P73 PREDICTORS OF FLARES AFTER GLUCOCORTICOID DISCONTINUATION IN REMITTED PATIENTS WITH SLE

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Objective Data on the safety and feasibility of the discontinuation glucocorticoids (GCs) in remitted patients with SLE are scanty and controversial results have been published. Our aim was to assess the predictors of flares after GCs discontinuation in a large cohort of prospectively followed patients.

Methods Patients diagnosed after 1980 and followed-up until 2023 who achieved remission lasting at least 6 months at least once during their disease course were included. Remission was defined as clinical SLEDAI-2K=0 on a stable immunosuppressive and/or antimalarial therapy and/or prednisone ≤5 mg/day. Flares were defined as any increase in clinical SLEDAI-2K>0 or the need for changes in SLE medications. Remitted patients who flared after discontinuing GCs were compared with patients who did not flare. Logistic regression was used to identify predictors of flare and Cox-regression to identify predictors of flare-free survival.

Results Prospectively collected data from 484 patients who achieved remission at least once during follow-up were retrospectively analysed. Three-hundred-eighty patients achieved remission off-GCs and were analysed. During a mean observational time of 87 (±76) months, 48 flares were observed, meaning an annual flare rate of1.65 flare/100 patients/year. At multivariate logistic regression analysis, predictors of flares after GC withdrawal were low C3 levels (OR 0.007, CI 95% 0.00–0.188, p=0.007), arthritis (3.108, 1.096–8.811, p=0.033), leukopenia (2.146, 1.030–4.472, p=0.041), vasculitis (2.650, 1.037–6.773,p =0.042), and remission duration (0.987, 0.980–0.995, p<0.001). By Cox regression analysis, predictors of shorter flare-free remission were thrombocytopenia (HR 2.446, CI 95% 1.106–5.410, p=0.027), vasculitis (3.033, 1.262–7.432, p=0.013), disease duration (0.943, 0.892–0.998, p=0.054), and positive anti-U1RNP (1.973, 0.988–3.940, p=0.054).

Conclusion According to our results, remission off-GCs is an achievable outcome in SLE, especially in patients without serological activity and in those with stable remission.

P74 LATE-ONSET MONOGENIC LUPUS: A CASE-BASED REVIEW

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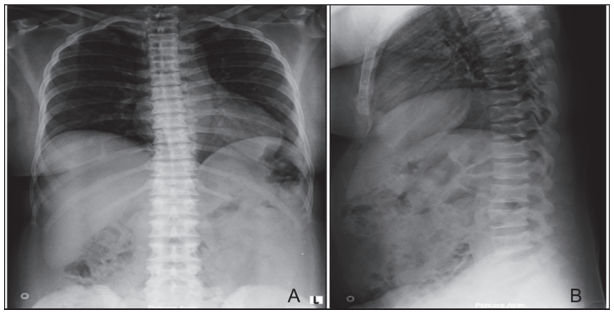
Objective To highlight Mendelian inheritance in adult-onset systemic lupus erythematosus with a unique case of spondyloenchondrodysplasia-immune dysregulation and explore the potential of baricitinib as a promising treatment.

Methods We present a case of spondyloenchondrodysplasia-immune dysregulation in an adult patient, born to nonconsanguineous parents. The patient was diagnosed with spondyloenchondrodysplasia-immune dysregulation and exhibited immune neutropenia, anti-dsDNA positivity, platyspondyly (figure 1), immune deficiency, and a homozygous variant (c.155A > C, p.Lys52Thr) in the ACP5 gene, previously classified as pathogenic. Baricitinib, a Janus kinase inhibitor known for its potential efficacy in managing interferonopathies like spondyloenchondrodysplasia-immune dysregulation, was initiated after the diagnosis.



Abstract P74 Figure 1 Patient's vertebral imaging on admission showed platyspondyly as seen in (A) and (B). This condition is characterized by flattened vertebral bodies throughout the axial skeleton and is a radiographic indication of spondyloenchondrodysplasia

Results Systemic lupus erythematosus typically manifests as a multifactorial disease in adulthood, with monogenic forms contributing to 10–25% of cases in childhood. Spondyloenchondrodysplasia, a rare monogenic lupus subtype, has been reported in only 22 patients to date, the majority of whom were diagnosed during childhood, except for one case diagnosed at 19. Our case is unique as it displayed the complete clinical spectrum of monogenic systemic lupus erythematosus at 35 years of age, deviating from the typical age of presentation described in the literature. During follow-up, our patient achieved successful clinical management through the initiation of baricitinib treatment.

Conclusion Spondyloenchondrodysplasia-immune dysregulation represents an uncommon cause of systemic lupus erythematosus in adulthood. Clinicians should be vigilant of underlying Mendelian inheritance when encountering patients with associated immunodeficiency, skeletal abnormalities, and neurological issues. Although current treatment modalities for Mendelian and non-Mendelian systemic lupus erythematosus are similar, Janus kinase inhibitors, such as baricitinib, hold promise as a viable treatment option for monogenic systemic lupus erythematosus cases exhibiting an interferon signature.

P75 MOLECULAR EXPLORATION OF SYSTEMIC LUPUS ERYTHEMATOSUS (SLE) SEXUAL DIMORPHISM UNCOVERS A CRUCIAL ROLE OF SMC1A IN TRANSCRIPTIONAL REGULATION OF INFLAMMATION

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