Successful treatment of SAPHO syndrome with apremilast


Synovitis, acne, pustulosis, hyperostosis and osteitis (SAPHO) syndrome is a rare disease with inflammatory osteoarticular and skin involvement. The pathogenesis of SAPHO syndrome remains unclear, but evidence suggests it may be an autoinflammatory disease triggered upon exposure to infectious agents in genetically predisposed individuals. Induction of the IL-23/Th17 axis as well as neutrophil activation seem to play a key role, and therapies targeting these immunological pathways, including TNF-inhibitors, ustekinumab, secukinumab and the IL-1 inhibitor anakinra are potential treatment options that need further investigation. Here we report a case of a 24-year-old woman suffering from SAPHO syndrome who presented at our clinic with palmoplantar pustulosis and sternoclavicular joint involvement. Previous treatments with topical steroids and keratolytics combined with NSAIDs, intravenous methylprednisolone, methotrexate and salazopyrin had all failed to improve symptoms. Therapy with etanercept was not tolerated, and due to a previous demyelinating peripheral neuropathy, further treatment with TNF inhibitors was avoided. We initiated ustekinumab 45mg, which improved skin manifestations but not joint pain. Dose escalation to 90mg initially improved joint pain, but the dose had to be reduced to 45mg again due to increased infections. During subsequent 45mg ustekinumab treatment joint pain exacerbated so we switched to secukinumab, which improved skin and joint symptoms significantly but was associated with a pustular hypersensitivity reaction. Finally, we began treatment with apremilast, a pan-cytokine approach, resulting in stabilization of the skin and joint symptoms without side effects. To our knowledge, this is the first case report of apremilast as a treatment for SAPHO syndrome. This article is protected by copyright. All rights reserved.