

Butterfly Rash

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A 32-year-old married Indian woman on treatment for hypothyroidism presented with a photosensitive rash associated with a burning sensation over the cheeks for the preceding two months. Notably, she also complained of flushing before the present symptoms. Her symptoms aggravated on taking spicy food and alcoholic beverages. The facial rash was not associated with any history of

fever, arthralgia, proximal muscle weakness, or other features suggestive of systemic involvement. She denied any history of miscarriage or topical steroid application. An indurated, erythematous, mildly edematous plaque studded with multiple inflammatory papules, tiny telangiectatic vessels, and pustules involving bilateral cheek, chin, and bridge of the nose but sparing the nasolabial folds was observed [Figure 1]. There was no oral ulceration. Other mucocutaneous sites were unaffected. Systemic examinations were within normal limits. Routine laboratory investigations were notable for mildly elevated erythrocyte sedimentation rate (19 mm/h; reference range 0–15 mm/hour). Anti-nuclear antibody testing was negative. Urinalysis did not reveal any abnormalities.



Figure 1: Erythematous plaque with superimposed papules and pustules over malar prominences, sparing nasolabial furrows. The presence of inflammatory papules and pustules was also observed over the chin.

Question

1. What is the most likely diagnosis?
 - a. Systemic lupus erythematosus (SLE).
 - b. Dermatomyositis.
 - c. Papulopustular rosacea (PPR).
 - d. Seborrhoeic dermatitis.
 - e. Erysipelas.

Answer

- c. Papulopustular rosacea (PPR).

Based on the above findings, a diagnosis of PPR was established. Pustulations and erythema subsided with judicious sunscreen use, topical metronidazole gel, and oral doxycycline (100 mg twice-daily) within a fortnight.

DISCUSSION

The papulopustular subset of rosacea is characterized by fixed centropacial erythema surmounted by

multiple erythematous and pinpoint pustules. The initial telangiectasia usually gets obscured by persistent erythema. This can be associated with facial flushing (triggered by spicy food, heat, and alcohol), non-pitting facial edema, and symptoms of burning and stinging sensations. Ocular symptoms like dryness, gritty sensation, crusting of eyelid margins, and frequent styes may sometimes be associated, although extrafacial involvement is rarely encountered. The condition commonly affects middle-aged women. Abnormal vasomotor response to thermal and other stimuli, dysregulation of the innate immune system and neurovascular control have been documented in the pathogenesis, although the exact cause remains unknown. Commensal microbes of the pilosebaceous unit, *Staphylococcus epidermidis*, and *Demodex* mites may also act as triggers. Apart from the papulopustular variety, other clinical variants of rosacea include erythematotelangiectatic, phymatous, and ocular rosacea.^{1,2}

Malar rash is a typical facial presentation of multiple disorders. SLE is an autoimmune disease that involves multiple organs in which the immune system produces numerous autoantibodies that attack different tissues such as the kidneys, joints, skin, brain, and heart. The stereotypical malar rash is the prototype of acute cutaneous lupus erythematosus. This is characterized by a flat or raised fixed erythema over the malar prominences that spares the nasolabial folds in SLE. As per the Systemic Lupus International Collaborating Clinics criteria, the malar rash is one of the criteria for SLE.³ Dermatomyositis is a disease of presumed autoimmune pathogenesis that presents with a symmetric, proximal, extensor inflammatory myopathy and characteristic cutaneous eruption (photodistributed pink-violet poikiloderma favoring the periocular region (heliotrope rash), chest (V-sign) and upper back (shawl sign), extensor surfaces (Gottron papules), and distinctive nail fold changes).¹ Seborrheic dermatitis is characterized by a red greasy scaling rash on the scalp, face involving

the nasolabial fold, and torso.⁴ Erysipelas is a skin infection that affects the superficial lymphatics and upper dermis. The classic manifestation includes a painful, rapidly progressive, well-circumscribed, shiny, erythematous plaque accompanied by edema. Systemic symptoms like fever, chills, and malaise are usually present.⁵

Strict photoprotection, avoidance of exacerbating factors (stress, alcohol, and spicy food), and specific measures like topical and/or systemic antibiotics show excellent response. Topical ivermectin (1% cream) is a recently approved treatment modality for PPR that has anti-inflammatory and antiparasitic activities. At the initial consultation, the patient should be made aware of the chronic relapsing nature of the condition and the need for maintenance therapy even when in remission.^{1,2}

PPR can seriously impair the quality of life of those affected. Therefore, a prompt clinical diagnosis is essential to obviate the need for any unnecessary investigative work-up and thereby limit the economic burden on patients.

Disclosure

The authors declared no conflicts of interest. Written consent was obtained from the patient.

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