Epidermolytic Ichthyosis Improvement with the Combination of Belatacept and Acitretin

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Introduction

- Epidermolytic ichthyosis (EI) is a rare, autosomal dominant keratinization disorder characterized by scaling, desquamation, and blistering.
- Monoclonal antibodies have been repurposed as a treatment for EI alongside standard therapies, such as oral retinoids.
- IL-17, IL-12, IL-23 and IL-4 receptor inhibitors have been reported to be efficacious in EI.

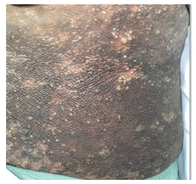
Case Presentation

- A 46-year-old woman with a history of epidermolytic ichthyosis and a deceased donor kidney transplant (DDKT) presented for a dermatology follow-up visit.
- Seven years prior, she underwent a DDKT and was started on prednisone and mycophenolic acid.
- The same year, a shave biopsy of the right arm confirmed epidermolytic hyperkeratosis and the recommendation was to use emollients.
- One-year post-transplant, due to the development of chronic active antibody-mediated rejection, she began belatacept infusions.
- Five years post-transplant, she was seen in dermatology clinic and started on acitretin 10 mg due to extensive hyperkeratosis on the trunk and extremities despite emollient use (Fig. 1A-C).
- At the dermatology follow-up three years later, she reported significant improvement with minimal ichthyosis.
- Her current medications included prednisone 5 mg daily, mycophenolate 540 mg twice daily, belatacept 5 mg/kg monthly, and acitretin 10 mg daily.
- A review of systems was negative for headache, vision changes, abdominal pain, and arthralgias.
- Examination revealed mild xerosis and slight desquamation of the trunk and bilateral lower extremities (Fig. 2A-C).
- Complete blood count, basic metabolic panel, lipid panel, and hepatic panel labs were all within normal limits.
- The patient was told to continue treatment with acitretin 10 mg daily and frequent use of thick emollient cream.

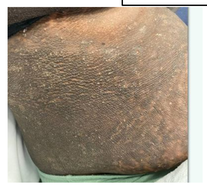
Clinical Photos

Fig. 1 Pre-addition of acitretin, taken on June 6, 2022- Diffuse ridging and hyperkeratosis

1A- Mid to lower back

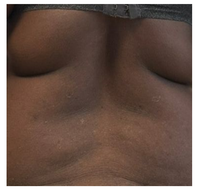


1B- Left Flank

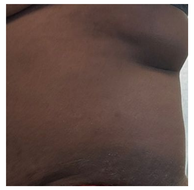


1C- Right flank

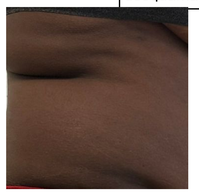
Fig. 2 Post-addition of acitretin, taken on June 9, 2025- Mild xerosis and slight desquamation



2A- Mid to lower back



2B- Left Flank



2C- Right flank

Clinical Timeline

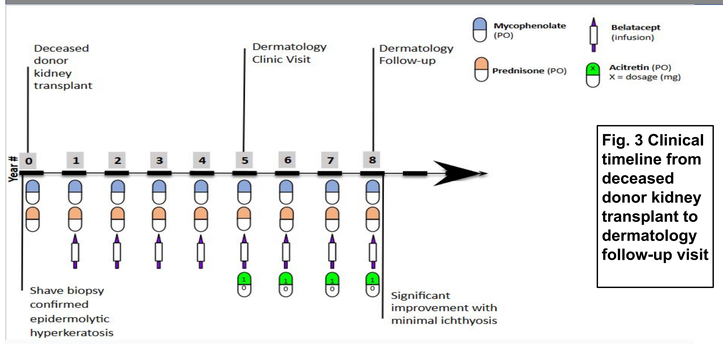


Fig. 3 Clinical timeline from deceased donor kidney transplant to dermatology follow-up visit

Discussion

Traditionally, acitretin has been used individually or in combination with other therapies for EI management. Significant improvement in scaling and hyperkeratosis was common with acitretin, but complete clearance was rare. The recommended acitretin dosage was 0.5 mg/kg/day with careful monitoring. This patient weighed around 80.9 kg and needed a starting dose of 40.45 mg daily; however, she was started on a lower dose of 10 mg daily and had almost complete clearance of her EI.

Additionally, monoclonal antibodies have been repurposed as a treatment for EI alongside acitretin. A 2025 case found that the three-month use of anti-IL-17A monoclonal antibody, vunakizumab, significantly reduced cutaneous keratinization and desquamation in an EI patient. Notably, in this case, the patient was taking belatacept for broad immunosuppression in the setting of her kidney transplant. The mechanism of action of belatacept is to bind CD80 and CD86 on antigen-presenting cells, preventing their interaction with CD28 on T-cells, and overall reducing T-cell proliferation and cytokine production. Potentially, the combination of the belatacept and low-dose acitretin led to the significant clearance of her EI.

Although this case introduces the possibility of a new medication being utilized in the management of EI, there are limitations. This case is limited by the concomitant use of and broad immunosuppression by prednisone and mycophenolate. Due to these additional variables, the effectiveness of the combination of low-dose acitretin and belatacept cannot be clearly defined. Future studies that evaluate the efficacy and safety profile of low-dose acitretin and belatacept in the management of EI are needed to come to a definitive conclusion.

Conclusion

This report serves to increase awareness of the combination of low-dose acitretin and belatacept as a potential therapy in the management of EI. However, due to the concomitant immunosuppression with prednisone and mycophenolate, future studies are needed to elucidate this finding.

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Acknowledgements

- **Special acknowledgement to the Department of Dermatology at the Virginia Commonwealth University Health System.**

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