti-Mi2 and Anti-MDA-5: –ve	Unknown	Unknown	Steroid, AZA, Methotrexate, CY	Complete clinical response	[7]
28/M	• Heliotrope rash, periungual erythema and erythematous rash over dorsum of hands, Gottron's papules	• Fatigue, Proximal muscle weakness of limbs	• Interstitial pneumonitis	ESR: 30 mm/h, CK: 287 U/L, AST: 93 U/L, ALT: 49 U/L	ANA, Anti-Jo-1, RF: –ve	CXR: Mild alveolar and interstitial pattern in both lower lobes	Muscle biopsy: Degeneration of muscle fibers and a mild degree of mononuclear cell infiltration	Steroid	Initial diagnosis → Steroid management → ILD, skin and muscular changes →Cyclosphosphamide and steroid therapy → Complete resolution of subcutaneous emphysema	[28]

Table 2: *(Continued)*

Age/ Gender	Symptoms/Signs			Investigations				Initial management prior to pneumothorax, pneumomediastinum or subcutaneous emphysema	Progression	References
	Dermatological	Muscular	Respiratory	Bloods	Autoimmune panel	Imaging	Biopsy			
30/M	• Heliotrope rash, Gottron's papules, Periungual telangiectasia	• No proximal muscle weakness	• SOB	CK: Normal, LDH: Normal, Aldolase: Normal	Unknown	CXR: subcutaneous emphysema in the laterocervical spaces HRCT Chest: scattered "ground glass" opacities, subcutaneous emphysema and a pneumomediastinum	Unknown	Steroid	Initial diagnosis → Steroid therapy for 1 month→Dyspnoea Steroid and CYC therapy → Resolved Dyspnoea→ Relapse of ILD and pneumomediastinum 2 years later MMF → Radiological resolution at 6 months and 12 months follow-up.	[20]
30/M	• Cutaneous vasculopathy	• NIL	• Interstitial pneumonitis	CK: 403 IU/L	Unknown	CT Chest: Air around trachea and in anterior mediastinum. Honeycomb pattern in posterior bases of lungs	Muscle biopsy: No abnormal findings	Steroid, CSA	Initial diagnosis → Steroid & CSA management → Progression of interstitial pneumonitis with subsequent development of pneumomediastinum and subcutaneous emphysema → Steroid and CY management → Resolution of symptoms	[30]
31/F	• Gottron's papules	• Nil	• Late inspiratory crackles	Unknown	Anti-Jo-1: –ve, ANA: 1:80 +ve, KL-6: 3,050 U/mL	CT scan: Subpleural patchy ground glass attenuation and consolidation in both lobes, with bronchiectasis	Unknown	Steroid, CY	Initial diagnosis → Steroid and CY therapy → Massive pneumothorax, subcutaneous emphysema → Compression of major vessels and main bronchi → Deceased	[25]
31/M	• Heliotrope rash • Gottron's signs • Periungual hypertrophy	• Proximal upper and lower limbs weakness	• SOB, Interstitial pneumopathy, Pulmonary fibrosis	CK: 120 U/L	Anti-Mi2 and Anti-MDA-5: –ve	Unknown	Unknown	Steroid, AZA, Methotrexate, Leflunomide, CY	Clinical remission	[7]
33/M	• Heliotrope rash • Gottron's signs • Periungual hypertrophy	• No weakness	• SOB, Interstitial pneumopathy	CK: 124 U/L	Anti-Mi2 and Anti-MDA-5: –ve	Unknown	Unknown	Steroid, AZA	Deceased	[7]
33/M	• Heliotrope rash, Gottron's lesion	• Slow progressive weakness in shoulders, hip girdle and thighs • Weak neck muscles	• Subcutaneous emphysema	CK: 171 U/L, LDH: 1195 U/L, AST: 554 U/L	Extractable nuclear antigens: –ve	HRCT Chest: pneumomediastinum, non-specific interstitial pneumonia (NSIP) with ground glass opacity opacity; features of bronchiolitis obliterans with organizing pneumonia	Muscle biopsy: Mild fiber atrophy	Steroid	Initial diagnosis → Steroid treatment → Worsening symptoms & subcutaneous emphysema →Deflazacort and MMF management → Symptomatic improvement on follow-up	[31]

Table 2: *(Continued)*

Age/ Gender	Symptoms/Signs			Investigations				Initial management prior to pneumothorax, pneumomedia-stinum or subcutaneous emphysema	Progression	References
	Dermatological	Muscular	Respiratory	Bloods	Autoimmune panel	Imaging	Biopsy			
34/M	• Macular erythema at neck and arm extensors • Gottron papules	• NIL	• SOB with bilateral fine crackles	Biochem: N, ESR 70 mm/h, CRP: 32.3 mg/dL, CK: Normal, Aldolase: Normal	ANA, ANCA, RF, and other immune markers were all negative Anti-SS-A(Ro) :+ve	CXR: Bilateral opacities, CT: Bilateral ground glass opacity in subpleural areas, consolidation, and pneumomediastinum	Skin biopsy: No specific pathology	Sulfasalazine for arthritis	Initial presentation with respiratory symptoms → ICU support for severe hypoxemic respiratory failure → Steroid + CYC management → Discharged on MMF and Steroids → Re-presentation for refractory severe hypoxemic respiratory failure 1 month later → Given immunosuppressive therapy and mechanical ventilation → Deceased	[1]
36/M	• Unknown	• Unknown	• Interstitial lung disease	Unknown	Unknown	Unknown	Unknown	Unknown	Deceased	[10]
38/F	• Erythema on the eyelids, fingers, elbows and knees • Gottron's sign • Skin ulcers on fingertips, both elbows and knees	• Proximal muscle weakness on both lower and upper extremities	• Acute chest pain and SOB	CK: 131 IU/L, Aldolase: 13.2 IU/L AST: 43 U/L, ALT: 55 U/L, LDH: 277 IU/L	ANA, Anti-SS-A, Anti-SS-B, Anti-Sm, Anti-RNP, Anti-Scl70, Anti-Jo-1, RF: –ve	CXR: Pneumothorax, pneumomediastinum and subcutaneous emphysema CT scan: Multiple ground glass opacity in both mid and lower lung fields, subcutaneous emphysema and pneumomediastinum	Muscle biopsy: Consistent with dermatomyositis	Steroid, Hydroxychloroquine	Initial diagnosis → Steroid and Hydroxychloroquine management → Acute chest pain and SOB → Addition of CY → Symptom resolution & recurrence free at 7 months follow-up	[13]
38/F	• Heliotrope rash, • Gottron's sign, • Periungual erythema	• Muscle weakness	• Unknown	CK: 2379 U/L	Anti-MDA5: +ve, Anti-ARS, Anti-SRP, Anti-TIFγ, Anti-Mi2, Anti-SAE, Anti-NXP2: –ve	Unknown	Unknown	Unknown	Deceased	[12]
38/M	• Periungual erythema on fingers • Typical Gottron's papules on MCP, PIP and extensor areas of elbows and knees	• Proximal muscle weakness	• Radiological pneumomedia-stinum and cervical subcutaneous emphysema	ESR: 27 mm/h, CRP: Normal, CK: 227 IU/L, LDH: 311 IU/L, Aldolase: 6.6 IU/L, AST: 56 IU/L, ALT: 80 IU/L	ANA, dsSNA, Anti-Jo-1, ANCA, RF: –ve	CT Chest: Pneumomediastinum and cervical subcutaneous emphysema	Muscle biopsy: Slight degeneration and atrophy of muscle fibers, and infiltrates of mononuclear cells, such as lymphocytes, plasma cells, and macrophages around the small vessels in connective tissue around the muscle	Steroid	Initial diagnosis → Steroid therapy → Initial improvement, followed by worsening myalgia and radiological evidence of pneumomediastinum and subcutaneous emphysema → CYC, Steroid and Methotrexate therapy → Radiological improvement with no recurrence	[29]
39/F	• Unknown	• Unknown	• Interstitial lung disease	Unknown	Unknown	Unknown	Unknown	Unknown	Deceased	[10]

Table 2: *(Continued)*

Age/ Gender	Symptoms/Signs			Investigations				Initial management prior to pneumothorax, pneumomediastinum or subcutaneous emphysema	Progression	References
	Dermatological	Muscular	Respiratory	Bloods	Autoimmune panel	Imaging	Biopsy			
39/F	• Gottron's signs • Periungual hypertrophy	• Proximal upper and lower limbs weakness	• SOB, Interstitial pneumopathy, Pulmonary fibrosis	CK: 674 U/L	Anti-Mi2 and Anti-MDA-5: –ve	Unknown	Unknown	Steroid, AZA, CYC	Complete clinical response	[7]
40/F	• Heliotrope rash, • Gottron's sign, • Periungual erythema, • Skin ulcer	• No muscle weakness	• Subcutaneous emphysema	CK: 1170 U/L	Anti-Mi2: +ve, Anti-ARS, Anti-SRP, Anti-TIFγ, Anti-MDA5, Anti-SAE, Anti-NXP2: –ve	Unknown	Unknown	Unknown	Alive	[12]
41/F	• Cutaneous vasculopathy	• Unknown	• Interstitial lung disease	CK: Normal, Aldolase: Normal	Unknown	CXR: Pneumomediastinum, HRCT Chest: Pneumomediastinum	Tracheal biopsy: Unspecified inflammatory disease with predominant polymorphonuclear infiltrate	Steroid & immunosuppressives	Initial diagnosis → Steroid and immunosuppressive management → Interstitial lung disease → Deceased	[19]
42/F	• Unknown	• No muscle weakness	• Unknown	CK: Normal	Unknown	HRCT Chest: Ground glass opacities, subpleural blebs	Muscle biopsy: No changes	Unknown	Lost to follow-up	[6]
42/F	• Unknown	• Muscle weakness	• Unknown	CK: Elevated (5 x Normal)	Unknown	HRCT Chest: Diffuse opacities predominant in the basal area	Muscle biopsy: Typical inflammatory changes	Unknown	Resolution	[6]
42/F	• Unknown	• Muscle weakness	• Unknown	CK: Elevated (5 x Normal)	Unknown	HRCT Chest: Diffuse opacities predominant in the basal area	Muscle biopsy: Typical inflammatory changes	Unknown	Resolution	[6]
42/F	• Unknown	• No muscle weakness	• Unknown	CK: Normal	Unknown	HRCT Chest: Ground glass opacities, paracardiac blebs	-	Unknown	Deceased 9 months after pneumomediastinum	[6]
42/F	• Unknown	• No muscle weakness	• Unknown	CK: Normal	Unknown	HRCT Chest: Ground glass opacities, paracardiac blebs, honeycomb cysts	Muscle biopsy: Typical inflammatory changes	Unknown	Deceased 2 months after pneumomediastinum	[6]
42/F	• Heliotrope rash, • Chest or back erythema, • Gottron's sign	• Muscle weakness	• Unknown	CK: 67 U/L	Anti-MDA5: +ve, Anti-ARS, Anti-SRP, Anti-TIFγ, Anti-Mi2, Anti-SAE, Anti-NXP2: –ve	Unknown	Unknown	Unknown	Alive	[12]
42/M	• Unknown	• No muscle weakness	• Unknown	CK: Normal	Unknown	HRCT Chest: Ground glass opacities	Muscle biopsy: No changes	Unknown	Resolution	[6]
42/M	• Unknown	• No muscle weakness	• Unknown	CK: Normal	Unknown	HRCT Chest: Ground glass opacities, paracardiac blebs	Muscle biopsy: No changes	Unknown	Resolution	[6]

Table 2: *(Continued)*

Age/ Gender	Symptoms/Signs			Investigations				Initial management prior to pneumothorax, pneumomedia-stinum or subcutaneous emphysema	Progression	References
	Dermatological	Muscular	Respiratory	Bloods	Autoimmune panel	Imaging	Biopsy			
42/M	• Unknown	• Muscle weakness	• Unknown	CK: Normal	Unknown	HRCT Chest: Honeycomb cysts, paracardiac blebs	Muscle biopsy: Typical inflammatory changes	Unknown	Resolution	[6]
42/M	• Gottron's papules on the MCP and proximal joints, • Periorbital heliotrope rash	• Mild myalgias and moderate proximal muscular weakness	• Anterior neck pain and SOB	Unknown	RF, Anti-platelet antibodies, cyroglobulin, ANA, ANCA, Anti-cardiolipin: all –ve	CXR & CT scan: Subcutaneous emphysema, pneumomediastinum and diffuse reticulonodular infiltration in both lungs	Muscle biopsy: Moderate necrosis of the muscular fibers	Steroid, Methotrexate, Hydroxychloro-quine	Initial diagnosis → Steroid, Methotrexate and Hydroxychloroquine therapy SOB, neck pain &bilateral inspiratory crackles → IV CYC & IVIG → Severe condition but stable at 1 year after diagnosis	[26]
42/M	• Heliotrope rash, • Gottron's papules	• Lower limbs weakness	• Asymptomatic subcutaneous emphysema	CK: 2260 IU	Unknown	CXR: Increased interstitial markings	Unknown	Steroid, AZA	Initial diagnosis → Steroid management → Readmission due to muscle weakness → IV steroid management → Development of bilateral aspiration pneumonia → Steroid and AZA management → Development of a long sinus tract with purulent drainage and local cellulitis → Increase in AZA and reduction of Steroid → Asymptomatic subcutaneous emphysema and extensive pneumomediastinum managed as outpatient as per patient → Reduction in subcutaneous emphysema on 1 month follow-up	[5]
42/M	• Chest or back erythema • Gottron's sign • Periungual erythema, • Skin ulcer	• No muscle weakness	• Unknown	CK: 1127 U/L	Anti-MDA5: +ve, Anti-ARS, Anti-SRP, Anti-TIFγ, Anti-Mi2, Anti-SAE, Anti-NXP2: –ve	Unknown	Unknown	Unknown	Alive	[12]
43/M	• Heliotrope rash, • Chest or back erythema, • Gottron's sign	• Muscle weakness	• Subcutaneous emphysema	CK: 4306 U/L	Anti-MDA5: +ve, Anti-ARS, Anti-SRP, Anti-TIFγ, Anti-Mi2, Anti-SAE, Anti-NXP2: –ve	Unknown	Unknown	Unknown	Deceased	[12]
44/M	• Chest or back erythema • Gottron's sign • Periungual erythema	• Muscle weakness	• Subcutaneous emphysema	CK: 3467 U/L	Anti-MDA5: +ve, Anti-ARS, Anti-SRP, Anti-TIFγ, Anti-Mi2, Anti-SAE, Anti-NXP2: –ve	Unknown	Unknown	Unknown	Deceased	[12]

Table 2: *(Continued)*

Age/ Gender	Symptoms/Signs			Investigations				Initial management prior to pneumothorax, pneumomediastinum or subcutaneous emphysema	Progression	References
	Dermatological	Muscular	Respiratory	Bloods	Autoimmune panel	Imaging	Biopsy			
44/M	• Unknown	• Unknown	• Interstitial lung disease	Unknown	Unknown	Unknown	Unknown	Unknown	Deceased	[10]
45/F	• Vasculitis-like skin lesions on the dorsum of both hands • Heliotrophic-like skin rash	• NIL	• Increased interstitial lung infiltrates in both lower lung fields	CK: Normal, LDH: Normal	ANA, dsDNA, Anti-Smith, SSA, SSB, Jo-1, Scl-70, Centromere, ANCA, RF, Anti-CCP: –ve	HRCT Chest: Reticulonodular and scattered ground grass appearance in the lower lung fields	Lung biopsy: Mild chronic inflammatory cell infiltrate admixed with the spindle cells	Steroid, AZA	Initial diagnosis → Steroid and AZA management → Diagnosis of Invasive breast cancer → Chemotherapy → Subcutaneous emphysema and pneumomediastinum→ Continuation of adjuvant chemotherapy as no severe respiratory symptoms → Improvement on follow-up CT scan	[11]
45/M	• Heliotrope rash • Periungual hypertrophy	• No weakness	• SOB, Interstitial pneumopathy	CK: 120 U/L	Anti-Mi2 and Anti-MDA-5: –ve	Unknown	Unknown	Steroid, CY, CYC	Complete clinical response	[7]
46/F	• Skin vasculopathy	• Myositis	• Head and neck were swollen and subcutaneous emphysema and crepitance observed	CK: 1280 U/L	Unknown	CT Scan: Pneumomediastinum& Subcutaneous emphysema from head to upper arm	Unknown	Steroid, CY	Initial diagnosis → Steroid and CY therapy → Subcutaneous emphysema and crepitance on day 30 of admission → Improvement of pneumomediastinum and subcutaneous emphysema → Rapid deterioration of ILD → Deceased	[27]
46/M	• Heliotrope rash • Gottron's papules	• Mild proximal muscle weakness • Mild atrophy of quadriceps, iliopsoas, and gastrocnemius	• SOB on re-presentation	Biochem: ?, ESR: 35 mm/h, CRP: Negative, CK: 395 IU/L (elevated), LDH: 228 IU/L (elevated), Aldolase: Myoglobin 89.0 ng/mL Transaminase elevated	ANA, SS-A, SS-B, Sm, dsDNA, RNP, Scl-70, Jo-1 & RF: –ve	CXR: Pulmonary fibrosis in lower lobes of lungs, CT scan: honey-comb pattern in the bilateral lower lobes	Muscle biopsy: Small groups of necrotic fibers, some variation in fiber size, muscle fibrosis, and mononuclear cell infiltration of lymphocytes around the small vessels	Steroid	Initial diagnosis → Steroid management for 14 days → subcutaneous emphysema → Steroid + Cyclosporin A therapy → Resolution of symptoms and signs with nil recurrence on 2 month follow-up	[4]
46/M	• Heliotrope rash • Gottron's signs • Periungual hypertrophy	• Proximal upper and lower limbs weakness	• SOB, Interstitial pneumopathy, Pulmonary fibrosis	CK: 170 U/L	Anti-Mi2 and Anti-MDA-5: –ve	Unknown	Unknown	Steroid, AZA, Methotrexate, CYC, CY	Complete clinical response	[7]
50/M	• Facial erythema, scaly papules on the fingers, helicotrope rash, shawl sign, Gottron papules	• Weakness of flexors of neck, shoulders and hips	• Generalized hypoventilation with Velcro rales in both lung bases	ESR: positive, CRP: positive, CK: 309 IU/L, AST: 74 IU/L	ANA: 1/160, positive speckled and nucleolar	CT scan: Pulmonary interstitial involvement with little ground glass, pneumomediastinum associated with pericardial compromise	NIL	Steroid, AZA	Initial diagnosis → Steroid and AZA management → Symptomatic resolution	[9]

Table 2: *(Continued)*

Age/ Gender	Symptoms/Signs			Investigations				Initial management prior to pneumothorax, pneumomediastinum or subcutaneous emphysema	Progression	References
	Dermatological	Muscular	Respiratory	Bloods	Autoimmune panel	Imaging	Biopsy			
51/F	• Gottron's sign • Skin ulcer	• Muscle weakness	• Pneumothorax and Subcutaneous emphysema	CK: 5800 U/L	Anti-MDA5: +ve, Anti-ARS, Anti-SRP, Anti-TIFγ, Anti-Mi2, Anti-SAE, Anti-NXP2: –ve	Unknown	Unknown	Unknown	Deceased	[12]
52/F	• Confluent and violaceous erythema on upper chest • Violaceous, ulcerated plaques on MCP joints, DIP joints, and elbows bilaterally	• Nil	• SOB on re-presentation	CK: Normal, Aldolase: Normal,	Anti-Jo-1: –ve	CT scan: Bilateral pneumomediastinum with subcutaneous emphysema	Skin Biopsy: consistent with dermatomyositis	Steroid, Hydroxychloroquine, Clobetasol	Initial diagnosis → Steroid management → Addition of HydroxychloroquineandClobetasol → SOB, facial and neck edema→ Bilateral chest tubes and CY → Discharged with significant improvement	[8]
54/F	• Heliotrope rash • Erythematous rashes over anterior chest wall	• NIL	• Sudden onset of SOB	ESR: 48mm/h, CK: 4332 IU/L (on diagnosis) → 35 IU/L (latest), AST: normal, ALT: normal	Anti-Ro: 52 (positive), Anti-smRNP, SS-A/B, Jo-1, ANA: –ve	CXR: Right sided pneumothorax affecting 50% of right hemithorax	Muscle biopsy: Perifascicular atrophy	Steroid	Diagnosis → Steroid management → SOB, tachypnea and cyanosis → Chest drain insertion → Respiratory failure despite resuscitation → Deceased	[23]
56/M	• Heliotrope rash, • Gottron's sign, • Skin ulcer	• No muscle weakness	• Subcutaneous emphysema	CK: 570 U/L	Anti-MDA5: +ve, Anti-ARS, Anti-SRP, Anti-TIFγ, Anti-Mi2, Anti-SAE, Anti-NXP2: –ve	Unknown	Unknown	Unknown	Deceased	[12]
57/F	• Heliotrope rash • Gottron's signs • Shawl sign • Periungual hypertrophy	• Proximal upper and lower limbs weakness	• SOB, Interstitial pneumopathy, Pulmonary fibrosis	CK: 41 U/L	Anti-Mi2 and Anti-MDA-5: –ve	Unknown	Unknown	Steroid	Deceased	[7]
57/M	• Skin ulcers • Cutaneous erythema • Facial and neck swelling	• Muscle weakness	• Interstitial lung disease	Unknown	Unknown	CXR: Worsening ILD and Extensive Subcutaneous emphysema CT Chest: pneumomediastinum, subcutaneous emphysema, and ground glass opacities corresponding to interstitial lesions with honeycomb pattern	Unknown	Steroid, CYC	Initial diagnosis → Steroid and CYC management → Subcutaneous emphysema → Addition of Rituximab → Symptomatic resolution	[37]

Age/ Gender	Symptoms/Signs		Investigations				Initial management prior to pneumothorax, pneumomedia-stinum or subcutaneous emphysema	Progression	References
	Dermatological	Muscular	Respiratory	Bloods	Autoimmune panel	Imaging	Biopsy			
58/F	• Unknown	• Unknown	• Neck swelling, Dysphagia	Unknown	Unknown	CXR & CT Chest: Extensive pneumomediastinum and subcutaneous emphysema. Bibasilar ground-glass and reticular opacities.	Unknown	CYC	Initial diagnosis → CYC therapy → Radiological resolution and discharged	[34]
59/F	• -	• No muscle weakness	• Interstitial pneumonitis	CK: 3501 IU/L	Unknown	Unknown	Unknown	Steroid	Initial diagnosis → Steroid management → Alive	[30]
59/M	• Unknown	• Unknown	• Interstitial lung disease	Unknown	Unknown	Unknown	Unknown	Steroid, CYC, AZA, IVIG	Initial diagnosis Steroid, CYC, AZA and IVIG management → Interstitial lung disease progression → Rituximab management → Improved lung function	[32]
60/M	• Gottron's sign • Periungualery-thema	• No muscle weakness	• Unknown	CK: 25 U/L	Anti-MDA5: +ve, Anti-ARS, Anti-SRP, Anti-TIFγ, Anti-Mi2, Anti-SAE, Anti-NXP2: –ve	Unknown	Unknown	Unknown	Alive	[12]
64/F	• Rash on forearm and lower extremities	• No muscle weakness	• Radiological evidence of pneumomediastinum	ESR: 114 mm, CRP: 34.2 mg/dL, CK: Normal, Aldolase: 15.5 U/L	Anti-Jo, Anti-SSA, MPO antibody, RF: +ve ANA, ANCA, Myositis, MDA-5, RF: –ves	CT scan: Extensive subcutaneous emphysema in the neck, groin, and buttocks, pneumomediastinum, and air in the bladder wall and retroperitoneum. Lower lobe predominant chronic interstitial infiltration and mild bronchiectasis.	Skin biopsy: Minimal perivascular inflammation and hemorrhagic crust not consistent with vasculitis	Steroid, AZA	Initial diagnosis → Steroid, AZA therapy → Several major complications including diverticular perforation and CMV reactivation	[35]
66/M	• Rash	• Residual muscle weakness	• SOB	CK: 69 IU/L	Unknown	CXR & CT scan: Severe Pneumomediastinum and ILD, mainly in the lower lobes	Unknown	Steroid, Methotrexate	Initial diagnosis → Steroid and Methotrexate management →Presented to hospital with dysphagia, dysphonia and dyspnea over 2 weeks → Given pulse steroid therapy → Discharged on high dose steroids →Weaned off steroids → Increased symptomatic resolution at 6 months follow-up, off methotrexate and tapered off steroids	[24]

Table 2: *(Continued)*

Age/ Gender	Symptoms/Signs			Investigations				Initial management prior to pneumothorax, pneumomedia-stinum or subcutaneous emphysema	Progression	References
	Dermatological	Muscular	Respiratory	Bloods	Autoimmune panel	Imaging	Biopsy			
74/M	• Chest or back erythema • Gottron's sign • Periungual erythema	• No muscle weakness	• Subcutaneous emphysema	CK: 144 U/L	Anti-MDA5: +ve, Anti-ARS, Anti-SRP, Anti-TIFγ, Anti-Mi2, Anti-SAE, Anti-NXP2: –ve	Unknown	Unknown	Unknown	Deceased	[12]
?/M	• Periorbital heliotrope rash, • Gottron's papules, • Vasculitic ulcers	• Proximal muscle weakness	• Interstitial lung disease	CK: 347 IU/mL, LDH: 437 IU/mL	Anti-SRP, Anti-PL7, Anti-Ro52: +ve	CXR: pneumomediastinum with pneumopericardium without pneumothorax, HRCT Chest: Usual interstitial pneumonia pattern of interstitial lung disease	Unknown	Steroid, CYC	Initial diagnosis → steroid & CYC management → SOB → Continued current management → Improvement of pneumomediastinum with good clinical response on 1 month follow-up.	[38]

autoantibodies. There are six subtypes of DM-specific autoantibodies, namely, anti-aminoacyl transfer RNA synthetase (ARS) (including Anti-Jo-1) anti-DNA helicase (anti-Mi2); anti-melanoma differentiation-associated gene 5 (MDA5); anti-transcription intermediary factor (TIF-1γ); anti-nuclear matrix protein-2 (NXP-2); and anti-small ubiquitin-like modifier activating enzyme (SAE) [15]. DM-associated antibodies include anti-Ku, which is involved in DNA repair, antinuclear antibody (ANA), and anti-SSA/Ro, which are antibodies for ribonucleoprotein complexes with small cytoplasmic RNAs (hY-RNA) [15, 16].

In terms of imaging modalities, chest radiograph (CXR) has a low sensitivity for early detection but is useful as a baseline assessment of the lungs and to assess for significant ILD. When diagnosed, serial CXR should be conducted to not only assess the progression of the disease, but also to determine the presence of complications, such as spontaneous pneumothorax (PTX), PnM, SE, and infection [17]. High-resolution CT (HRCT) of the chest can provide a better assessment of ILD, including findings of irregular linear opacities, consolidation, ground glass opacities, pleural effusion, and honeycombing, as well as providing information about the location and extent of PTX, PnM, and SE, when CXR is inconclusive [18]. The utility of bronchoscopy includes assessment of the site and size of laryngeal lesions, bronchial wall necrosis and the ability to perform a histological assessment when lung biopsy is performed [6, 10, 19]. Histological findings may include nonspecific interstitial pneumonia, organizing pneumonia, diffuse alveolar damage, and usual interstitial pneumonia [1]. Bronchoalveolar lavage plays a supportive role, as it may provide some information regarding disease progression, however, there is no characteristic BAL cell profile for parenchymal involvement in DM [20].

The management of DM with ILD is complex and involves a multi-disciplinary approach. With regard to dermatological presentation, non-pharmacological management includes avoidance of sunlight and using protective clothing. For extensive erythematous lesions and muscle weakness, steroids are titrated to CK levels. The addition of immunosuppressive agents and anti-pruritic agents are given as per treating rheumatologist or dermatologist recommendations, with due consideration of patient's tolerability and medication side effects [21]. The most effective treatment for ILD has yet to be decided. Currently, patients are treated with corticosteroids as first-line management. However, since high dose steroids alone are associated with poorer prognosis, patients are often given one or more immunosuppressive agents including azathioprine (AZA), cyclophosphamide (CYC), cyclosporine (CS), mycophenolate mofetil (MMF), and/or intravenous immunoglobulins (IVIG) [13, 21].

CONCLUSION

In conclusion, PnM is a rapid progressive complication of DM with concomitant ILD. A respiratory physician should regularly follow up patients with PnM to ensure that the condition can be monitored and respiratory function optimized.

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