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Pre-Capillary Pulmonary Hypertension in a Patient With Idiopathic Inflammatory Myopathy Without Extensive Pulmonary Involvement

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

ABDEF Steve O. Nwokeocha
BDE Liana Abduova
DEF Shalini Thiruvarudchelvam
DEF Ashlesha Chaudhary
EF Raghini Ranganathan
ADEF Bailey O'Donnell

Department of Internal Medicine, Bassett Medical Center, Cooperstown, NY, USA

Corresponding Author: Steve O. Nwokeocha, Bassett Medical Center, 1 Atwell Road, Cooperstown, NY, USA, 13326, Phone: +1-607-282-2114, e-mail: Steve.Nwokeocha@bassett.org

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Patient: Female, 71-year-old
Final Diagnosis: Dermatomyositis • heart failure • pulmonary hypertension
Symptoms: Dysphagia • fatigue • leg swelling • proximal muscle weakness • shortness of breath
Clinical Procedure: Cardiac catheterization • CT chest • echocardiography • laboratory investigations • MRI • right and left heart catheterization
Specialty: Critical Care Medicine • Pulmonology • Rheumatology

Objective: Rare coexistence of disease or pathology

Background: Dermatomyositis is an idiopathic inflammatory myopathy (IIM) that presents commonly with cutaneous, myopathic, and systemic manifestations. Cardiopulmonary involvement is usually paired with interstitial lung disease (ILD), which when severe can lead to pulmonary hypertension (PH) with heart failure, contributing to morbidity, poor quality of life, and mortality. However, dermatomyositis is rarely associated with PH in the absence of extensive ILD, and is infrequently considered in patients who present PH in the absence of significant lung findings.

Case Report: We present the case of a 71-year-old woman who was admitted with signs and symptoms of heart failure after months of progressive dysphagia, proximal muscle weakness, and shortness of breath. She was subsequently diagnosed with dermatomyositis, as well as pre-capillary PH, through right-heart catheterization, but had no features of extensive ILD on imaging. She was treated with intravenous diuretics, intravenous methylprednisolone 1 g daily for 3 days, followed by oral prednisone 60 mg, and 2 doses of 75 g of intravenous immunoglobulins (IVIG), resulting in overall clinical improvement. Further IVIG treatment and re-evaluation of the need for vasodilator therapy was planned, but unfortunately the patient died following admission with hyperosmolar hyperglycemia and aspiration pneumonia after declining resuscitation and intubation.

Conclusions: PH in the absence of extensive ILD in dermatomyositis may be under-recognized. IIM evaluation should be considered in the rheumatological work-up of pre-capillary and combined pre-/post-capillary PH within the appropriate clinical context. Gaps exist in the understanding of the pathophysiology of PH in IIMs and its management, prognosis, and outcomes.

Keywords: Dermatomyositis • Heart Failure • Pulmonary Arterial Hypertension • Pulmonary Heart Disease • Pulmonologists • Rheumatology

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Introduction

Dermatomyositis is an idiopathic, inflammatory myopathy (IIM) that is often associated with cutaneous, myopathic, and systemic manifestations, presenting classically with proximal muscle weakness and cutaneous rash, but capable of extending to cardiovascular, gastrointestinal, and pulmonary systems. Pulmonary involvement often includes hypoventilation, aspiration pneumonia, and interstitial lung disease (ILD) [1]. However, a rare complication of dermatomyositis can involve development of pulmonary hypertension (PH) in the absence of, or with minimal, concurrent ILD [2]. Although there are case studies illustrating this rare association, the mechanism is not clearly defined. However, there are emerging studies which show myositis-specific/myositis-associated antibodies (MSAs/MAAs) that are contributory to the development of ILD and IIMs and that may also have an association with the development of PH [2].

Pulmonary arterial hypertension (PAH) is a pulmonary vascular disease associated with significant vascular remodeling, and evidence suggests inflammatory cytokines and immune-mediating and modulatory cells may be contributory to the development of PH in the setting of dermatomyositis [3], including high prevalence of MSA/MAA [2], and elevated IL-6 and IL-10 [3]. Herein, we present this case report which details an unusual manifestation of an IIM that led to an initial presentation with heart failure, PH, and subsequent evaluation leading to dermatomyositis as the underlying cause of the patient's clinical presentation and progression.

Case Report

We present the case of a 71-year-old White female with a past medical history significant for hypertension, type 2 diabetes mellitus, Raynaud's syndrome, hypothyroidism with Hashimoto thyroiditis, and recent diagnosis of oropharyngeal dysphagia. She was admitted to the hospital due to a 2- to 3-week history of progressively worsening bilateral lower extremity swelling. She had shortness of breath on exertion. She denied orthopnea, paroxysmal nocturnal dyspnea, chest pain, palpitations, or cough. She was started on furosemide tablets by her primary care physician but only took 1 dose and discontinued.

On further questioning, she mentioned concerns of generalized fatigue that had been worsening over a period of about 6 months. This started alongside progressively worsening bilateral upper and lower limb proximal muscle weakness, with weakness of the muscles of the neck described as being unable to hold her head up, as well as voice changes. Shortly after this, she developed progressive oropharyngeal dysphagia. This was managed with frequent visits to the speech and language therapist due to reduced oral intake that led to a loss of 14-17 kg in weight over 6 months. She had been seen by the otolaryngologist, and subsequently the gastroenterologist, where an upper endoscopy was offered, but she declined. She stated marked functional decline due to significantly reduced mobility and oral intake, which reduced her quality of life. She had no family history of autoimmune diseases. She never smoked or drank alcohol. She was not up to date with cancer screenings and vaccinations.

A rheumatologic review of systems was positive for infrequent dry mouth, infrequent dry eyes, hair thinning, muscle weakness, and Raynaud's phenomenon. She also had chronic dry skin. The review was negative for recurrent aphthous ulcers, photosensitive rash, inflammatory eye disease, serositis, arthralgias, venous thromboembolism, cerebrovascular accident (CVA), and seizures.

On physical examination, she was breathing on room air and her vital signs were normal. Her weight was 74.8 kg and her body mass index was 30 kg/meter^2. She appeared well without signs of distress. She had hypophonia. There was no palpable cervical or axillary lymphadenopathy. Cardiovascular exam showed normal rate, regular rhythm with S1 and S2 present. Her respiratory exam revealed mild fine inspiratory crackles at the bases of both lungs. Her abdomen was soft and non-distended. There was 2+ pitting edema in lower extremities up to the thigh. Skin exam showed no psoriasiform, malar, or lupus-like rash, or palpable purpura. She had faint reddish/purplish macular areas over both elbows and the extensor surfaces of the bilateral metacarpophalangeal (MCP) and proximal interphalangeal (PIP) joints, consistent with Gottron's sign (**Figure 1**). There were suggestions of possible mechanic hands bilaterally with hyperkeratotic, scaly, and fissured skin on the tips and sides of the fingers and thumbs (**Figure 2**). There were also scattered clear fluid-filled blisters on the shin and foot, some ruptured and crusted. She had no obvious telangiectasias or nailfold capillary abnormalities, and no scleroderma skin thickening or calcinosis. No heliotrope rash or holster sign. On neurological exam, she was alert and oriented. Power with any movement at the shoulder or hip joint was 3-/5. She had good hand grip and pedal strength bilaterally. Sensation was intact. Cranial nerves II – XII were intact.

On day 1, her laboratory work-up for complete blood count, kidney function, electrolytes, hepatic panel, and thyroid function were unremarkable. High-sensitivity troponin was mildly elevated at 33.4 pg/mL; repeat was 36.3 pg/mL. BNP was mildly elevated at 126 pg/mL. Portable chest X-ray demonstrated cardiomegaly, peribronchial cuffing, and early septal thickening. Electrocardiogram (EKG) showed sinus rhythm with rSr' in V1 without ST-segment or T-wave changes. A transthoracic echocardiogram showed normal left ventricular ejection fraction

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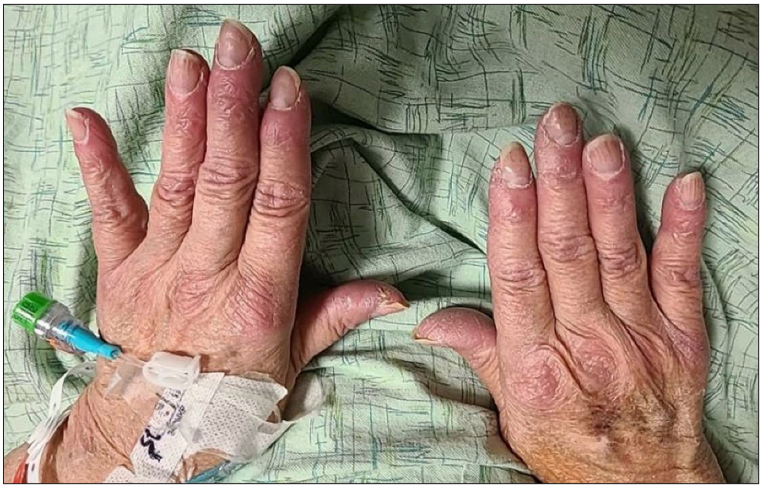

Figure 1. Faint Gottron's sign over the dorsal aspect of both hands.

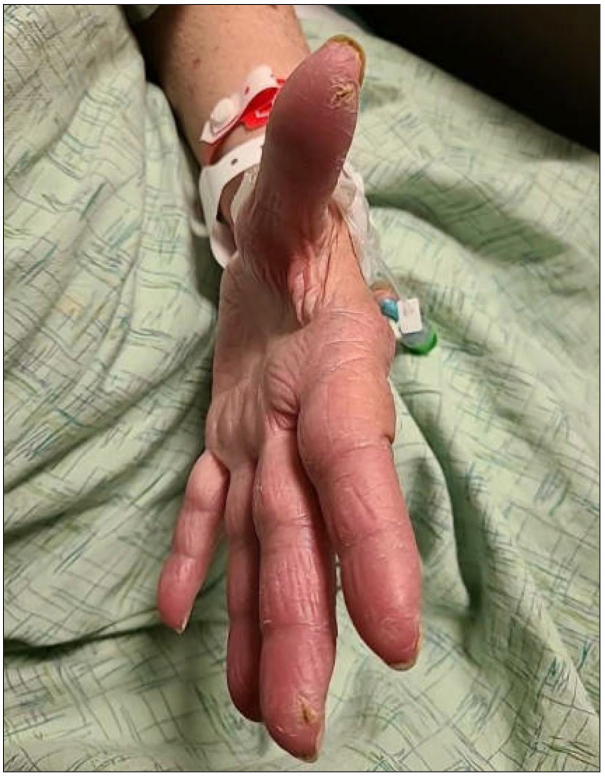

Figure 2. Thickened and flaky skin suggestive of possible mechanic hands.

greater than 55% and normal left ventricle wall thickness. The interventricular septum was flattened, consistent with elevated right ventricle (RV) pressure/volume overload, moderate RV dilatation with normal RV wall thickness, and estimated systolic pulmonary pressure of 80 mmHg, indicating PH.

Her presentation was concerning for severe PH with right-heart failure alongside an underlying neuromuscular and autoimmune process. She was started on boluses of intravenous (IV) furosemide 40 mg daily which she received for the first 3 days. Comprehensive neuromuscular and auto-immune work-up was sent with the subsequent results as listed in **Table 1**, most of which were obtained between day 1 and day 7 of admission, with the myositis panel results obtained after 2 weeks (**Table 1**).

Prior to the availability of the results of the extensive testing, neurology was consulted, who at the time suspected underlying myopathy. However, they opted to wait for the results of creatine phosphokinase (CPK), aldolase, acetylcholine receptor antibody, erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP). Cardiology and pulmonology were consulted for evaluation of her severe PH with right-heart failure. A pulmonary function test could not be appropriately done due to poor ability to put in effort for the study secondary to weakness. A computed tomography angiography with pulmonary embolism protocol showed no acute venous thromboembolism, with shallow inspiratory effort and mild bilateral basal interstitial changes which were felt to be secondary to early developing ILD. A ventilation/perfusion (V/Q) scan showed no evidence of chronic thromboembolic pulmonary hypertension. Given findings of severe PH in the absence of overt left-sided heart disease, or overt significant chronic lung disease, both cardiology and pulmonology recommended right-heart catheterization.

On day 8 of admission, with the combination of the elevated CPK, aldolase, anti-dsDNA, extractable nuclear antigen antibodies, and antinuclear antibodies (ANA) results (**Table 1**), there was a concern for possible dermatomyositis/polymyositis, though it was felt that this diagnosis would not explain the presence of PH. A consultation with rheumatology was placed. Pulmonology reviewed further differential diagnoses of precapillary PH including idiopathic PAH, connective tissue disease-associated PAH, chronic thromboembolic disease, porto-PH, human immunodeficiency virus (HIV)-associated PAH, drug-induced PAH, and rare pulmonary veno-occlusive disease; all

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Table 1. Neuromuscular and auto-immune work-up panel.

Laboratory investigation	Laboratory value	Reference range
Creatine phosphokinase (CPK)	1293 U/L	67-95 U/L
Aldolase	53.1 U/L	<8.1 U/L
Erythrocyte sedimentation rate (ESR)	42 mm/h	<30 mm/h
C-reactive protein (CRP)	1.6 mg/dL	0.1-1.0 mg/dL
Acetylcholine receptor antibody	Negative	Negative
Antinuclear antibodies (ANA)	Positive	Negative
Anti-double stranded DNA (dsDNA)	16 IU/mL	<15 IU/mL
Extractable Nuclear Antigen antibodies (ENA) screen	6.4	<1.0
SSA-A/Ro antibodies	178 U/mL	<7 U/mL
RNP70 IgG	1.5 U/mL	<7 U/mL
Smith antibodies	<0.7 U/mL	<7 U/mL
U1RNP IgG	2.1 U/mL	<5 U/mL
Centromere antibody IgG	0.6 U/mL	<7 U/mL
SS-B/La antibodies IgG	1.7 U/mL	<7 U/mL
Anti-PM/Scl-100 antibody	>200 units	<20 units
Anti-SS-A 52kD antibody	175 units	<20 units
Anti-Scl-70 antibodies	0.9 U/mL	<7 U/mL
Anti-Jo-1 antibody	<20	<20
Anti-PL-7 antibody	Negative	Negative
Anti-PL-12 antibody	Negative	Negative
Anti-EJ antibody	Negative	Negative
Anti-OJ antibody	Negative	Negative
Anti-SRP antibody	Negative	Negative
Anti-Mi-2 antibody	Negative	Negative
Anti-TIF-1-gamma antibody	<20	<20
Anti-mda-5 antibody	<20	<20
Anti-NXP-2 (P140) antibody	<20	<20
Anti-SAE1 antibody, IgG	<20	<20
Anti-Ku antibody	Negative	Negative
Anti-U1 ribonucleoprotein antibody	<20	<20
Anti-U2 ribonucleoprotein antibody	Negative	Negative
Anti-U3 ribonucleoprotein antibody (Fibrillarin)	<20	<20

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of which were not very likely in this patient based on her in-depth history, examination, and work-up thus far. HIV testing was negative. An arterial blood gas test (ABG) demonstrated pH 7.45, $paCO_2$ 46 mmHg, paO_2 59 mmHg, HCO_3^- 32 mmol/L, and base excess of +7 mmol/L. Due to hypoxemia on the ABG, she was placed on 2 L of oxygen with a nasal cannula.

Rheumatology's assessment noted that the faint macular regions of the elbow, MCP,

and PIP were consistent with Gottron's sign, in addition to possible mechanic's hands, history of Raynaud's, and proximal muscle weakness. Alongside the positive ANA and SS-A antibodies (**Table 1**) her examination findings were considered most consistent with dermatomyositis. Based on the European League Against Rheumatism and the American College of Rheumatology (EULAR/ACR) scoring for adult and juvenile IIMs, she had a score of 11.7 (without muscle biopsy, age >40 years, proximal upper and lower extremity weakness, weak neck flexors, proximal lower extremity muscles weaker than distal muscles, Gottron's sign, dysphagia, and elevated CPK). This score placed her within the definite IIM diagnostic category, and indicated dermatomyositis within the classification tree for subgrouping. The myositis panel was still pending at this time. Rheumatology also stated that PH was considered to be potentially associated with dermatomyositis, though with the understanding that both are rarely reported in the literature without extensive ILD as in her case.

Magnetic resonance imaging (MRI) of the left thigh was done to evaluate for signs of myositis, which in addition to the available information would prevent the need for a muscle biopsy. The left thigh was selected as a representative proximal muscle group for evaluation as opposed to scanning all limbs. The MRI showed soft-tissue edema and changes consistent with inflammatory myositis without abscess or mass. Given the severity of her disease, IV methylprednisolone 1 g daily was started on day 8 and continued for 3 days, followed by oral prednisone 60 mg daily, with plans for intravenous immunoglobulins (IVIG) or rituximab after screening microbiological work-up. Due to an association of dermatomyositis with cancer, cancer screening was recommended to be pursued on discharge. Commencement of steroids was met with noticeable gradual improvements in weakness and shortness of breath by the patient, with continued dysphagia. Diuresis recommenced on day 10 of admission.

On day 12 she underwent left and right-heart catheterization, which demonstrated widely patent coronary arteries, with hemodynamic indices as follows: RV 89/- mmHg, right atrium 12/- mmHg, pulmonary artery pressure (PAP) 91/30 mmHg, mean PAP (mPAP) 54 mmHg, pulmonary vascular resistance (PVR) 6.6 Wood units (WU), pulmonary arterial wedge pressure (PAWP) 13 mmHg, cardiac output 6.14 L/min, cardiac index 3.61 L/min/m^2. Based on recent thresholds set for the definitions of PH, she had pre-capillary PH; it was felt that the degree of PH based on the mPAP was unlikely to be driven by left heart disease, and there were no signs of significant lung disease. Cardiology strongly recommended transfer to a dedicated PH center. Eventually, she agreed to be transferred to a tertiary care center and was transported on day 16.

At the PH center she received 2 doses of 75 g of IVIG on consecutive days alongside continued diuresis, resulting in clinical improvement in strength, breathing, and edema. Dysphagia persisted with continued aspiration noted on modified barium swallow. CPK came down to 347 U/L (reference range: 26-192 U/L) and inflammatory markers improved. Given normal cardiac output and cardiac index, PAH-targeted vasodilator therapy was deferred with plans for repeat right-heart catheterization and vasodilatory testing when she reached euvolemia with diuresis. She was discharged with plans for close follow-up.

At a 1-month rheumatology clinic follow-up visit, she was seen to be lethargic, providing some history with her husband supplementing. She reported initial improvement in strength and function after IVIG, including the ability to climb a few stairs which she was unable to do for a long time prior to the admission. However, she subsequently declined in strength and energy over the week preceding the next clinic visit. She no longer required supplemental oxygen, and her peripheral edema was markedly improved, but she was profoundly weak in the proximal muscles, requiring assistance for transfers and wheelchair mobility. CPK levels had improved to 146 U/L (ref 67-95 U/L). CRP was 0.5 mg/dL and ESR was 55 mm/h. Plans were made for further IVIG treatment at 50 g once every 4 weeks for 6 months.

Two days later, she was admitted in a hyperosmolar hyperglycemic state, with severe hypernatremia, acute kidney injury, and aspiration pneumonia. She initially improved almost to baseline, and subsequently agreed to a percutaneous feeding tube due to continued dysphagia. She was close to being discharged when she developed severe acute hypoxic respiratory failure with escalating oxygen requirements, assessed to be secondary to aspiration. She subsequently decided against resuscitation and mechanical or non-invasive ventilation, and eventually died. The family declined an autopsy.

Discussion

Dermatomyositis is a form of IIM with skin involvement that has a highly variable incidence estimated to be about 0.2 to 2 per 100 000 person-years, and an estimated prevalence of 13 per 100 000, highest in the United States [1,4,5,6]. The

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prevalence of PH with dermatomyositis is even less so, often occurring in the presence of severe, extensive ILD, and very rarely as PAH in the absence of severe ILD [2]. In a French PH registry of over 5000 patients, 34 had IIMs, out of which only 3 had evidence of PAH without extensive ILD; all 3 patients had dermatomyositis [7].

Autoimmune diseases cause PH through various mechanisms. PAH has a well-recognized association with conditions like systemic sclerosis and mixed connective tissue disease, with pathophysiology hypothesized to be multifactorial, including interactions between the immune system (through cells, cytokines, and interleukins) and vascular cells, causing vascular remodeling within the pulmonary arteries, leading to precapillary PH, all of which may play a similar role in dermatomyositis [3,4,8,9].

IIMs are diagnosed based on the 2017 classification criteria set by EULAR/ACR, where patients with characteristic clinical findings of weakness, rash, and laboratory results such as elevated CPK, are scored based on the presence or absence of a muscle biopsy [4]. Our patient had a score that placed her within the definite category without need for a muscle biopsy, and within the sub-group of dermatomyositis [4]. In IIMs, PH is not a readily recognized or appreciated phenomenon. It is not considered within the 2017 EULAR/ACR classification criteria for IIMs [4]. The learning point from our case, however, is that it may be under-recognized, especially in dermatomyositis, where its pathogenesis is postulated to primarily involve endothelial and microvascular injury [11,12].

In our case, significant left-sided heart disease was not supported by the wedge pressure. Precapillary or combined pre- and post-capillary PH can be defined by PVR greater than or equal to 3 WUs, with mPAP above 20 mmHg and PAWP below or equal to 15 mmHg if pre-capillary PH; or PAWP above 15 mmHg if combined pre-and-post-capillary PH [13]. Her right-heart catheterization was suggestive of pre-capillary PH. Chronic thromboembolic disease was excluded by negative V/Q imaging, and severe parenchymal lung disease was not evident on chest imaging. HIV testing was negative, and there was no history suggestive of portal hypertension or a causative drug exposure. Sjögren syndrome was considered, but her symptoms of dry eyes and dry mouth were not persistent enough to be considered as concerns for Sjögren syndrome by our rheumatologist. Upon testing for Sjögren's-syndrome-related antigen A (SSA) antibodies, she had positive anti-SSA/Ro antibodies, but they can also be seen in dermatomyositis [14].

Tobal et al performed a retrospective cohort study in patients with either pre-capillary or combined pre- and post-capillary PH without prior diagnosis of auto-immune systemic diseases, and compared them to matched cohorts of patients with suspected IIM without PH [2]. It showed statistically significantly increased prevalence of myositis-specific antibodies (MSAs) and myositis-associated antibodies (MAAs) in the PH cohort who had radiological signs of ILD, and an increased prevalence in weakly positive MSAs/MAAs in patients without radiologic evidence of ILD [2]. There was statistically significantly increased prevalence of weakly positive dermatomyositis-associated MSAs in the PH cohort compared with the matched IIM/ILD cohort [2].

Tobal et al's study and our case add to a growing body of evidence suggesting that, in addition to the already recognized rheumatological etiologies of PH, perhaps IIMs and MSAs/MAAs should be actively considered as part of work-up in patients presenting with PH, especially with PVR above 3 WUs [2,7,12,15-17]. In our case, hypoxemia, noted on her ABG in the absence of significant ILD on imaging, perhaps played a role. It is, however, unclear if a mild degree of oxygen requirement would have such an impact on her mPAP without other co-existing pre-capillary etiologies.

Current management of dermatomyositis is centered on rapid control of the inflammatory myopathy, commonly done with high-dose corticosteroids [1,18]. In severe or refractory dermatomyositis, intravenous immunoglobulin and rituximab can be used [1,18]. There are no clear guidelines for the management of patients with isolated PH, but vasoactive therapy has been used alongside immunosuppressive agents used in dermatomyositis treatment [15].

Our case also highlights that gaps exist within the literature on PH in IIMs that should be readily explored. For instance, people with PH and weakly positive MSAs/MAAs without obvious symptoms of IIMs may indicate an early disease process prior to the development of symptomatic IIM with ILD [2]. A study on the disease course in these kinds of patients, and on the impact of early treatment with immunological therapies on the development or progression of their IIMs and PH, would be useful for practice. Additional research gaps include the pathophysiological process of PH in these patients, particularly MSAs or MAAs associated with increased incidence or risk of PH with or without extensive ILD. In addition, the appropriate management approach for patients with symptomatic IIMs and PH without significant ILD needs to be established.

Conclusions

Pre-capillary PH in the absence of significant lung disease in patients with dermatomyositis and other IIMs is a potentially under-recognized association. In patients with unexplained pre-capillary PH and potential features of dermatomyositis or other IIMs, clinicians should consider formal IIM evaluation to establish an etiology. There are a multitude of research gaps in this

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association to be further explored, including the underlying pathophysiology, optimal management, disease course, and outcomes.

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Department and Institution Where Work Was Done

Internal Medicine Department, at Basset Medical Center, Cooperstown, NY, USA.

Patient Consent

Consent was obtained from the patient.

Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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