Butterfly Rash

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Case description

A 32-year-old married Indian lady, on treatment for hypothyroidism, presented with photosensitive rash associated with burning sensation over the cheeks for the preceding 2 months. Notably, she also complained of flushing prior to the present symptoms. Her symptoms aggravated on taking spicy food and alcoholic beverages. The facial rash was not associated with any history fever, arthralgia, proximal muscle weakness, or other features suggestive of systemic involvement. She denied any history of miscarriage or topical steroid application. On examination, an indurated, erythematous, mildly oedematous plaque studded with multiple inflammatory papules, tiny telangiectatic vessels, and pustules involving bilateral cheek, chin and bridge of nose but sparing the nasolabial folds was observed. [Fig. 1] There was no oral ulceration. Other mucocutaneous sites were unaffected. Systemic examinations were within normal limits. Routine laboratory investigations was notable for mildly elevated erythrocyte sedimentation rate (19 mm/hr; reference range 0-15 mm/hour);

Anti-nuclear antibodies testing was negative. Urinalysis did not reveal any anbormalities.

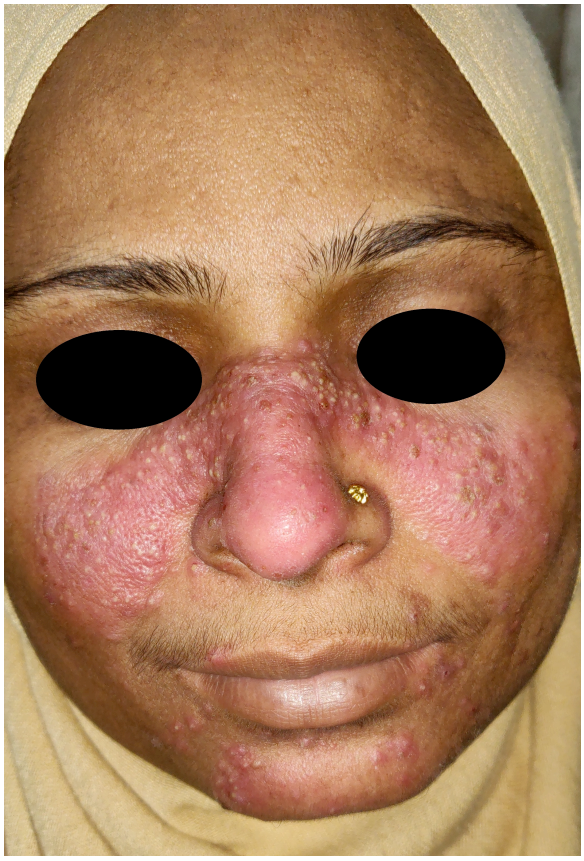

Figure 1: Erythematous plaque with superimposed papules and pustules over malar prominences, sparing nasolabial furrows; presence of inflammatory papules and pustules also observed over the chin.

Question

What is the most likely diagnosis?

A. Systemic lupus erythematosus (SLE)

B. Dermatomyositis

C. Papulo-pustular rosacea

D. Seborrhoeic dermatritis

E. Erysipelas

Answer:

C. Papulo-pustular rosacea.

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

Discussion:

The papulopustular subset of rosacea is characterized by fixed centrofacial erythema surmounted by multiple erythematous papules and pinpoint pustules. The initial telangiectasia usually gets obscured by the persistent erythema. This can be associated facial flushing (triggered by spicy food, heat, alcohol) and non pitting facial oedema, along with symptoms of burning and stinging sensation. Ocular symptoms like dryness, gritty sensation, crusting of eyelid margins, frequent stye may be associated at times, 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 exact cause remains unknown. Commensal microbes of pilo-sebaceous 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 auto-antibodies that attack different tissues such as 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 systemic lupus erythematosus. As per the Systemic Lupus International Collaborating Clinics (SLICC) criteria, 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 favouring the periocular region (heliotrope rash), chest (V-sign) and upper back (shawl sign), extensor surfaces (Gottron papules), and distinctive nail fold changes). [1] Seborrhoeic dermatitis is characterized by red greasy scaling rash on scalp, face involving the nasolabial fold, and torso. [4] Erysipelas is a type of skin infection that involves the superficial lymphatics and upper dermis. The classic manifestation includes a painful, rapidly progressive, well-circumscribed, erythematous shiny plaque accompanied with edema. Systemic symptomis 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 maintenence therapy even when in remission. [1, 2]

PPR can seriously impair the quality of life of those affected. A prompt clinical diagnosis is essential to obviate the need for any unnecessary investigative workup and thereby limit the economic burden on patients.

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