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Case presentation

Possible photoactivated dermatitis with features of post-inflammatory pigmentary alteration (PIPA) and rosacea

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Abstract

Cutaneous flushing and facial erythema are common dermatologic conditions that elicit a wide differential diagnosis that includes rosacea, seborrheic dermatitis, photodermatitis, connective-tissue diseases, carcinoid syndrome, and mastocytosis. Herein we present an unusual case of a mask-like rosacea-PIPA overlap that occurred in a patient with prior history of rectal carcinoid tumor and a negative systemic evaluation.

Case synopsis

History: A 48-year-old woman with prior history of a resection of a carcinoid tumor of the rectum was referred to the Skin and Cancer Unit by her primary dermatologist for further evaluation of facial erythema with mild flushing that had been present for at least two years. Her dermatologist first noticed changes to her face incidentally on a routine skin examination. The patient stated that her face felt full at times and she sometimes experienced mild flushing and burning around the time of her menstruation. She noted over the past two years that others frequently commented that she appeared more youthful with fewer fine lines. She denied any procedures to the face, such as chemical peels, lasers, or the use of topical medications. The patient denied wearing makeup and used Cetaphil cleanser to wash her face; she used a sunscreen periodically. She denied photosensitivity or worsening of the condition with sun exposure, spicy foods, or alcohol. After having spinal fusion surgery to correct cervical disc disease, the patient began to note generalized fatigue and aches. She underwent rheumatologic and neurologic evaluations and was initially diagnosed with undifferentiated connective-tissue disease owing to a positive anti-nuclear antibody (ANA) of 1:80 with a borderline pattern. However, repeat ANA was negative and no objective inflammatory signs were found. Trials of methotrexate, sulfasalazine, prednisone, and non-steroidal anti-inflammatory drugs had no effect on her aches or facial erythema. A punch biopsy was obtained of the skin overlying the left mandible.

Physical examination: On the face there is a red-brown, faintly edematous, mask-like plaque that spares the orbits with sharp demarcation at the edges of the face with scattered, prominent, small telangiectases throughout.

Laboratory Data: A complete blood count, comprehensive metabolic panel, C-reactive protein, erythrocyte sedimentation rate, anticardiolipin, anti-double stranded DNA, anti-Smith, anti-ribonucleoprotein, anti-Ro, and anti-La antibodies; myositis-specific panel; aldolase; myoglobin; anti-cyclic citrullinated peptides; serum protein electrophoresis; hepatitis C antibody; hepatitis B antibody; human immunodeficiency virus antibody; and angiotensin converting enzyme were normal or negative.

The minimum erythema dose to ultraviolet B was normal at 800 mJ/cm² and the minimal erythema dose to ultraviolet A was normal at 30 J/cm². Visible light test was negative. Photopatch tests showed reactions to chlorpromazine hydrochloride and promethazine; these results were thought not to be clinically relevant.

Histopathology: There is a sparse, perivascular and perifollicular, lymphocytic infiltrate with focal vacuolar changes that involve follicular epithelium. There is no appreciable dyskeratosis. A colloidal iron stain shows increased dermal mucin. A periodic acid-Schiff with diastase stain fails to show a thickened basement membrane.



Figures 1 and 2. Sharply demarcated, erythematous facial plaque

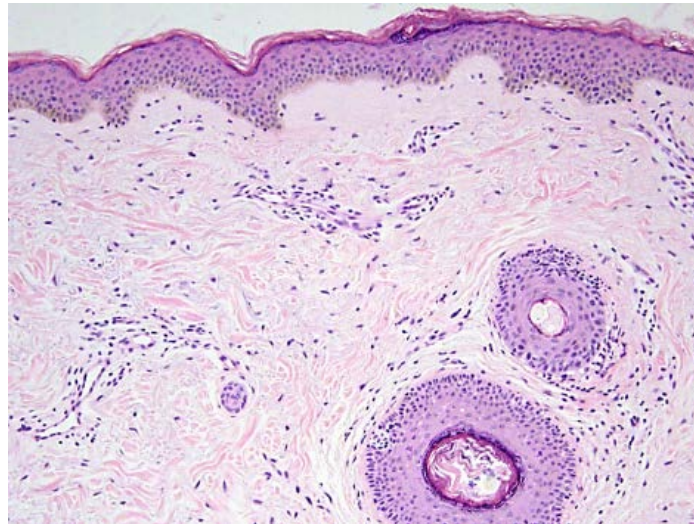


Figure 3. Focal vacuolar changes and dermal mucin

Discussion

Diagnosis: Possible photoactivated dermatitis with features of post-inflammatory pigmentary alteration (PIPA) and rosacea

Comment: Erythema of the face is a common dermatologic complaint, with the differential diagnosis including physiologic or climacteric flushing, rosacea, seborrheic dermatitis, photodermatitis, connective-tissue disease (lupus erythematosus or dermatomyositis), carcinoid syndrome, and mastocytosis [1, 2]. Whereas most patients will not undergo extensive workup for persistent mild facial erythema, it should be considered in selected patients to rule out more serious underlying conditions [1, 2].

Extensive systemic evaluation in this patient was unrevealing as to the trigger of her unique eruption. Although the patient could not recall a specific sun exposure, the location and sharply demarcated borders suggested a photoprecipitated event. Dermatopathologic evaluation suggested both a component of PIPA and rosacea. These findings correspond with the patient's clinical observation that she experiences flushing and burning sensation with specific triggers, such as menstruation.

Photoexposure is known to exacerbate rosacea, but to our knowledge there is no literature to suggest that a precipitating phototoxic or photoallergic dermatitis can incite the disease. Chronic sun exposure contributes to development of the disease through activation of the innate immune system in genetically predisposed individuals [3-6]. In rosacea, the dermal vasculature demonstrates alterations in the blood flow, vasodilatation, and angiogenesis that may lead to permanent erythematous changes [5, 6].

Although our patient's findings were consistent with rosacea and PIPA, her presentation also merited an evaluation for connective-tissue diseases. Unlike rosacea, the facial erythema of lupus erythematosus often has violaceous qualities and often is accompanied by mild edema, exacerbated by photoexposure, and has prominent mucin as a histopathologic feature [7, 8]. Positive antinuclear antibody and anti-Ro/La antibodies usually are observed as well [7, 8]. Reticular erythematous mucinosis, although rarely observed on the face, has many overlapping features with lupus erythematosus but usually lacks autoantibodies [8]. Unlike our patient, the facial erythema of dermatomyositis often includes the eyelids and may share the same violaceous hue as lupus erythematosus.

This patient had a history of rectal carcinoid tumor that preceded the onset of her mask-like eruption. It is unclear what effect her prior condition could have had on her persistent telangiectases and erythema because rectal carcinoid is not thought to elicit carcinoid syndrome [9]. However, the development of fixed facial erythema from vasoactive flushing conditions previously has been noted [1, 9].

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