IMMUNOMODULATORY THERAPY FOR IDIOPATHIC RECURRENT ERYTHEMA MULTIFORME: A CASE REPORT

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Background: Erythema multiforme (EM) is an immune-mediated condition characterized by targetoid skin lesions. The EM major subtype involves mucous membranes and associated systemic symptoms. The vast majority of cases are self-limited; a small proportion can recur or persist impacting patient quality of life (QoL). In cases where a trigger is not clear, management can be challenging.

Materials/Methods: We report the presentation, management, and outcomes of a patient with idiopathic recurrent EM major.

Results: A 16 year old female with no significant past medical history or medication exposures presented with a rash associated with oral and genital pain. On exam, she had dusky bullous lesions surrounded by erythematous halos on the palms and web spaces of the feet. She also had exquisitely painful oral ulcers and erosions of her oral and genital mucosa interfering with PO and voiding. She reported 2 similar episodes within the previous year that self-resolved. Herpes simplex virus and Mycoplasma pneumoniae testing were negative. There was no serologic or clinical suspicion of systemic lupus erythematosus, although anti-nuclear antibodies were detected (1:160). Her symptoms resolved with supportive care. However, 3 months later, she experienced a fourth recurrence. Interestingly, the skin lesions recurred in the same sites each flare. Skin biopsy showed non-specific chronic inflammation without evidence of vasculitis, mast-cell reaction, or local infection. She was diagnosed with EM major based on clinical morphology of the lesions with associated mucosal involvement. The frequent recurrences were significantly impacting her QoL. The lack of an identified trigger, either infectious or other exposure, suggested immune dysregulation and she was started on azathioprine. Azathioprine prevented recurrences during periods of medication adherence. However, 3 subsequent flares requiring admission for pain management and hydration occurred in the setting of medication non-adherence. Her skin lesions continue to appear in the same locations with each recurrence.

Conclusions: This case illustrates an unusual presentation of recurrent EM major, where skin lesions recur in the same locations with each flare. It also highlights that this condition can have a significant impact on QoL, so that, while management can be challenging in idiopathic cases, immune modulation should be pursued. Our experience suggests that azathioprine can help prevent recurrences and improve patient well-being in recurrent EM major.

Images / Graph / Table
Figure 1. Oral mucosal lesions and palmar and toe-web skin lesions at presentation