Reply to the comment on: Successful management of pulmonary hypertension with baricitinib in a dermatomyositis patient

Sirs,

We read with great interest the reply by Pinto Oliveira *et al*. (1) concerning our recently published manuscript describing the successful use of baricitinib in a patient with dermatomyositis (DM) and pulmonary hypertension (PH). We appreciate the thoughtful comments and would like to take this opportunity to clarify some of the observations raised.

We fully acknowledge the classification of PH as outlined in the reply. Our team operates within a multidisciplinary framework that includes rheumatologists, radiologists, pulmonologists, and cardiologists, all highly experienced in the diagnosis and management of PH. Our Unit has developed considerable expertise in handling such complex clinical scenarios, and this case was no exception.

The diagnosis of Group 1 PH, namely pulmonary arterial hypertension (PAH), in our patient, although rare in the context of DM, was well substantiated and is supported by similar cases in the literature (2). Both idiopathic inflammatory myopathies (IIM) and PAH were fully characterised, excluding other causes of PH, such as overlap with systemic sclerosis or ILD. Moreover, our patient was positive for anti-Ku antibodies, which have already been described in patients with IIM and PAH (3, 4). The decision-making process regarding diagnostic procedures was carefully evaluated. Given the absence of any clinical or instrumental signs suggestive of chronic thromboembolic disease, a ventilation/perfusion lung scan was not deemed necessary. The patient was a young adult with no personal history of thrombosis or risk factors associated with a prothrombotic state. In this context, the clinical judgment supported by the multidisciplinary team led to a reasoned and cautious diagnostic approach.

Therapeutically, intravenous immunoglobulins, part of the recommended treatment options for inflammatory myopathies, were administered with benefit. Baricitinib was selected after careful consideration, and we considered it both safe and appropriate in this clinical setting. Importantly, the therapeutic strategy was not limited to the management of PH alone. The patient presented with severe and refractory cutaneous involvement, persistently elevated inflammatory markers, and had previously experienced a significant cutaneous flare and gastrointestinal side effects following the administration of PAH treatments. These complexities prompted a comprehensive internal discussion within our Myositis Unit, along with an extensive review of the existing literature (5), to identify a safe and effective treatment path.

Given the patient's young age, the psychological impact of his condition, and the invasive nature of right heart catheterisation, the decision not to repeat this procedure was supported by the clear resolution of PAH as documented by echocardiography repeatedly performed by an expert cardiologist, and by the patient's evident and sustained clinical improvement. He remained in stable condition under close and continuous follow-up.

In conclusion, our report highlights a novel and promising therapeutic use of baricitinib in a DM patient with PAH, a potential application supported by preclinical evidence that warrants further investigation. This case highlights the importance of individualised care and confirms the essential role of a highly specialised multidisciplinary team in the management of complex patients with IIM.

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References

1. RIZZO C, CAMARDA F, LA BARBERA L, GUGGINO G: Successful management of pulmonary hypertension with baricitinib in a dermatomyositis patient. *Clin Exp Rheumatol* 2025; 43(2): 390. https://doi.org/10.55563/clinexprheumatol/c6an8c
1. PINTO OLIVEIRA C, PASCOAL D, PRATA AR, CARVALHO D, CASTRO G, DOURADO E: Comment on: Successful management of pulmonary hypertension with baricitinib in a dermatomyositis patient. *Clin Exp Rheumatol* 2026; 44(2): 413.
2. SANGES S, YELNIK CM, SITBON O *et al*.: Pulmonary arterial hypertension in idiopathic inflammatory myopathies: Data from the French pulmonary hypertension registry and review of the literature. *Medicine* 2016; 95(39): e4911. https://doi.org/10.1097/md.0000000000004911
3. RIGOLET A, MUSSET L, DUBOURG O *et al*.: Inflammatory myopathies with anti-Ku antibodies: a prognosis dependent on associated lung disease. *Medicine* 2012; 91(2): 95-102. https://doi.org/10.1097/md.0b013e31824d9cec
4. ISERN RA, YANEVA M, WEINER E *et al*.: Autoantibodies in patients with primary pulmonary hypertension: association with anti-Ku. *Am J Med* 1992; 93(3): 307-12. https://doi.org/10.1016/0002-9343(92)90238-7
5. RIZZO C, GRAZZINI S, CONTICINI E *et al*.: Small molecules in idiopathic inflammatory myopathies: a systematic review and a multicenter case series about Janus kinase inhibitors and apremilast. *Reumatismo* 2025; 77(2). https://doi.org/10.4081/reumatismo.2025.1718