

Pyoderma gangrenosum as a Rare Complication of
Chronic Recurrent Multifocal Osteomyelitis successfully treated with
Upadacitinib as steroid-sparing agent

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Background

Pyoderma gangrenosum (PG)

- rare, inflammatory skin disorder,
- particularly uncommon in childhood
- associated with chronic inflammatory conditions, most notably inflammatory bowel diseases, rheumatoid arthritis and myelodysplastic disorders.
- Treatment options in adults: corticosteroids and biologics such as TNF inhibitors and others¹

Case Report

- 11 years old girl
- complicated and intermittently therapy-refractory CRMO
- Extraosseous manifestations with severe psoriasis and arthritis
- several treatments
- Development of pyoderma gangrenosum under treatment with the IL-12/23 inhibitor Ustekinumab
- significant bone and joint pain: Assessment of disease activity/pain 7/10 (visual analogue scale), multiple skin ulcerations in addition to her pre-existing psoriasis, elevated inflammatory markers, and MRI findings indicative of active multifocal osteomyelitis.
- Introduction of methotrexate (MTX) and upadacitinib, a Janus kinase (JAK) inhibitor, as bridging agent
- dramatic improvement within a few days after initiating Upadacitinib without any worsening of the underlying osteomyelitis or psoriasis.
- no infections or other side effects during upadacitinib treatment.
- No flare of psoriasis or pyoderma gangraenosum so far
- Osteomyelitis still active in last MRI (although improvement), without any clinical pain.

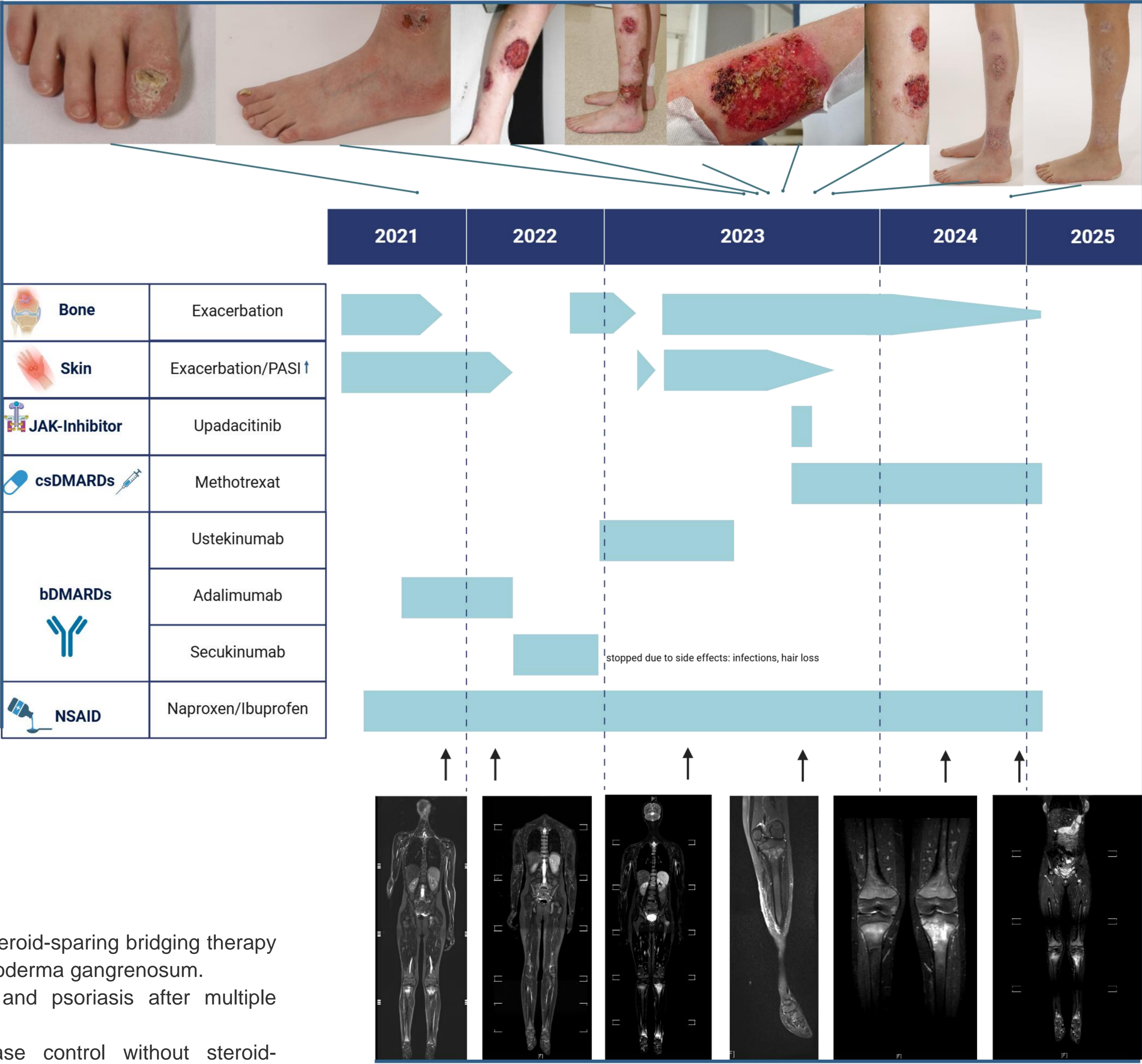
Comprehensive diagnostic workup, including evaluation for underlying genetic disorders, immune deficiencies, and malignancies, yielded no significant findings.

Conclusion

- This case demonstrates
- the successful off-label use of upadacitinib as a steroid-sparing bridging therapy in a pediatric patient with refractory CRMO and pyoderma gangrenosum.
 - Prompt stabilization of both her osteomyelitis and psoriasis after multiple disease flares
 - Upadacitinib treatment showed effective disease control without steroid-depending rebound.
 - While upadacitinib is not yet approved for use in children under 12 years of age, this case highlights the potential of JAK inhibitors as a valuable therapeutic option in complex cases of autoinflammatory diseases with extraosseous manifestations and complications.
 - Patient assessment of disease activity/pain dropped to zero (visual analogue scale), while no complete remission of the osteomyelitis was achieved via MRI.

Chronic Recurrent Multifocal Osteomyelitis (CRMO)

- autoinflammatory bone disorder
- presents with extraosseous manifestations, including skin conditions like psoriasis
- Management options in severe cases include disease-modifying antirheumatic drugs (DMARDs) like methotrexate or sulfasalazine, TNF inhibitors, especially in cases with extra-articular manifestations as well as bisphosphonates



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