Unveiling the pulmonary burden of idiopathic inflammatory myopathies in South Africa

The idiopathic inflammatory myopathies (IIMs) are a group of rare, chronic, autoimmune diseases characterised by inflammation and progressive weakness of the skeletal muscles. The primary forms of IIM are dermatomyositis (DM), polymyositis, inclusion body myositis, the antisynthetase syndrome, overlap syndromes, and clinically amyopathic dermatomyositis.[1] The IIMs are of relevance to the pulmonologist because of their frequent association with pulmonary complications, in particular interstitial lung disease (ILD), which contributes significantly to the morbidity and mortality of affected individuals.

In this issue of *AJTCCM*, Hes *et al.*[2] retrospectively describe the spectrum and prevalence of these pulmonary manifestations in a local patient population, offering crucial insights in a region-specific context. They studied a cohort of 77 patients diagnosed with IIM, all presenting with respiratory complaints at a tertiary care facility in Johannesburg. Most patients (almost two-thirds) had DM. Pulmonary complications were documented in 85% of patients, with a higher incidence in the DM subgroup compared with other IIMs. As expected, ILD was the most prevalent pulmonary complication,[3] present in ~70% of patients, with nonspecific interstitial pneumonitis (NSIP) being the predominant pattern of ILD, present in just over one-third of patients. This finding is consistent with the global understanding of ILD in IIM, where NSIP is frequently observed.[4] Infection and pulmonary hypertension also contributed to the burden of disease. Relevant to the South African (SA) context is that ~10% of patients presented with pulmonary tuberculosis (TB) as an immunosuppression-associated infection, underscoring the importance of vigilant screening and management of TB in this population. In addition to ILD, alveolar hypoventilation secondary to diaphragmatic and chest wall muscle weakness is another cause of respiratory insufficiency in IIMs, but this complication was not reported by Hes *et al.*[2]

A few points are worthy of note. Firstly, the study confirmed the importance of clinical assessment of patients with IIMs, with dyspnoea and cough, and the presence of bibasal crackles on auscultation, independently associated with physiological restriction and reduced diffusing capacity. Other clinical features associated with poor outcome in ILD in IIMs reported in the literature include progressive pulmonary restriction after 3 months, heliotrope erythema, delay in diagnosis, and Raynaud's phenomenon.[5] Secondly, the authors found that in approximately one-fifth of patients, respiratory symptoms predated the onset of clinical myopathy, in contrast to studies from other settings that suggest that they occur contemporaneously.[6] Thirdly, the authors replicated the strong association between anti-Jo1 antibody (histidyl-tRNA synthetase)-positive status and elevated acute inflammatory markers and muscle enzymes, indicating a more aggressive disease phenotype.[7] The presence of this serological marker underscores the need for tailored therapeutic strategies to mitigate the inflammatory cascade and preserve pulmonary function in these patients. And lastly, the study emphasises the increased susceptibility of patients on chronic immunosuppressive drugs to TB, where SA presents a particularly high-risk environment. No data are given on the prescription of TB prophylaxis in this cohort, but the rationale for its use may be extrapolated from other studies of vulnerable populations, such as patients on chronic haemodialysis or the recipients of solid organ transplants, where it may mitigate the risk of latent TB reactivation and new TB infections.[8-10]

In conclusion, and notwithstanding the limitations of the retrospective design, this study provides important insights into the pulmonary complications in SA patients with IIM. The high prevalence of ILD, coupled with significant rates of pulmonary infections and pulmonary hypertension, underscores the multifaceted nature of pulmonary involvement in IIM. The findings advocate for heightened awareness, early diagnostic evaluations, and prompt therapeutic interventions to mitigate the pulmonary morbidity in this patient population. As we deepen our understanding of the regional manifestations of IIM, tailored strategies can be developed to improve patient outcomes and quality of life.

Gregory L Calligaro, BSc Hons, MB ChB, Dip PEC (SA), MMed (Med), FCP (SA), Cert Pulmonology (SA) Phys
Centre for Lung Infection and Immunity, Division of Pulmonology, Department of Medicine, Groote Schuur Hospital and UCT Lung Institute, University of Cape Town, South Africa
greg.calligaro@uct.ac.za

1. Milone M. Diagnosis and management of immune-mediated myopathies. Mayo Clin Proc 2017;92(5):826-837. https://doi.org/10.1016/j.mayocp.2016.12.025
2. Hes T, Wong M, Tikly M, Govind N. Pulmonary manifestations of the idiopathic inflammatory myopathies in a South African population. Afr J Thoracic Crit Care Med 2024;30(3):e1663. https://doi.org/10.7196/AJTCCM.2024.v30i3.1663
3. Long K, Danoff SK. Interstitial lung disease in polymyositis and dermatomyositis. Clin Chest Med 2019;40(3):561-572. https://doi.org/10.1016/j.ccm.2019.05.004
4. Kiely PD, Chua F. Interstitial lung disease in inflammatory myopathies: Clinical phenotypes and prognosis. Curr Rheumatol Rep 2013;15(9):359. https://doi.org/10.1007/s11926-013-0359-6
5. Cobo-Ibáñez T, López-Longo F-J, Joven B, et al. Long-term pulmonary outcomes and mortality in idiopathic inflammatory myopathies associated with interstitial lung disease. Clin Rheumatol 2019;38(3):803-815. https://doi.org/10.1007/s10067-018-4353-2
6. Vu TTT, Brown KK, Solomon JJ. Myositis-associated interstitial lung disease. Curr Opin Pulm Med 2023;29(5):427-435. https://doi.org/10.1097/MCP.0000000000001000
7. Chang S, Shin MH, Shin JH, Park MS. Clinical characteristics of anti-Jo-1-positive interstitial lung disease. Eur Respir J 2019;54(Suppl 63):PA5183. https://doi.org/10.1183/13993003.congress-2019.PA5183
8. Ndamase S, Okpechi I, Carrara H, Black J, Calligaro G, Freercks R. Tuberculosis burden in stage 5 chronic kidney disease patients undergoing dialysis therapy at Livingstone Hospital, Port Elizabeth, South Africa. S Afr Med J 2020;110(5):422-426. https://doi.org/10.7196/SAMJ.2020.v110i5.14035
9. Torre-Cisneros J, Doblas A, Aguado JM, et al. Tuberculosis after solid-organ transplant: Incidence, risk factors, and clinical characteristics in the RESITRA (Spanish Network of Infection in Transplantation) cohort. Clin Infect Dis 2009;48(12):1657-1665. https://doi.org/10.1086/599035
10. Yuan Z, Chao S, Xu Y, Niu Y. Chemoprophylaxis for the prevention of tuberculosis in kidney transplant recipients: A systematic review and meta-analysis. Front Pharmacol 2023;14:1022579. https://doi.org/10.3389/fphar.2023.1022579

Afr J Thoracic Crit Care Med 2024;30(3):e2682. https://doi.org/10.7196/AJTCCM.2024.v30i3.2682