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BIOLOGICS IN A VERY REFRACTORY SAPHO SYNDROME CASE

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BACKGROUND

The aim of this study is to report a case of synovitis-acne-pustulosis-hyperostosis-osteitis (SAPHO) syndrome that was refractory to conventional treatment and more than two classes of biologics.

CASE REPORT

A 42-year-old woman from the north of Brazil was referred to the rheumatology clinic in 2015 because of severe back and parasternal region pain, particularly in the upper part of her trunk, that were refractory to nonsteroidal anti-inflammatory drugs (NSAIDs). She had a history of acne during adolescence and recurrent pustulosis in hands and feet. She denied lumbar or sacroiliac joint (SIJ) pain and there were no gastrointestinal or ocular complaints. In the first visit, the main diagnostic hypothesis was SAPHO syndrome or an axial form of psoriatic arthritis. She returned 15 days later with some results: erythrocyte sedimentation rate of 9 mm/h, C-reactive protein of 13.2 mg/L (normal: until 5 mg/L). Magnetic resonance imaging (MRI) of thoracic spine revealed osteitis in vertebral bodies from T2 to T6 (Fig. 1). Besides, there were no bone marrow edema or structural lesions in sacroiliac joint MRI. Bone scintigraphy showed the characteristic bull's head sign (Fig. 2).

After these results, SAPHO syndrome was diagnosed and naproxen 550 mg BID and endovenous zoledronic acid 5 mg were prescribed. After 3 months, there was just partial response and we decided to start sulfasalazine 1.5 g. Less than 3 months later, the pain was much more intense and antitumor necrosis factor (anti-TNF) etanercept was indicated. As soon after the first two doses, she had an important worsening of her pain and pustulosis. The drug was interrupted and prednisone 20 mg with methotrexate 15 mg were started. Because of paradoxical effect of the anti-TNF, we decided to try secukinumab. She maintained the drug (dose augmented until 300 mg) for 8 months. However, she was never free of prednisone. Secukinumab was suspended and adalimumab was prescribed, but she had no response after 4 months. Because of anemia and persistent plaquetosis, we decided to try tocilizumab 8 mg/kg. After more 6 months of treatment, she was still taking prednisone 10-15 mg every day. Because of pain in her back and costosternal joints were persistent, we decided to try another anti-interleukin 17. Ixekizumab 80 mg was prescribed and zoledronic acid was repeated. After more than 6 months of treatment, she was finally free of corticosteroids.

Figure 1. STIR sequence showing corner inflammatory lesions (red arrows) in thoracic spine.

Figure 2. Bone scintigraphy showing the characteristic bull's head sign (red arrow).

CONCLUSION

There are refractory SAPHO patients, and anti-IL17 seems to be an option even for those that were refractory to anti-TNF and IL-6 drugs.