Pneumomediastinunm: 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

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.

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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\textsubscript{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 represented 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 pneumonia, 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 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 diagnosis 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 |
|                               | More commonly in Juvenile DM | Lower extremities: Holster sign   |
|                               | Rare                        | Cutaneous calcinosis             |
| **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 | Initial diagnosis | References |
|-------|--------|---------------------------------------------------|----------------|-----------------------------------------------------------------------------------------|-------------|------------------|------------|
| 10/F  | F      | - Goltz’s papules, photosensitivity, armpits       | CT Chest       | - Initial management of chloroquine and prednisone                                     | Steroid     | -                | [31]       |
| 16/M  | M      | - Periungual erythema, Gottron’s papules          | CXR: LR       | - Initial presentation > Strict bed rest for 2 months > Ongoing steroids and intravenous  | - Steroid   | -                | [22]       |
| 18/F  | F      | - Scarring lesions in the finger pads, periungual | ESR: 84 mm/h,  | - Initial presentation > Steroid and Methotrexate management > Partial improvement of SOB | -          | -                | [33]       |
| 38/F  | F      | - Heliotrope rash, Gottron’s signs, Shawl sign    | CK: 3031 U/L   | - Initial presentation > Steroid and AZA management > Radiological improvement > SOB     | -          | -                | [7]        |
| 20/F  | F      | - Proximal upper and lower limb weakness          | CK: 43 U/L    | - Initial presentation > Steroid and Methotrexate management > Partial improvement of SOB | -          | -                | [5]        |
| Age/Gender | Symptoms/Signs | Investigations | Biopsy | Initial management prior to pneumothorax, pneumomediastinum or subcutaneous emphysema | Progression |
|------------|----------------|----------------|--------|--------------------------------------------------------------------------------------|-------------|
| 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 | Progression of subcutaneous emphysema, pneumomediastinum, thickening of interlobular septa, and a reticulonodal pattern | Muscle biopsy: No abnormal finding | Steroid, AZA | Initial diagnosis → Steroid & AZA therapy → Ongoing cutaneous manifestation → Addition of Hydroxychloroquine → Deceased due to 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 |
| 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 |
| 23/M       | Cutaneous vasculopathy | No muscle weakness | Interstitial pneumonitis | CK 219 IU/L | Unknown | Unknown | Unknown | Steroid, AZA, CSA | Initial diagnosis → Steroid & AZA therapy → Alive |
| 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 |
| 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, consistent with dermamyositis | Muscle biopsy: No abnormal finding | NIL | Initial presentation and diagnosis → Steroid and MMF management → Symptomatic improvement |
| 27/M       | Heliotrope rash, Gottron’s signs | Proximal upper and lower limb weakness | SOB, Interstitial pneumonopathy, Pulmonary fibrosis | CK: 190 U/L | Anti-Mi2 and Anti-MDA-5: -ve | Unknown | Unknown | Steroid, AZA, Methotrexate, CY | Clinical remission |
| 28/M       | Heliotrope rash, Gottron’s signs, Shawl sign | Proximal upper and lower limb weakness | SOB, Interstitial pneumonopathy | CK: 65 U/L | Anti-Mi2 and Anti-MDA-5: -ve | Unknown | Unknown | Steroid, AZA, Methotrexate, CY | Complete clinical response |
| 28/M       | Heliotrope rash, periangual erythema and erythematous rash over dorson 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 asebor 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 → Cyclophosphamide and steroid therapy → Complete resolution of subcutaneous emphysema |
| Age/Gender | Symptoms/Signs | Investigations | Autoimmune panel | Imaging | Biopsy | Initial management prior to pneumothorax, pneumomediastinum or subcutaneous emphysema | Progression | References |
|------------|----------------|----------------|------------------|---------|--------|-----------------------------------------------|-------------|-----------|
| 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 | [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: 3,090 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 pneumonopathy, 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 pneumonopathy | 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, hipgirdle and thighs | • Weak neck muscles | CK: 173 U/L, LDH: 1995 U/L, AST: 554 U/L | Extractable nuclear antigens: -ve | HRCT Chest: pneumomediastinum, HRCT Chest: pneumomediastinum, Mild fiber atrophy of non-specific interstitial pneumonia (NSIP) with ground glass opacity; features of bronchiolitis obliterans with organizing pneumonia | Muscle biopsy: Steroid | Initial diagnosis → Steroid treatment → Worsening symptoms & subcutaneous emphysema → Deflazacort and MMF management → Symptomatic improvement on follow-up | [31] |
| Age/Gender | Symptoms/Signs | Investigations | Autoimmune Panel | Imaging | Biopsy | Initial management prior to pneumothorax, pneumomediastinum or subcutaneous emphysema | Progression | References |
|------------|----------------|---------------|------------------|---------|--------|-----------------------------------------------|-------------|-----------|
| 34/M       | MACULAR ERYTHEMA AT NECK AND ARM EXTENSORS | SOB WITH BILATERAL FINE CRACKLES | BIOSPHER: N, ESR 70 MM/H, CRP 32.3 MG/DL, CK: NORMAL, AKDLASE: NORMAL | ANA, ANCA, RF, AND OTHER AUTOIMMUNE 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 | SULTASALAZINE FOR ARTHRITIS |
| 36/M       | UNKNOWN | UNKNOWN | INTERSTITIAL LUNG DISEASE | UNKNOWN | UNKNOWN | ANA, ANTI-SS-A, ANTI-SS-B, ANTI-SM, ANTI-RNP, ANTI-SC170, ANTI-JO-1 | CXR: PNEUMOTHORAX, PNEUMOMEDIASTINUM AND SUBCUTANEOUS EMPHYSEMA | STEROID, HYDROXYCHLOROQUINE |
| 38/F       | HELIOtrope rash, GOTTORn’s sign, PERIUNGUAL erytheMA | MUSCLE WEAKNESS | CK: 2379 U/L, ANTI-MDA5: +VE, ANTI-ARS, ANTI-SRP, ANTI-Mi2, ANTI-SAE, ANTI-NXP2: -VE | Unknown | Unknown | CT CHEST: PNEUMOMEDIASTINUM AND CERVICAL SUBCUTANEOUS EMPHYSEMA | STEROID, HYDROXYCHLOROQUINE |
| 38/M       | PERIUNGUAL erytheMA on fingers | PROXIMAL MUSCLE WEAKNESS | ESR: 27 MM/H, CRP: NORMAL, CK: 2379 U/L, LDH: 311 IU/L, AKDLASE: 6.6 IU/L, ALT: 86 IU/L | ANA, dsSNA, ANTI- Jo-1, ANCA, RF: -VE | CT CHEST: PNEUMOMEDIASTINUM AND CERVICAL SUBCUTANEOUS EMPHYSEMA | MUSCLE BIOPSY: SLEIGHT 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, HYDROXYCHLOROQUINE MANAGEMENT | ACUTE CHEST PAIN AND SOB → ADDITION OF CY → SYMPTOM RESOLUTION & RECURRENCE FREE AT 7 MONTHS FOLLOW-UP |
| 39/F       | UNKNOWN | UNKNOWN | INTERSTITIAL LUNG DISEASE | UNKNOWN | UNKNOWN | ANA, ANCA, RF, AND OTHER AUTOIMMUNE 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 | SULTASALAZINE FOR ARTHRITIS |

References:

[1] Initial presentation with respiratory symptoms → ICU support for severe hypoxic respiratory failure → Steroid + CYC management → Discharged on MMF and Steroids → Re-presentation for refractory severe hypoxic respiratory failure 1 month later → Given immunosuppressive therapy and mechanical ventilation → Deceased

[10] Initial presentation with respiratory symptoms → ICU support for severe hypoxic respiratory failure → Steroid + CYC management → Discharged on MMF and Steroids → Re-presentation for refractory severe hypoxic respiratory failure 1 month later → Given immunosuppressive therapy and mechanical ventilation → Deceased

[12] Initial presentation with respiratory symptoms → ICU support for severe hypoxic respiratory failure → Steroid + CYC management → Discharged on MMF and Steroids → Re-presentation for refractory severe hypoxic respiratory failure 1 month later → Given immunosuppressive therapy and mechanical ventilation → Deceased

[29] Initial presentation with respiratory symptoms → ICU support for severe hypoxic respiratory failure → Steroid + CYC management → Discharged on MMF and Steroids → Re-presentation for refractory severe hypoxic respiratory failure 1 month later → Given immunosuppressive therapy and mechanical ventilation → Deceased
| Age/Gender | Symptom/Signs | Dermatological | Muscular | Respiratory | Autoimmune panel | Imaging | Biopsy | Initial management prior to pneumothorax, pneumomediastinum or subcutaneous emphysema | Progression | References |
|------------|--------------|----------------|----------|-------------|------------------|--------|--------|---------------------------------------------|------------|-----------|
| 39/F       | Gottron's signs  | Proximal upper and lower limbs weakness | SOB, Interstitial pneumonopathy, 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 | 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 | 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]         |
| Age/Gender | Dermatological Symptoms/Signs | Muscular Symptoms/Signs | Respiratory Symptoms/Signs | Bloods | Autoimmune panel | Imaging | Biopsy | Initial management prior to pneumothorax, pneumomediastinum or subcutaneous emphysema | Progression | References |
|------------|-----------------------------|------------------------|-----------------------------|--------|-------------------|---------|--------|------------------------------------------------|-------------|-----------|
| 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 | RF, Anti-platelet antibodies, cryoglobulin, 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, Hydroxychloroquine | [26] 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 |
| 42/M       | • Heliotrope rash, Gottron’s papules | • Lower limbs weakness | • Asymptomatic subcutaneous emphysema | CK: 2260 IU | Unknown | CXR: Increased interstitial markings | Unknown | Steroid, AZA | [5] Initial diagnosis → Steroid management → Readmission due to muscle weakness → IV steroid management → Development of bilateral aspiration pneumonia → Steroid and AZA management → Development of a lung 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 |
| 42/M       | • Chest or back erythema, Gottron’s sign, Periungual erythema | • 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: 3457 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 |
|------------|----------------|----------------|----------------------------------------------------------------------------------------|-------------|------------|
| 44/M       | Unknown        | Interstitial lung disease | CT: Normal, LDH: Normal | Unknown | Unknown | Steroid, AZA | [10] |
| 45/F       | Vasculitis-like skin lesions on the dorsum of both hands | Increased interstitial lung infiltrates in both lower lung fields | 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 | [11] |
| 45/M       | Heliotrope rash | No weakness | CK: 120 U/L | Anti-Mi2 and Anti-MDA-5: −ve | Unknown | Complete clinical response | [7] |
| 46/F       | Skin vasculopathy | Head and neck were swollen and subcutaneous emphysema and crepitance observed | CK: 1280 U/L | CT Scan: Pneumomediastinum & Subcutaneous emphysema from head to upper arm | Unknown | Steroid, CY | [27] |
| 46/M       | Heliotrope rash | Mild proximal muscle weakness | CK: 170 U/L | Anti-Mi2 and Anti-MDA-5: −ve | Unknown | Steroid, AZA | [4] |
| 46/M       | Gottron’s papules | Proximal upper and lower limb weakness | CK: 170 U/L | Anti-Mi2 and Anti-MDA-5: −ve | Unknown | Complete clinical response | [7] |
| 50/M       | Facial erythema, scaly papules on the fingers, heliotrope rash, shawl sign, Gottron papules | Weakness of flexors of neck, shoulders and hips | CK: 309 IU/L (elevated), LDH: 228 IU/L (elevated), Aldolase: Myoglobin 89.0 ng/mL, Transaminase elevated | 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, AZA | [9] |
| Age/Gender | Symptoms/Signs | Investigations | Autoimmune panel | Imaging | Biopsy | Initial management prior to pneumothorax, pneumomediastinum or subcutaneous emphysema | Progression | References |
|------------|----------------|----------------|------------------|---------|--------|-------------------------------------------------|-------------|------------|
| 51/F       | Gottron’s sign  | • Pneumothorax  | Anti-MDA5: +ve,  | Unknown | Unknown| Deceased                                       |             | [12]       |
|            | Skin ulcer      | and Subcutaneous| Anti-ARS, Anti-SRP, Anti-TIFγ, Anti-Mi2, Anti-SAE, Anti-NXP2: -ve | Unknown | Unknown|                                                |             |            |
|            | Muscle weakness | emphysema       |                   |         | Unknown|                                                |             |            |
| 52/F       | Confluent and   | • SOB on        | 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 Hydroxychloroquine and Clobetasol → SOB, facial and neck edema → Bilateral chest tubes and CY → Discharged with significant improvement | [8]         |
|            | violaceous      | representation  |                   |         |        |                                                |             |            |
|            | erythema on     | CK: Normal,     |                   |         |        |                                                |             |            |
|            | upper chest     | Aldolase: Normal,|                   |         |        |                                                |             |            |
|            | • Nil           |                   |                   |         |        |                                                |             |            |
| 54/F       | Heliotrope rash | • Sudden onset  | Anti-Ro: 52       | 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]        |
|            | Erythematous     | of SOB           | (positive),       |                   |        |                                                |             |            |
|            | rashes over     |                   | Anti-smRNP, SS-A/B, Jo-1, ANA: -ve |                   |         |                                                |             |            |
|            | anterior chest   |                   |                   |         |        |                                                |             |            |
|            | wall            |                   |                   |         |        |                                                |             |            |
| 56/M       | Heliotrope rash,| • No muscle      | Anti-MDA5: +ve,  | Unknown | Unknown| Deceased                                       |             | [12]       |
|            | Gottron’s sign,  | weakness         | Anti-ARS, Anti-SRP, Anti-TIFγ, Anti-Mi2, Anti-SAE, Anti-NXP2: -ve | Unknown | Unknown|                                                |             |            |
|            | Skin ulcer      |                  |                   |         |        |                                                |             |            |
| 57/F       | Heliotrope rash | • Proximal upper| Anti-Mi2 and      | Unknown | Unknown| Steroid                                         | Deceased    | [7]        |
|            | Gottron’s signs | and lower limbs | Anti-MDA5: -ve    | Unknown | Unknown|                                                |             |            |
|            | Shallow sign     | weakness         |                   |         |        |                                                |             |            |
|            | Periungual       |                  |                   |         |        |                                                |             |            |
|            | hypertrophy      |                  |                   |         |        |                                                |             |            |
| 57/M       | Skin ulcers     | • Muscle         | CK: 41 U/L        | Unknown | Unknown| Steroid, CYC                                    | Initial diagnosis → Steroid and CYC management → Subcutaneous emphysema → Addition of Rituximab → Symptomatic resolution | [37]        |
|            | Cutaneous        | weakness         |                   |         |        |                                                |             |            |
|            | erythema         |                  |                   |         |        |                                                |             |            |
|            | Facial and neck  |                  |                   |         |        |                                                |             |            |
|            | swelling         |                  |                   |         |        |                                                |             |            |
| Age/Gender | Dermatological | Muscular | Respiratory | Symptoms/Signs | Investigations | Autoimmune panel | Imaging | Biopsy | Initial management prior to pneumothorax, pneumomediastinum or subcutaneous emphysema | Progression | References |
|------------|----------------|----------|-------------|----------------|----------------|-------------------|---------|--------|-----------------------------------------------|------------|-----------|
| 58/F       | Unknown        | Unknown  | Unknown     | Neck swelling, Dysphagia | Unknown | Unknown | CXR & CT | Chest: Extensive pneumomediastinum and subcutaneous emphysema. Bibasilar ground-glass and reticular opacities. | CYC | [34] |
| 59/F       | Unknown        | Unknown  | Unknown     | Interstitial pneumonitis | CK: 3501 IU/L | Unknown | Unknown | Unknown | Steroid | Initial diagnosis → Steroid management | Alive | [30] |
| 59/M       | Unknown        | Unknown  | Unknown     | Interstitial lung disease | Unknown | Unknown | Unknown | Unknown | Steroid, CYC, AZA, IVIG | Initial diagnosis → Steroid, CYC, AZA and IVIG management | Intestinal lung disease progression → Rituximab management → Improved lung function | [32] |
| 60/M       | Gottron’s sign | No muscle weakness | Unknown | CK: 25 U/L | Anti-MDA5: +ve, Anti-ARS, Anti-SRF, 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 | Anti-Jo, ANA, ANCA, Myositis, MDA-5, RF: -ve | 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 | Unknown | CXR & CT scan: Severe pneumomediastinum and ILD, mainly in the lower lobes | 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] |
| Age/Gender | Symptoms/Signs | Initial management prior to pneumothorax, pneumomediastinum or subcutaneous emphysema | Progression | References |
|------------|---------------|-------------------------------------------------------------------------------------|-------------|------------|
| 74/M       | Chest or back erythema, Gottron’s sign, Periungual erythema | | Deceased | [12] |
|            | 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 |
| 7/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-aminocyl 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 size and site 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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