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Interstitial Lung Disease in Dermatomyositis and Polymyositis in Patients from Saudi Arabia: A Single Tertiary Centre Retrospective Study

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Abstract **Background:** Interstitial lung disease (ILD) is a major extra-muscular manifestation of dermatomyositis (DM) and polymyositis (PM), yet data from Middle Eastern populations remain sparse. We aimed to characterize radiologic ILD patterns, severity, clinical-serologic correlates, and treatment usage among Saudi patients with PM/DM-ILD managed at a tertiary centre. **Methods:** We conducted a retrospective cohort study (2006-2022) at a single tertiary hospital in Riyadh, Saudi Arabia. Adult patients (≥18 years) with PM/DM by 2017 ACR/EULAR classification were re-adjudicated, and ILD was confirmed by high-resolution CT (HRCT) after blinded re-review by a thoracic radiologist. ILD patterns followed ATS/ERS categories (NSIP, UIP, OP, RB-ILD, unclassifiable). Severity was treated separately from pattern and operationalized as "mild" if HRCT parenchymal involvement was <20% or, when imaging was equivocal, if FVC ≥80% predicted. Demographics, clinical features, standard serologies, pulmonary function tests (PFTs), echocardiography, and treatments were abstracted using a standardized form with double-entry checks. Analyses were descriptive and exploratory; between-group comparisons were limited to subgroups with n≥5 and adjusted for multiple testing (Holm). **Results:** Thirty-five patients met criteria (80% female; DM 60%, PM 40%). On HRCT rereview, NSIP was the most frequent pattern (37.1%, 13/35), followed by unclassifiable (14.3%, 5/35), UIP (5.7%, 2/35), RB-ILD (2.9%, 1/35), and OP (2.9%, 1/35). In parallel, severity was mild in 37.1% (13/35) across patterns. ILD timing relative to myositis was PM/DM-preceding in 48.6%, concomitant in 31.4%, and ILD-preceding in 20%. Exertional dyspnoea (74.3%) and non-productive cough (62.9%) were frequent; heliotrope rash and Gottron papules each occurred in 34.3%. ANA was positive in 82.9%; anti-Jo-1 in 34.3%. FVC (mean±SD) approximated 71% predicted overall; DLCO ~90% predicted where available. As pre-specified, no inferential tests were performed for subgroups with n<5 (UIP, RB-ILD, OP), and signals from small cells were treated descriptively only. Glucocorticoids were universally used; 77.1% were maintained at ≤7.5 mg/day. Mycophenolate mofetil (54.3%) and rituximab (48.6%) were the most common steroid-sparing agents; azathioprine (40%), methotrexate (34.3%), and IVIG (25.7%) were also used; antifibrotics were prescribed in 11.4%. **Conclusion:** In this Saudi tertiary-care cohort, NSIP predominated as the ILD pattern, while mild severity at presentation was common but distinct from pattern. The dataset underscores heterogeneity in serology and treatment and highlights the need for standardized radiologic severity grading, prospective PFT capture, and multicentre registries to refine prognosis and guide therapy in regional PM/DM-ILD.

Key Words Myositis, Interstitial Lung Disease, High-Resolution Computed Tomography (HRCT), Pulmonary Function Tests, Autoimmune Lung Disorders

INTRODUCTION

Polymyositis (PM) and dermatomyositis (DM) are the most common subtypes of idiopathic inflammatory myopathy. The autoimmune disorders PM and DM are systemic in nature; they can impact the skin and lungs and are marked by persistent inflammation of the striated muscles [1-2]. DM

is distinguished by a skin condition known as Gottron's rash or heliotrope periorbital rash, which occurs in conjunction with the muscular illness. Adults with PM, on the other hand, acquire proximal muscle weakening over weeks to months without skin lesions [3,4]. Both share several systemic disease manifestations such as arthritis, Raynaud's syndrome, dysphagia caused by pharyngeal and oesophageal involvement, cardiac dysfunction, and different forms of pulmonary disease [5]. Lung involvement tends to be a component of early PM/DM, in which, in approximately 20% of cases, lung disease precedes the onset of muscle or skin disease [3]. Interstitial lung disease (ILD) in PM and DM tends to be the most common form of lung involvement, with a prevalence of 23-41%, with a higher prevalence being reported in people of Asian descent [1-4]. Several recent studies have increased our understanding of this particular disease component [3]. Patients with PM are more often compared to patients with DM, and somehow female individuals are affected more often than are male individuals [5]. The underlying mechanism for ILD and PM/DM remains unknown. Immune mechanisms generally have key roles in the pathogenesis of the inflammation of the lungs in genetically susceptible individuals [1].

Genetic background likely contributes meaningfully to the striking geographical variability observed in both the prevalence and radiologic patterns of ILD among patients with polymyositis and dermatomyositis (PM/DM). Differences in HLA haplotypes, ancestry-related variation in immune-response genes, and population-level distributions of myositis-specific/associated autoantibodies can shape the pulmonary phenotype, while environmental co-exposures (e.g. dust, biomass smoke, viral triggers) and health-system factors (screening intensity, referral pathways) modulate detection and apparent frequency [1]. Against this backdrop, the principal ILD patterns encountered in PM/DM, defined according to ATS/ERS HRCT lexicon, include nonspecific interstitial pneumonia (NSIP), usual interstitial pneumonia (UIP), organizing pneumonia (OP), and diffuse alveolar damage (DAD) [1]. Less commonly reported phenotypes include airway-centred/respiratory bronchiolitis-associated ILD and unclassifiable ILD, which arise when imaging features are mixed or incomplete [3-6]. Of note, pulmonary vasculitis has been described only rarely: in one autopsy series it was observed in 5 of 65 PM/DM cases, a finding that has not been consistently reproduced in other cohorts and therefore should be interpreted with caution as an infrequent or terminal-event phenomenon rather than a routine clinicoradiologic pattern [1].

Clinically, PM/DM-associated ILD spans a wide spectrum, from asymptomatic disease detected on screening HRCT/PFTs to rapidly progressive respiratory failure with hypoxemia and high short-term mortality. Based on presenting tempo, three pragmatic clinical courses are commonly described: (i) acute/rapid-onset disease with precipitous dyspnoea and gas-exchange impairment (often corresponding to OP or DAD on imaging); (ii) chronic, slowly progressive disease more typical of fibrosing NSIP; and (iii) subclinical/asymptomatic disease in which abnormal chest imaging or pulmonary physiology uncovers ILD before symptoms declare themselves [7]. While cough and exertional dyspnoea are the most frequent complaints, the absence of respiratory symptoms does not exclude ILD. Indeed, one cohort reported that ~27% of patients with imaging-confirmed ILD were asymptomatic, whereas a considerable fraction of patients without radiographic ILD endorsed respiratory symptoms attributable to alternative mechanisms (airway disease, deconditioning, anaemia), underscoring the imperfect overlap between symptoms and parenchymal involvement [7]. This dissociation justifies systematic pulmonary screening in PM/DM, particularly at diagnosis and during early follow-up, even when the clinical examination is unrevealing.

Serologically, anti-aminoacyl tRNA synthetase antibodies are the strongest predictors of ILD in myositis, with anti-histidyl tRNA synthetase (anti-Jo-1) being the most frequently detected specificity; across studies, anti-Jo-1 positivity has been associated with ILD prevalences exceeding 70% [8]. Patients with anti-synthetase antibodies commonly manifest the anti-synthetase syndrome, characterized by arthritis/arthralgia, Raynaud phenomenon, fever, and "mechanic's hands," features that can aid early recognition in pulmonary clinics [9]. Other myositis-specific antibodies (e.g. PL-7, PL-12, MDA5, Mi-2, NXP2, TIF1-γ, SAE1) show variable associations with ILD presence, tempo, and pattern; for example, rapidly progressive phenotypes have been linked to certain serologies in specific populations, whereas classic dermatomyositis antibodies (e.g. Mi-2) tend to correlate less strongly with extensive lung disease. Nevertheless, given inter-laboratory variability and small subgroup sizes in many reports, these associations should guide clinical suspicion rather than function as deterministic rules.

Because published PM/DM-ILD data from Arab populations, and Saudi Arabia in particular, are scarce, important questions remain about regional pattern distributions, severity at presentation, and serologic correlates within local practice contexts. Differences in ancestry, environmental exposures (e.g. desert dust), and care pathways (e.g. early referral to tertiary centres) could plausibly shift both what is biologically present and what is clinically detected. To address this gap, we undertook a single-centre study in Riyadh with the primary objective of rigorously assessing ILD patterns on HRCT among patients with PM/DM using standardized ATS/ERS criteria and radiologist validation. In line with contemporary best practice and to avoid conceptual errors, pattern (NSIP, UIP, OP, DAD, airway-centred/unclassifiable) was treated as distinct from severity (extent/physiology). In addition, we characterized disease severity and described clinical-serologic features and real-world treatments as secondary, exploratory aims. By delineating the true pattern profile and its clinical context in a Saudi cohort, this report adds population-specific evidence to the literature and establishes a foundation for subsequent multicentre, longitudinal studies in the region.

METHODS

Study Design

This was a retrospective cohort study of adults who had dermatomyositis (DM) or polymyositis (PM) and developed interstitial lung disease (ILD). The research sampled only one tertiary referral centre, the Rheumatology Department, King Faisal Specialist Hospital & Research Centre (KFSH-RC), Riyadh, Saudi Arabia, taking advantage of its integrated rheumatology-pulmonology care approaches. The study plan conformed to the STROBE statement checklist for observational studies. Operating choices (definitions, abstraction, and analysis rules) were agreed upon a priori and written in an internal protocol.

Clinical Setting and Time Frame

The sampling frame from January 2006 to December 2022 encompassed a period when diagnostics and therapeutics were changing. Since clinical practice evolved over time, we prespecified era annotations to provide descriptive background. The retrospective phase of collecting data (chart review, HRCT re-adjudication, and cleaning of the database) took 12 months to finish.

Eligibility Criteria

Inclusion Criteria

- Age ≥18 years when assessed for ILD
- PM/DM classification according to the 2017 ACR/EULAR criteria. Historical classification initially given by Bohan-Peter re-adjudicated against ACR/EULAR to standardize
- HRCT-confirmed ILD, re-reviewed as detailed below

Exclusion Criteria

- Drug-induced myopathies or primary alternate diagnoses for muscle disease
- Overlap connective tissue disorders without clear PM/DM classification
- Paediatric cases (<18 years)
- Insufficient records preventing ILD adjudication or core variable abstraction

Workflow for case ascertainment

We sifted through rheumatology clinic lists, service logs for ILD, and imaging repositories for likely cases, then Verified eligibility through structured review of records. Denominators in each step as well as reasons for exclusion were recorded for clarity.

Pattern Taxonomy, Severity Grading, and Diagnosis of ILD

Primary Descriptor: ILD patterns were attributed according to consensus radiologic categories as follows: nonspecific interstitial pneumonia (NSIP), usual interstitial pneumonia (UIP), organizing pneumonia (OP), respiratory bronchiolitis-associated ILD (RB-ILD/airway-centred disease), diffuse alveolar damage (DAD), or unclassifiable when the features were mixed/insufficient. Pattern attribution was based on HRCT.

Independent Descriptor

Severity of ILD was addressed independently of pattern. We defined "mild" ILD as ≤20% parenchymal involvement on HRCT visual score; when extent was in between or not assessable, FVC ≥80% predicted served as a prespecified physiologic tie-breaker. All others were non-mild (moderate-severe). This classification schema carefully avoids defining "mild ILD" as a pattern.

HRCT Re-review, Blinding, and Adjudication

All HRCT scans available were reread by a thoracic radiologist who was blind to treatment and serology. A second reader was consulted if disagreement occurred. Differences were settled by consensus, with discrepancy log (date, location in slices, radiologic features, disposition final) maintained. When prior reports were sparse in description, we placed highest priority on direct reevaluation of images; in cases in which images were unrecoverable, we used most authoritative previous radiology report available and noted case as lower-certainty in sensitivity notes.

Variables and Measurement Framework

We used a standardized case report form and data dictionary to collect:

- **Demographics**: age at ILD diagnosis, gender, nationality
- **Disease timeline**: time of ILD in relation to PM/DM (before, simultaneously, or after diagnosis of myositis), disease duration in general
- **Clinical signs**: cough, dyspnoea on a standardized ordinal scale (e.g. MRC for dyspnoea if reported), weakness, fatigue, arthralgia/arthritis, Raynaud phenomenon, "mechanic's hands," skin signs (heliotrope rash, Gottron)
- **Serology**: ANA and myositis-specific/associated antibodies routinely available in the centre (e.g. anti-Jo-1, PL-7, PL-12, Mi-2, MDA5, SRP, NXP2, TIF1-γ, SAE1, PM-Scl)
- **Pulmonary function**: FVC and DLCO (percent predicted), collected closest to HRCT. Since the availability of PFTs was inconsistent over the long-time interval, the PFTs were rated as available-case with HRCT severity as the anchor
- **Cardiopulmonary imaging**: RVSP by transthoracic echocardiography when obtainable; ancillary chest imaging
- **Therapies**: glucocorticoids (dose strata at last follow-up), and steroid-sparing agents (mycophenolate, azathioprine, methotrexate, calcineurin inhibitors), biologics (e.g. rituximab), IVIG, cyclophosphamide, and antifibrotics

Sources of Data, Abstraction Workflow, and Quality Controls

Primary information was collected from electronic medical records; paper records from previous years were consulted

in case digital records were lacking. A piloted form was utilized by abstractors, while a 10-15% random review by a seasoned reviewer was completed in significant fields (pattern/severity, serology, PFTs, therapies). Double-entry by two distinct abstractors was done wherever feasible for critical variables, which were adjudicated by source verification in case of discordances.

Inter-rater Reliability

For the HRCT rereview subset with dual reads, we estimated simple agreement and recorded discrepant domains (e.g. ground-glass vs predominance of reticulation, traction bronchiectasis, honeycombing). Since the small rereview subset prohibited exact kappa estimation, we predefined escalation rules: recurrent disagreement invoked a consensus meeting with explicit adjudication based on features. Regular data validation checks (range, internal consistency, cross-field logic tests) were executed on database lock.

Definitions of Outcomes and Prespecified Analytic Object

Primary aim was to define HRCT patterns of ILD in PM/DM patients, independent of severity. Other, exploratory aims were to:

- Define severity on presentation;
- Report distributions of serology;
- Offer summary of symptoms and extrapulmonary disease;
- Report usage of treatment in clinical practice; and
- Offer descriptive background for timing of ILD in relation to onset of myositis

Statistical Strategy and Multiple-Testing Control

We outlined the screening cascade from initially sampled charts through to the last analytic cohort, noting reasons for exclusion (e.g. alternative diagnosis, incompleteness of imaging, age <18, non-PM/DM CTD). We appreciated subspecialty referral bias in interpretation due to the centre's subspecialty composition. We treated missing data by available-case analysis; we did not impute values due to the modest number of examples and non-random past capture of historical data. Sensitivity notes indicated where inferences were based on smaller denominators (e.g. DLCO, RVSP).

Statistical Plan and Multiple-Testing Control

Descriptive and exploratory analyses were carried out. We present continuous variables as mean±SD (or median [IQR] when non-normal); categorical as n (%). Comparisons between groups were restricted to subgroups with $n \geq 5$ to preserve validity. For related families of comparisons, we applied Holm adjustment to control the family-wise error rate. We did not apply inferential tests in very small pattern cells (e.g. UIP, OP, RB-ILD) and instead presented these purely descriptively. All p-values were interpreted circumspectly in light of sample size as well as retrospective study.

Era Effects and Descriptions of Sensitivity

Because diagnostic methods and therapies evolved from 2006 through 2022, we recorded cases by diagnostic/therapeutic period. Where period-specific variables (e.g. expanded antibody panels or adoption of mycophenolate/rituximab therapy) realistically affected ascertainment, we added narrative sensitivity background rather than overestimating statistical differences.

RESULTS

Our study included 35 ILD patients who were suffering from myositis, where 28 (80%) were females and 7 (20%) were males. Majority of the patients (97.1%) were Saudi citizens. The type of myositis showed that 21 (60%) had Dermatomyositis and remaining 14 (40%) had Polymyositis. The most common ILD was Non-specific Interstitial pneumonia (37.1%), followed by Unclassified (14.3%), Usual interstitial pneumonia (5.7%), Respiratory bronchiolitis ILD (2.9%) and Cryptogenic organizing pneumonia (2.9%) (Table 1). The onset of ILD showed that 48.6% were PM/DM-preceding ILD, 31.4% were concomitant and 20% were ILD proceeding PM/DM. The distribution of ILD type based on two type of myositis is given in Table 2 and, which showed no statistically significant differences.

The assessment of clinical features in these patients are as follows: Fatigue (82.9%), Fever (11.4%), Weight loss (31.4%), Weakness (88.6%), Dysphagia (28.6%), exertional dyspnoea (74.3%), Non-productive cough (62.9%), Myocarditis (0%), Pericarditis (2.9%), Arthralgia (77.1%) and pulmonary hypertension (23.5%). However, we didn't find any statistically significant differences in their distribution between type of ILD ($P>0.05$), except for pulmonary hypertension, which was found to be significantly higher among Usual interstitial pneumonia cases ($p=0.040$) (Table 3).

The distribution of skin manifestations according to ILD type are given in Table 4. The distribution is as follows: Heliotrope rash (34.3%), Gottron papules (34.3%), mechanic hand (20%), V-sign (14.3%), shawl sign (11.4%), holster sign (2.9%), Raynaud phenomena (14.3%), Fingers tips ulcers (8.6%), and Calcinosis (8.6%). No statistically significant differences in distribution according to ILD type was observed ($p>0.05$).

Table 1: Demographic data and medical history

Parameters	Variables	N	%
Gender	Male	7	20.0
	Female	28	80.0
Nationality	Non-Saudi	1	2.9
	Saudi	34	97.1
Myositis type	Dermatomyositis	21	60.0
	Polymyositis	14	40.0
Comorbidities	Diabetes	6	17.1
	Hypertension	6	17.1
	Dyslipidaemia	7	20.0
	Thromboembolic Disease	5	14.3
	Hypothyroidism	5	14.3
	Osteoporosis	5	14.3
	Any malignancy	3	8.6
Type of ILD	Non-specific Interstitial pneumonia	13	37.1
	Usual interstitial pneumonia	2	5.7
	Mild ILD	13	37.1
	Respiratory bronchiolitis ILD	1	2.9
	Cryptogenic organizing pneumonia	1	2.9
	Unclassified	5	14.3
ILD onset	ILD-preceding	7	20.0
	Concomitant	11	31.4
	PM/DM-preceding	17	48.6

Table 2: Distribution of ILD based on myositis type

Parameters	ILD Type						P value
	NSIP	UIP	Mild ILD	RB-ILD	COP	Unclassified	
Dermatomyositis	9 (69.2%)	0 (0%)	7 (53.8%)	0 (0%)	1 (100%)	4 (80%)	
Polymyositis	4 (30.8%)	2 (100%)	6 (46.2%)	1 (100%)	0 (0%)	1 (20%)	0.247

Table 3: Clinical presentation according to ILD type

Parameters		ILD Type						Total	P values
		NSIP	UIP	Mild ILD	RB-ILD	COP	Unclassified		
Fatigue		11	2	10	1	1	4	29	
		84.6%	100.0%	76.9%	100.0%	100.0%	80.0%	82.9%	0.944
Fever		3	0	1	0	0	0	4	
		23.1%	0.0%	7.7%	0.0%	0.0%	0.0%	11.4%	0.687
Weight loss		3	1	5	1	0	1	11	
		23.1%	50.0%	38.5%	100.0%	0.0%	20.0%	31.4%	0.552
Weakness		10	2	12	1	1	5	31	
		76.9%	100.0%	92.3%	100.0%	100.0%	100.0%	88.6%	0.687
Type weakness	Proximal	9	2	9	1	0	2	23	
		69.2%	100.0%	69.2%	100.0%	0.0%	40.0%	65.7%	
	Distal	0	0	1	0	0	0	1	
		0.0%	0.0%	7.7%	0.0%	0.0%	0.0%	2.9%	
	Both	4	0	3	0	1	3	11	
		30.8%	0.0%	23.1%	0.0%	100.0%	60.0%	31.4%	0.681
Arthralgia (joint pain)		12	1	8	1	1	4	27	
		92.3%	50.0%	61.5%	100.0%	100.0%	80.0%	77.1%	0.423
Dysphagia		3	0	5	0	1	1	10	
		23.1%	0.0%	38.5%	0.0%	100.0%	20.0%	28.6%	0.454
Exertional dyspnoea		12	1	7	1	0	5	26	
		92.3%	50.0%	53.8%	100.0%	0.0%	100.0%	74.3%	0.059
Nonproductive cough		11	1	5	1	0	4	22	
		84.6%	50.0%	38.5%	100.0%	0.0%	80.0%	62.9%	0.109
Pericarditis		0	0	1	0	0	0	1	
		0.0%	0.0%	7.7%	0.0%	0.0%	0.0%	2.9%	0.884
Myocarditis		0	0	0	0	0	0	0	
		0.0%	0.0%	0.0%	0.0%	0.0%	0.0%	0.0%	NA
Pulmonary hypertension		3	2	1	1	0	1	8	
		23.1%	100.0%	8.3%	100.0%	0.0%	20.0%	23.5%	0.040

Table 4: Skin manifestations according to ILD type

Parameters		ILD Type						Total	P value
		NSIP	UIP	Mild ILD	RB-ILD	COP	Unclassified		
Mechanic hand	N	4	1	1	0	1	0	7	
	%	30.8%	50.0%	7.7%	0.0%	100.0%	0.0%	20.0%	0.117
Heliotrope rash	N	2	0	6	0	1	3	12	
	%	15.4%	0.0%	46.2%	0.0%	100.0%	60.0%	34.3%	0.166
Gottron papules	N	5	0	4	0	1	2	12	
	%	38.5%	0.0%	30.8%	0.0%	100.0%	40.0%	34.3%	0.589
Holster sign	N	0	0	0	0	0	1	1	
	%	0.0%	0.0%	0.0%	0.0%	0.0%	20.0%	2.9%	0.289
V-sign	N	2	0	2	0	1	0	5	
	%	15.4%	0.0%	15.4%	0.0%	100.0%	0.0%	14.3%	0.195
Rash over upper back, shoulders, and back of the neck	N	1	0	2	0	0	1	4	
	%	7.7%	0.0%	15.4%	0.0%	0.0%	20.0%	11.4%	0.939
Raynaud phenomena	N	2	0	3	0	0	0	5	
	%	15.4%	0.0%	23.1%	0.0%	0.0%	0.0%	14.3%	0.801
Fingers tips ulcers	N	3	0	0	0	0	0	3	
	%	23.1%	0.0%	0.0%	0.0%	0.0%	0.0%	8.6%	0.352
Calcinosis	N	1	0	0	0	0	2	3	
	%	7.7%	0.0%	0.0%	0.0%	0.0%	40.0%	8.6%	0.161

Table 5: Lab investigations according to ILD type

Parameters	ILD Types							P value
	NSIP	UIP	Mild ILD	RB-ILD	COP	Unclassified	Total	
Anaemia	2	0	4	1	1	0	8	0.084
	15.4%	0.0%	30.8%	100.0%	100.0%	0.0%	22.9%	
Leukopenia	1	0	3	0	0	0	4	0.687
	7.7%	0.0%	23.1%	0.0%	0.0%	0.0%	11.4%	
Thrombocytopenia	2	0	1	0	0	0	3	0.898
	15.4%	0.0%	7.7%	0.0%	0.0%	0.0%	8.6%	
Raised Creatinine phosphokinase	11	2	11	1	1	5	31	0.906
	84.6%	100.0%	84.6%	100.0%	100.0%	100.0%	88.6%	
Raised ESR	9	2	9	1	1	3	25	0.852
	69.2%	100.0%	69.2%	100.0%	100.0%	60.0%	71.4%	
Raised CRP	9	2	9	1	1	3	25	0.741
	69.2%	100.0%	69.2%	100.0%	100.0%	60.0%	71.4%	
ANA - Positive	11	2	12	1	0	3	29	0.149
	84.6%	100.0%	92.3%	100.0%	0.0%	60.0%	82.9%	
Anti-Jo-1	6	2	2	1	0	1	12	0.087
	46.2%	100.0%	15.4%	100.0%	0.0%	20.0%	34.3%	
AntiMi2	1	0	0	0	1	1	3	0.023
	7.7%	0.0%	0.0%	0.0%	100.0%	20.0%	8.6%	
AntiTIF1	0	0	1	0	0	0	1	0.884
	0.0%	0.0%	7.7%	0.0%	0.0%	0.0%	2.9%	
AntiMDA5	2	0	0	0	0	1	3	0.670
	15.4%	0.0%	0.0%	0.0%	0.0%	20.0%	8.6%	
AntiNXP2	1	0	0	0	0	1	2	0.697
	7.7%	0.0%	0.0%	0.0%	0.0%	20.0%	5.7%	
AntiSAE1	1	0	0	0	1	0	2	0.003
	7.7%	0.0%	0.0%	0.0%	100.0%	0.0%	5.7%	
Anti-Ku	0	0	0	0	0	1	1	0.289
	0.0%	0.0%	0.0%	0.0%	0.0%	20.0%	2.9%	
PMScl75	3	0	1	0	0	0	4	0.687
	23.1%	0.0%	7.7%	0.0%	0.0%	0.0%	11.4%	
PL7	2	0	1	0	0	1	4	0.939
	15.4%	0.0%	7.7%	0.0%	0.0%	20.0%	11.4%	
PL12	1	0	0	0	0	1	2	0.697
	7.7%	0.0%	0.0%	0.0%	0.0%	20.0%	5.7%	
SRP	1	0	1	0	0	1	3	0.942
	7.7%	0.0%	7.7%	0.0%	0.0%	20.0%	8.6%	
Anti DNA Positive	0	0	1	0	0	1	2	0.697
	0.0%	0.0%	7.7%	0.0%	0.0%	20.0%	5.7%	
AntiU1RNP Positive	0	0	2	0	0	0	2	0.610
	0.0%	0.0%	15.4%	0.0%	0.0%	0.0%	5.7%	
Rheumatoid factor Ab	2	1	4	0	0	0	7	0.551
	15.4%	50.0%	30.8%	0.0%	0.0%	0.0%	20.0%	
Lupus anticoagulant-Positive	0	0	0	0	0	0	0	NA
	0.0%	0.0%	0.0%	0.0%	0.0%	0.0%	0.0%	
Anti Beta IgG Positive	1	0	1	0	0	0	2	0.981
	7.7%	0.0%	7.7%	0.0%	0.0%	0.0%	5.7%	
Anti_Beta_IgM	1	0	3	0	0	0	4	0.687
	7.7%	0.0%	23.1%	0.0%	0.0%	0.0%	11.4%	
Anti_Card_IgG Positive	0	0	1	0	0	0	1	0.884
	0.0%	0.0%	7.7%	0.0%	0.0%	0.0%	2.9%	
Anti_Card_IgM	2	0	0	0	0	1	3	0.670
	15.4%	0.0%	0.0%	0.0%	0.0%	20.0%	8.6%	
Anti Ro/SSA	5	2	4	0	0	2	13	0.438
	38.5%	100.0%	30.8%	0.0%	0.0%	40.0%	37.1%	
Anti_LA	0	0	0	0	0	0	0	NA
	0.0%	0.0%	0.0%	0.0%	0.0%	0.0%	0.0%	
C3	0	0	2	0	0	0	2	0.610
	0.0%	0.0%	15.4%	0.0%	0.0%	0.0%	5.7%	
C4	0	0	2	0	0	0	2	0.610
	0.0%	0.0%	15.4%	0.0%	0.0%	0.0%	5.7%	

Table 6: Comparison of Forced Vital capacity, Diffusing capacity for carbon monoxide and Echo-RSVP based on ILD type

Parameters	Variables	Mean	SD	P value
Forced Vital capacity %	Non-specific Interstitial pneumonia	0.613	0.189	0.009
	Usual interstitial pneumonia	0.490	-	
	Mild ILD	0.841	0.117	
	Respiratory bronchiolitis ILD	0.350	-	
	Cryptogenic organizing pneumonia	0.790	-	
	Unclassified	0.783	0.156	
Diffusing capacity of the lungs for carbon monoxide (DLCO) %	Non-specific Interstitial pneumonia	0.793	0.149	0.013
	Usual interstitial pneumonia	.	-	
	Mild ILD	1.036	0.139	
	Respiratory bronchiolitis ILD	-	-	
	Cryptogenic organizing pneumonia	0.820	-	
	Unclassified	0.875	0.258	
Pulmonary hypertension (mmHG)	Non-specific Interstitial pneumonia	26.462	9.052	0.005
	Usual interstitial pneumonia	44.725	62.614	
	Mild ILD	20.333	1.155	
	Respiratory bronchiolitis ILD	80.000	-	
	Cryptogenic organizing pneumonia	20.000	-	
	Unclassified	24.000	8.944	

Table 7: Use of medicine according to ILD type

Parameters		NSIP	UIP	Mild ILD	RB-ILD	COP	Unclassified	Total
Steroid use		13	2	13	1	1	5	35
		100.0%	100.0%	100.0%	100.0%	100.0%	100.0%	100.0%
Steroid maintenance dose	=<7.5	10	1	11	0	0	5	27
		76.9%	50.0%	84.6%	0.0%	0.0%	100.0%	77.1%
	>7.5	2	1	1	1	0	0	5
		15.4%	50.0%	7.7%	100.0%	0.0%	0.0%	14.3%
	off steroid	1	0	1	0	1	0	3
		7.7%	0.0%	7.7%	0.0%	100.0%	0.0%	8.6%
Mycophenolate mofetil		11	2	3	1	0	2	19
		84.6%	100.0%	23.1%	100.0%	0.0%	40.0%	54.3%
Rituximab		7	2	6	0	0	2	17
		53.8%	100.0%	46.2%	0.0%	0.0%	40.0%	48.6%
Immunoglobulin		1	1	4	1	1	1	9
		7.7%	50.0%	30.8%	100.0%	100.0%	20.0%	25.7%
Cyclophosphamide		0	0	0	0	0	1	1
		0.0%	0.0%	0.0%	0.0%	0.0%	20.0%	2.9%
Azathioprine		3	0	9	0	0	2	14
		23.1%	0.0%	69.2%	0.0%	0.0%	40.0%	40.0%
Calcineurin inhibitors		2	0	1	0	0	1	4
		15.4%	0.0%	7.7%	0.0%	0.0%	20.0%	11.4%
Methotrexate		2	0	8	0	1	1	12
		15.4%	0.0%	61.5%	0.0%	100.0%	20.0%	34.3%
Antifibrotic		4	0	0	0	0	0	4
		30.8%	0.0%	0.0%	0.0%	0.0%	0.0%	11.4%

The lab and other blood investigations based in type of ILDs are given in Table 5. The features are as follows: Anaemia (22.9%), Leukopenia (11.4%), Thrombocytopenia (8.6%), Raised Creatinine phosphokinase (88.6%), raised ESR (71.4%), raised CRP (71.4%), raised AST (54.3%), raised ALT (54.3%), raised LDH (80%), raised aldolase (8.6%).

The Autoimmune serology are as follows: Anti-nuclear antibody positive (82.9%), anti-Jo1 (34.3%),anti-Mi2 (8.6%), anti-TIF1 (2.9%), anti-MDA5 (8.6%), anti-NXP2 (5.7%), anti-SAE1 (5.7%), anti-Ku (2.9%), PM-Scl75 (11.4%), PL-7 (11.4%), PL-12 (11.4%), SRP (8.6%), anti-DNA (5.7%), anti-U1RNP (5.7%), Rheumatoid factor (20%), Lupus anticoagulant (0%), anti-beta 2-glycoprotien IgG (5.7%), anti-beta 2-glycoprotien IgM (11.4%), anti-cardiolipin Ab IgG (2.9%), anti-cardiolipin Ab IgM (8.6%), anti-Ro/SSA (37.1%), anti La/SSB (0%), low Complement C3 (5.7%), and low Complement C4 (5.7%). It was found that AntiMi2 was significantly higher in Cryptogenic organizing pneumonia compared to other types (p=0.023). Alsoanti-SAE1 was found to be significantly higher in Cryptogenic organizing pneumonia and Non-specific Interstitial pneumonia (p=0.003).

All 35 patients were found to be using steroids, with 77% of them maintaining their dosage at or below 7.5 mg, 14.3% above 7.5 mg, and 8.6% were off steroids. Patients who maintained on steroid dosage more than 7.5 mg daily are as; RB-ILD (100%), UIP (50%), NSIP (15%), Mild ILD (7.7%). Other medications use are as follows: Mycophenolate mofetil (54.3%), Rituximab (48.6%), Immunoglobulin (25.7%), Cyclophosphamide (2.9%), Azathioprine (40%), Calcineurin inhibitors (11.4%), Methotrexate (34.3%), and antifibrotic (11.4%) (Table 7).

The mean forced vital capacity (FVC) in ILD patients was found to be 0.7123±0.1956 %, which showed significantly lower was found in Respiratory bronchiolitis ILD (p=0.009). The mean diffusing capacity of the lungs for carbon monoxide (DLCO) was found to be 0.901±0.191, which was also significantly higher in mild ILD (p=0.013). The mean right ventricle systolic pressure on TT-Echo (Echo-RSVP) in these patients was found to be 26.39 ±16.78, which was significantly higher in respiratory bronchiolitis ILD (p=0.005) (Table 6).

DISCUSSION

Despite advances in treatment, pulmonary problems continue to be recognized as a major contributor to morbidity in people with PM/DM.ILD can cause symptoms such as cough, shortness of breath, and chest discomfort. These symptoms can be distressing and affect a person's ability to perform daily activities, leading to a reduced quality of life [12]. Reduced lung function and breathlessness can limit a person's ability to engage in physical activities and exercise [13]. This may lead to a decrease in overall fitness and physical well-being. Both PM/DM are associated with muscle weakness and fatigue, and the addition of ILD can exacerbate these symptoms [14]. Fatigue can further limit a person's ability to participate in activities and negatively impact their quality of life. Living with a chronic illness like dermatomyositis or polymyositis, along with ILD, can lead to increased stress, anxiety, and depression [15]. Coping with the physical and emotional challenges of these conditions can affect one's mental well-being and overall quality of life. The unpredictable nature of autoimmune diseases like PM/DM, along with ILD, can cause uncertainty about the future [14]. This uncertainty can lead to emotional distress and affect one's overall sense of well-being.

The comparison of clinical features will shed light on the distinct presentations of ILD in DM and PM patients. The presence of characteristic skin findings in DM patients and the absence of these in PM patients might contribute to different diagnostic challenges and prognostic implications. In this retrospective investigation, we included 35 PM/DM patients with ILD who were not specifically chosen for their pulmonary symptoms or other characteristics of PM/DM. This allowed us to rule out the possibility of a selection bias related to the severity of the disease, as indicated by either clinical or subclinical symptoms. Imaging patterns were classified as usual interstitial pneumonia (UIP) pattern (combination of reticular opacities, traction bronchiectasis, and honeycombing with basal and subpleural predominance, and minimal ground-glass opacities); nonspecific interstitial pneumonia (NSIP) pattern (combination of ground-glass attenuation and reticulation, with little if any honeycombing); organizing pneumonia (OP) pattern (patchy consolidation); or unclassifiable pattern [40].

The time at which ILD symptoms first appeared could not be used as a predictor of how well the lungs would function [22]. Several autoimmune diseases, including scleroderma, lupus, and rheumatoid arthritis, have been linked to interstitial pneumonitis [23,24]. However, the frequency with which polymyositis patients develop interstitial pneumonitis is unknown.

The relationship between dermatomyositis and ILD is complex, and not all individuals with DM/PM will develop ILD. ILD is the presenting feature and has been reported to precede signs of clinical myopathy in 7.2% to 37.5% of cases [42,43,44]. In our cohort, nearly half developing ILD after PM/DM (48.8%). Myositis-ILD can present in a variety of patterns, though NSIP is by far the commonest ILD pattern in DM/PM patients, comprising 61-81.8% of cases in the largest series [42, 45, 46]. In our cohort, the most common ILD was Non-specific Interstitial pneumonia (37.1%) and Mild ILD (37.1%), followed by Unclassified (14.3%), Usual interstitial pneumonia (5.7%) followed by Respiratory bronchiolitis ILD and Cryptogenicorganizing pneumonia (2.9%).

Myositis is twice as common in females as in males, with a peak incidence in the fifth and sixth decades of life [28]. Our study findings also showed a higher prevalence among females compared to males with a ratio of 4:1. Evidence shows that autoimmune diseases often have a higher incidence in women, possibly due to differences in immune responses between the sexes. Hormonal factors, including oestrogen, have been suggested to play a role in modulating immune function and may contribute to the sex bias [29]. It's important to note that the onset of DM/PM can be gradual or sudden, and the severity of symptoms can vary. Some people may initially have muscle symptoms, while others may notice skin changes first. In cases where ILD is associated with DM/PM, it may develop concurrently with muscle and skin symptoms or at a later stage [30].

The mean age of the individuals in the current cohort of 35 cases of ILD was 40 years old, and the maximum was 69 years, and about 80% of the patients were female. Non-productive cough and dyspnoea are the most prominent symptoms [31]. Predicting which people with PM-DM will get interstitial pneumonitis is challenging. Mende et al. reported that there appears to be no correlation between the severity or duration of the disease, the existence of positive serological tests, or the extent to which muscle enzymes are elevated [32].

Limitations of the study could include its retrospective nature, potential selection bias, and variations in treatment strategies among patients. Due to the study's single-centre design, there weren't enough patients for a reliable statistical analysis. Due to the retrospective nature of the study, PFTs were not performed at consistent intervals, and there was substantial between-time point variation in FVC and DLCO for certain individuals.

Clinical Recommendations

Management of clinical care in PM/DM-ILD consists of early detection, frequent follow-up, and individualized therapy. Pulmonary manifestations like cough, shortness of breath, and chest pain decrease the quality of life as well as potentially aggravate pre-existing fatigue and weakness. Since the extent of disease in the lungs is not always reflected in the symptoms, baseline HRCT scans and pulmonary function studies are done

at the time of diagnosis, even in symptomatic individuals, and are redone in intervals to monitor disease progression. Echocardiograms are performed in cases with suspicion of pulmonary hypertension.

Serologic tests, among them myositis-specific and associated antibodies, aid in diagnosis as well as in the detection of patients who are at increased risk, specifically those with anti-synthetase syndrome. Approach to therapy involves corticosteroids as first-line therapy with prompt switching to steroid-sparing drugs like mycophenolate or rituximab in most instances. Azathioprine, methotrexate, calcineurin inhibitors, IVIG, and in selected cases antifibrotics are employed according to disease phenotype, intensity, as well as tolerance.

Supportive treatment is essential: pulmonary rehab, vaccination, prevention of infection, and comorbidity management contribute to maintaining long-term health. Activity pacing and mental health referrals are taught to deal with the stress and unpredictability of chronic disease. If progression continues despite conventional treatment, cases are discussed in multidisciplinary meetings, with thought given to escalation to more advanced interventions like clinical trials or lung transplantation.

CONCLUSIONS

Our results further suggest that pulmonary function tests and high-resolution computed tomography (HRCT) of the lungs be part of the standard protocol for evaluating and monitoring individuals with polymyositis and dermatomyositis. Most of the patients found to have Non-specific Interstitial pneumonia pattern. The findings can aid clinicians in early diagnosis, appropriate treatment selection, and improved patient outcomes. Further prospective studies are warranted to validate these findings and explore potential molecular mechanisms underlying the distinct ILD manifestations in DM and PM.

Ethical Statement

The study was approved by Institutional Review Board (IRB) of KFSH-RC. Due to the retrospective nature, de-identified use of data, and low risk, the IRB provided Waiver of Informed Consent. All procedures conformed to institutional confidentiality and data-protection guidelines. We note specifically that over-testing and misclassification are risks in retrospect studies; our radiologist-verified HRCT rereview and formal structured data audit were implemented to reduce these risks and improve the credibility of assessed patterns and severity reporting.

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