Microscopic Polyangiitis associated Interstitial Lung Disease (MPA-ILD) vs Idiopathic Pulmonary Fibrosis (IPF)

Presenter: Dr. Dhwani Manoj Gohel, Advanced Trainee, Blacktown Hospital, Department of General/ Acute Medicine and Conjoint Associate Lecturer, University of New South Wales, Sydney

Authors: *Dr. Dhwani Manoj Gohel*, *Advanced Trainee*, *Blacktown Hospital, Department of General/ Acute Medicine and Conjoint Associate Lecturer, University of New South Wales, Sydney, Dr. Shakti Dabholkar, Blacktown Hospital, Department of Respiratory & Adjunct Lecturer, University of New South Wales, Sydney and Dr. Surjit Tarafdar, Blacktown Hospital, Department of Nephrology and General/ Acute Medicine*

Pulmonary fibrosis is a rare complication of Microscopic Polyangiitis (MPA)[1,2]. It is crucial for clinicians to suspect Microscopic Polyangiitis-Interstitial Lung Disease (MPA-ILD) as this group tends to have poorer prognosis than patients with Idiopathic Pulmonary Fibrosis (IPF)[3].

FE is a 79-year-old male ex-smoker who was diagnosed with MPA in 2015 when he presented with renal impairment. He underwent a renal biopsy which pathologically confirmed the diagnosis and was subsequently treated with Mycophenolate and Prednisolone. His other medical history includes chronic kidney disease, chronic inflammatory demyelinating polyneuropathy (CIDP), latent tuberculosis treated with Rifampicin, chronic Hepatitis B, thrombocytopenia, osteopenia and left cataract secondary to prednisone use. The initial suspicion of ILD was in 2019 when he was incidentally found to have fine bibasal inspiratory crepitations on auscultation. This was confirmed on high-resolution CT (HRCT) chest which showed findings consistent with Usual Interstitial Pneumonitis (UIP) with suspicion of MPA as the possible cause. He was continued on his immunosuppression at the time.

He recently presented with worsening dyspnoea on minimal exertion. Repeat HRCT chest performed in February 2023 demonstrated extensive honeycombing with a basal predominance along with interlobular reticular thickening consistent in a UIP pattern. Other known causes of ILD were ruled out by serological testing. After an extensive discussion in the ILD multidisciplinary meeting, it was concluded that his clinical pattern and imaging CT chest were more consistent with IPF rather than MPA-ILD.

This case highlights the difficulties in eliciting the aetiology of ILD in patients with autoimmune disease. Clinicians should be aware of the well-established yet lesser known association between MPA and ILD and the importance of establishing MPA-ILD vs IPF to guide management.