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A Rare Case of Bullous Eruptive Disseminated Porokeratosis

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History

- A 70-year-old female with a history of immunosuppression secondary to kidney transplant and disseminated superficial actinic porokeratosis (DSAP) presented for evaluation of a new blistering eruption on her lower extremities.
- She noted the abrupt onset of blisters and increased bilateral leg swelling one month prior to presentation during a hospitalization for herpes zoster ophthalmicus.
- The blistering eruption was unresponsive to high-dose furosemide, thrombo-embolic-deterrent compression stockings, and topical clobetasol 0.05% cream.

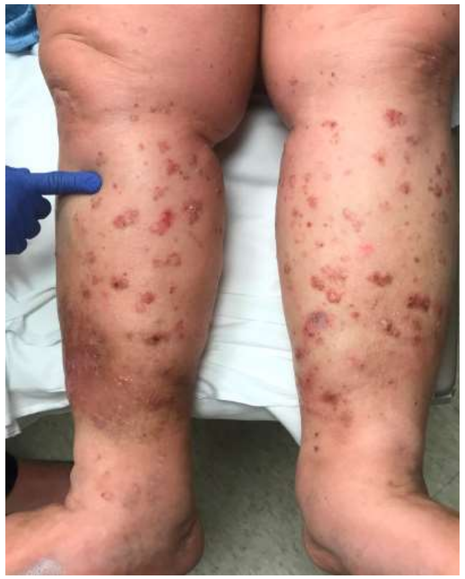
Examination

- Physical exam revealed numerous irregularly shaped, red-brown macules surrounded by a keratotic rim of scale, clinically consistent with porokeratosis, overlying the thighs, shins, and ankles bilaterally
- Within many of these characteristic porokeratosis lesions, tense vesicles and small bullae containing clear fluid were noted
- Lower extremity pitting edema to the knees bilaterally

Course and Therapy

- History and exam findings led to a diagnosis of eruptive disseminated porokeratosis (EDP) with the unusual feature of bulla formation in the setting of peripheral edema.
- Leg compression and elevation, medical management of multifactorial leg edema, and cessation of clobetasol application were advised, and the bullous EDP improved at follow up.

Clinical Photos



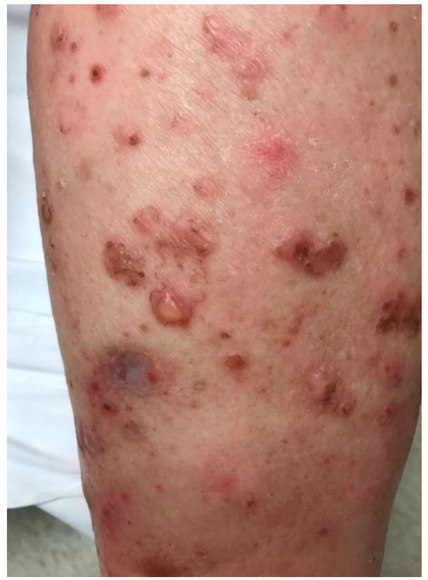


Figure 1 (left). Numerous irregularly shaped, red-brown macules surrounded by a keratotic rim of scale, clinically consistent with a disseminated eruption of porokeratosis.
Figure 1 (right). Tense vesicles and small bullae containing clear fluid were confined within many of the characteristic porokeratosis lesions.

References

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Discussion

- The term eruptive disseminated porokeratosis (EDP) has been used to describe all cases of acute, disseminated eruptions of porokeratosis. In a review of 35 patients with EDP, the presence of preexisting lesions of porokeratosis (especially DSAP) had been noted in about 30% of patients, and dissemination appeared to represent an inflammatory flare.[1] In our case, human herpesvirus 3 (HHV-3) reactivation was considered to be a potential trigger.
- While immunosuppression likely contributed to this patient's initial development of DSAP following kidney transplantation,[2] the HHV-3 reactivation of her concurrent herpes zoster ophthalmicus may have triggered the abnormal clones of keratinocytes within DSAP lesions to further proliferate and manifest as EDP. Although HHV-3 has never been directly implicated in the development of porokeratosis, herpes simplex virus infection has been reported as a possible trigger of EDP.[3]
- EDP with secondary bullous changes is an exceptionally rare variant with only two cases reported in the literature to date.[4,5]
- The bullous changes seen in our patient are suspected to be secondary to underlying edema and the localized extravasation of interstitial fluid in areas affected by porokeratosis. We postulate that the abnormal keratinocytes affected by porokeratosis failto provide adequate junctions between adjacent keratinocytes, allowing localized extravasation of interstitial fluid within the sites of reduced skin integrity.
- In summary, we report an unusual presentation of bullous EDP, likely triggered by herpes zoster ophthalmicus, in the setting of acute-on-chronic edema in a renal transplant recipient on systemic immunosuppression.
- Clinicians should be aware that EDP may present with bullous changes, particularly in the setting of edema.