A complex case of vulval pyoderma gangrenosum

A 53-year-old female presents with a 2-year history of recurrent deep vulval fissures and ulcerations that subsequently spread to the natal cleft, inframammary and inguinal folds. These were associated with severe pain, and a significant social and psychological impact on the patient. Multiple vulvae biopsies showed ulcerating chronic inflammation with focal non-suppurative, non-caseating granuloma formation. Infectious causes were excluded. CT enterography, colonoscopy and gastroscopy were performed to exclude Crohn's disease. An age appropriate malignancy screen was normal. A diagnosis of mixed vulval pyoderma gangrenosum and genital hidradenitis suppurativa was made.

The deep vulvoperineal ulcerations have been exquisitely responsive to both oral and intravenous pulse corticosteroids with reduction in pain and size, however, this has been complicated by steroid-induced vertebral and hip fractures despite bone prophylaxis, requiring multiple vertebroplasties. Multiple other therapeutics modalities have been trialled without success (or adverse effect), including various antibiotics, spironolactone, cyproterone acetate, colchicine, zinc, cyclosporine, mycophenolate mofetil, azathioprine, methotrexate, IVIG, ustekinumab (IV and SCT), infliximab (improvement noted but as with adalimumab, was complicated by severe palmoplantar pustulosis and generalised guttate psoriasis) and anakinra (complicated by pustular eruption at injection site and no clinical effect after 28 days). Thalidomide was complicated by urticarial and morbilliform eruption, even on rechallenge.

There has been no healing of the existing painful deep ulcerations. Current treatment includes ustekinumab, dapsone, metronidazole and ciprofloxacin. She continues to be a challenging and complex case.