Pyoderma Faciale

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Pyoderma faciale is a rare cutaneous disorder that predominantly affects women in their 20s and 30s and is characterized by the rapid appearance of coalescing nodules and draining sinuses, combined with livid erythema on the face. We describe a 40-year-old woman who presented with localized pyoderma faciale that worsened during treatment with oral and topical antibiotics and corticosteroids. Subsequent treatment with isotretinoin for 5 months resulted in dramatic and sustained improvement.

Cutis. 2008;81:488-490.

P yoderma faciale (rosacea fulminans) is an uncommon dermatosis characterized by the rapid appearance of coalescing nodules on the face and draining sinuses. The etiology of this disorder is unknown, though it has been considered an extreme form of rosacea.¹ Pyoderma faciale occurs most commonly in women in their 20s and 30s. Recommended therapy consists of a brief course of systemic or high-potency topical steroids followed by isotretinoin.²

Case Report

A 40-year-old white woman with no remarkable medical history was referred for evaluation. Two months prior to her presentation at our office, the patient was seen by another dermatologist for the abrupt appearance of multiple draining pustules with surrounding erythema on her right cheek. The clinical impression was folliculitis, and throughout the following month, the patient was treated with multiple courses of systemic antibiotics including doxycycline, dicloxacillin sodium, and ciprofloxacin hydrochloride, as well as topical antibiotics including clindamycin

Accepted for publication October 2, 2007.

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The authors report no conflict of interest.

and mupirocin. Nodules coalesced to a large plaque, raising concern of a deep fungal infection, and the patient was started on topical ciclopirox olamine and oral fluconazole, without effect. Results of our physical examination 2 weeks later showed a healthyappearing woman with papulopustules overlying a tender, deeply reddish purple, indurated plaque of 5-cm diameter on the right cheek (Figure 1). No other skin lesions were identified. The patient denied any ocular symptoms and refused a skin biopsy.

Pyoderma faciale was diagnosed based on the clinical appearance and history, and the patient was started on 0.8 mg/kg of prednisone (40 mg daily). The decision was made to treat the patient with isotretinoin, and she was educated about the iPLEDGETM qualification criteria for isotretinoin administration. As a female of childbearing potential, she was required to have 2 negative pregnancy tests at least 19 days apart and commit to the simultaneous use of 2 forms of effective contraception for 1 month prior to beginning treatment.³ A screening pregnancy test was negative, and the patient indicated her willingness to use latex condoms for pregnancy prevention. Her spouse had previously undergone a vasectomy.

One week later, the patient complained of worsened swelling of her right cheek, and physical examination showed increased numbers of erythematous nodules coalescing into a plaque, with overlying pustules. Despite continued treatment with prednisone, 2 weeks later, the patient noted new tender firm subcutaneous nodules on her left cheek, which coalesced to a nodular erythematous plaque and progressed to involve her chin (Figure 2). The patient was distressed by the appearance of these lesions and was seen in the emergency department for symptoms of an anxiety attack. Treatment with azithromycin dihydrate was initiated and prednisone was increased to 1 mg/kg (50 mg daily).

A second pregnancy test 1 month after initial presentation to our clinic was negative, and the patient was started on 0.8 mg/kg of isotretinoin (40 mg daily). Within 2 weeks, the patient noted substantial improvement in facial swelling and lesion induration. Monthly physical examinations over the

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Figure 1. Small pustules and nodules on an erythematous base on the patient's right cheek at initial presentation.



Figure 2. Progression of pyoderma faciale to involve both cheeks and the chin during treatment with corticosteroids.



Figure 3. Improvement in induration and acneform lesions with persistent erythema following 8 weeks of treatment with isotretinoin.



Figure 4. Atrophic scarring on left cheek 3 months after discontinuation of isotretinoin.

next several months showed steady improvement and reduction in the induration and acneform lesions (Figure 3). Isotretinoin was discontinued after 5 months. Her skin remained free of acute lesions 3 months after discontinuation of isotretinoin but with atrophic scarring (Figure 4).

Comment

First described by O'Leary and Kierland⁴ in 1940, pyoderma faciale is an uncommon, though almost certainly underreported,⁵ dermatosis that predominantly affects women in their 20s and 30s. Originally thought to be tuberculous in origin, pyoderma faciale was later considered a variant of acne conglobata.⁶ In 1992, Plewig et al¹ suggested that pyoderma faciale was a variant of rosacea, not acne, and they proposed the term rosacea fulminans. This condition is differentiated from acne and acne fulminans on the basis of its fulminating course in the absence of systemic symptoms; principal localization to the central face with sparing of the neck, back, and chest; and absence of comedones. Unlike acne, pyoderma faciale is predominantly a disorder of postadolescent women, though the condition has been reported in a 3-yearold girl⁷ and an adolescent boy.⁸

The etiology of pyoderma faciale is unknown. Hormonal influences have been suggested in light of the condition's almost exclusive occurrence in women; additionally, the eruption has been associated with pregnancy in a few cases.^{1,9-12} The condition also has been associated with inflammatory bowel disease^{9,13-17} and thyroid disease.⁹ High doses of vitamins B₆ and B₁₂,¹⁸ as well as pegylated interferon and ribavirin therapy for hepatitis C virus,^{19,20} have been implicated as triggers. No potential triggers were identified in our patient.

Although pyoderma faciale generally involves much of the central face (ie, forehead, nose, chin, cheeks), it rarely has been observed in a more limited form. Barker and Gould⁵ described 2 women with pyoderma faciale confined to the chin. Massa and Su⁹ reported a localized variant of pyoderma faciale in 4 patients that involved only the cheeks, jawline and chin, or one cheek; they noted that 3 of these 4 patients presented early, having received no previous treatment, and the researchers raised the possibility that this localized form would have spread without rapid therapy. Our case supports the hypothesis of Massa and Su⁹ that a localized form of pyoderma faciale may progress to more widespread disease in the absence of adequate treatment. Haugstvedt and Bjerke¹⁰ reported a case similar to ours in which a 35-year-old woman had spreading of lesions during treatment with oral prednisolone. Most reports describe a positive response to treatment with systemic corticosteroids.^{1,18} However, we observed exacerbation of disease during treatment with oral prednisone.

Many medications have been used in the treatment of pyoderma faciale, with varying success. In 1987, Cunliffe and Rowell²¹ reported successful treatment of pyoderma faciale with a 5-month course of isotretinoin at a dosage of 1 mg/kg daily. On the basis of their experience treating 20 patients with pyoderma faciale, Plewig et al¹ later proposed that a short course of topical and/or systemic corticosteroids followed by oral isotretinoin is the treatment of choice.

Conclusion

We present a case of pyoderma faciale that began as a localized eruption and was unresponsive to treatment with multiple medications. Substantial worsening of the disease was observed despite treatment with corticosteroids during the iPLEDGE qualification period. This disfiguring disorder may cause substantial psychological distress, and, although cosmetic outcome usually is favorable, patients may have substantial permanent scarring. Early treatment of pyoderma faciale may help to minimize scarring.²² Our patient ultimately responded well to treatment with isotretinoin.

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