# Granulomatous Periorificial Dermatitis

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GOAL

To gain an understanding of granulomatous periorificial dermatitis (GPD)

OBJECTIVES

Upon completion of this activity, dermatologists and general practitioners should be able to:

1. Discuss the presentation of GPD.
2. Describe the differential diagnosis for GPD.
3. Explain the treatment of GPD.

CME Test on page 398.

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Ms. Tarm and Drs. Creel, Krivda, and Turiansky report no conflict of interest. The authors report no discussion of off-label use. Dr. Fisher reports no conflict of interest.

*Granulomatous periorificial dermatitis (GPD) is a distinct facial eruption in prepubertal children that should be distinguished from granulomatous rosacea, perioral dermatitis, and cutaneous sarcoidosis. We describe a case of GPD and review the key distinguishing features of this condition.*

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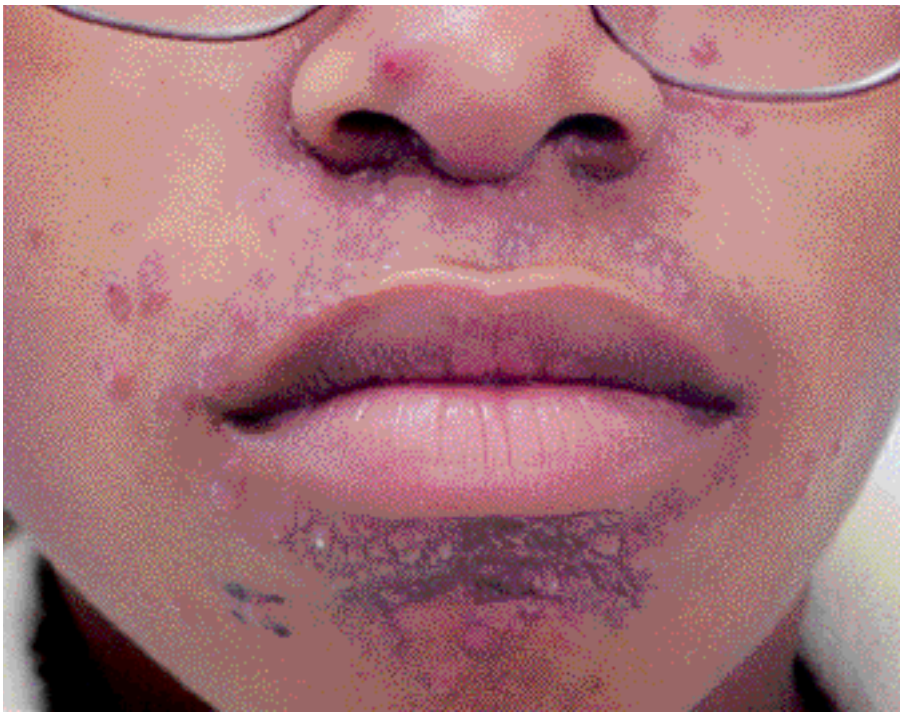
## Case Report

A healthy 12-year-old African American boy presented with a 3-month history of an asymptomatic, papular, perioral eruption. He had been previously treated by a primary care provider, who had prescribed a low-potency and then a medium-potency topical corticosteroid, without improvement. The patient's parents denied that he had used topical corticosteroids or other topical preparations prior to the eruption. He took no oral medications, and the findings of the review of systems were within reference range. Results of a physical examination



Numerous firm papules coalescing in a perioral distribution.

revealed numerous, discrete and coalescing, firm, pink, hyperpigmented papules ranging from 2 to 4 mm in diameter, primarily located in the perioral area (Figure). A few papules were noted around the nose and eyes. Results of a shave biopsy of a perioral papule revealed dermal granulomatous inflammation. The infiltrate consisted of histiocytes and lymphocytes and also included several focal collections of neutrophils. There was overlying parakeratosis. Results of a periodic acid–Schiff, Gomori methenamine-silver, Brown-Brenn, and Fite-Farraco stains were negative for organisms.

The patient was diagnosed with granulomatous periorificial dermatitis (GPD), and treatment with oral tetracycline 500 mg twice a day was initiated. Six days later, a tapering course of oral prednisone was added (consisting of 40 mg for 3 days, 30 mg for 3 days, 20 mg for 3 days, and 10 mg for 3 days) because of the severe extent of the eruption and the social distress it was causing the patient. Within 3 weeks, a dramatic decrease in the eruption was noted, with only a few small papules remaining. The patient was then switched from oral tetracycline to topical clindamycin twice a day for maintenance, but he did not return for follow-up.

## Comment

GPD is characterized by a monomorphic papular eruption occurring in the perinasal, perioral, and periocular areas. Gianotti et al[1] first described the condition in 5 children, ranging in age from 2 to 7 years, who had asymptomatic, distinctive, flesh-colored, "micronodular," periorificial eruptions. In the literature, the condition has been variably called *Gianotti-type perioral dermatitis*, *sarcoidlike granulomatous dermatitis*, *facial Afro-Caribbean childhood eruption*, and *childhood granulomatous perioral dermatitis*. It has been proposed that GPD is a form of perioral dermatitis with granulomatous histologic features.[2,3] GPD typically affects prepubertal children, predominantly African Americans and others with dark skin. Several cases involving fair-skinned children have been reported.[2] Both genders are equally affected. Typical lesions have been described as flesh-colored, yellow-brown, or red papules or micronodules.[2,3] Slight scaling of lesions or surrounding erythema may occur.[2] Scarring is absent in most cases; however, pinpoint atrophy or scarring occasionally occurs.[2] In addition to the characteristic facial distribution, extrafacial and generalized lesions on the trunk, extremities, and labia majora have been described.[2] In all reported cases, the lesions, including extrafacial lesions, were histologically similar and self-limited and were not accompanied by associated systemic symptoms. The presence of extrafacial lesions did not affect the duration of disease or the response to treatment.

The etiology of GPD is unknown. It may represent an unusual inflammatory granulomatous

response to allergens.[3] The initial allergen may cause inflammation and a focal disruption of the follicular wall, inciting a granulomatous reaction.[3] In 1978, Georgouras and Kocsard[4] described a case of Gianotti-type perioral dermatitis as an unusual reaction to bubble gum. Other reports have implicated reactions to formaldehyde, cosmetic preparations, and antiseptic solutions.[5] Topical corticosteroids may induce or exacerbate both perioral dermatitis and GPD.[3,6]

Histologic findings consist of upper dermal and perifollicular granulomas admixed with lymphocytes. The inflammation surrounding a focally disrupted hair follicle may range from a primarily lymphocytic inflammation with focal granuloma formation to a denser dermal granulomatous infiltrate. The presence of lymphocytic inflammation can help distinguish GPD from the "naked" granulomas in cutaneous sarcoidosis that typically lack inflammatory cells. GPD also may show epidermal change with mild to moderate spongiosis.[3,7]

The clinical differential diagnosis of granulomatous papules on the face of a child includes GPD, granulomatous rosacea, lupus miliaris disseminatus faciei, and cutaneous sarcoidosis. Other cutaneous diseases that may occur in a periorificial distribution include perioral dermatitis, telangiectatic fibromas, and trichoepitheliomas. Although similar in distribution to GPD, the primary lesions in these latter 3 conditions are not granulomatous and can often be distinguished clinically. Deep fungal infection, atypical mycobacterial infection, and leishmaniasis can be considered in the histologic differential diagnosis of granulomatous dermatitis, but these conditions usually present as nodules, plaques, or ulcers and not necessarily in a perioral distribution.

Granulomatous rosacea and GPD have a similar clinical and histopathologic presentation. Both conditions can present as red or yellow-brown, dome-shaped facial papules with histologic findings of a perifollicular lymphohistiocytic or granulomatous infiltrate. The major distinguishing features of granulomatous rosacea are erythema, telangiectasias, pustules, and edema.[8] Granulomatous rosacea is not characterized by a concentration of lesions in the perioral area, and it is most common in 30- to 50-year-old women.[9] Rosacea also involves the eyes in about 17% of patients seen by dermatologists.[10] Ocular rosacea can manifest as blepharitis, conjunctival injection, and chalazion.

Lupus miliaris disseminatus faciei is another chronic facial papular eruption with a high predilection for the eyelids. The lesions are usually red or yellow-brown dome-shaped papules. Histologic examination demonstrates well-formed granulomas with central caseation necrosis. Lesions resolve spontaneously in 12 to 24 months with scarring. To our knowledge, lupus miliaris disseminatus faciei has not been reported in children. It has been hypothesized that most patients with this eruption actually have a form of granulomatous rosacea.[7]

Sarcoidosis is a systemic granulomatous disease with cutaneous manifestations. Cutaneous involvement occurs in up to one third of patients and can present as macules, papules, nodules, plaques, subcutaneous nodules, infiltrative scars, and ichthyosis. Maculopapular lesions are the most common cutaneous manifestation of sarcoidosis and can occur anywhere on the skin.[11] Lupus pernio is a variant of sarcoidosis characterized by violaceous papules and plaques on the nasal alar rims, ears, and cheeks. This variant of sarcoidosis occurs most frequently in middle-aged women and is associated with chronic fibrotic respiratory tract involvement. Patients with cutaneous sarcoidosis may have systemic symptoms including weight loss, shortness of breath, cough, fatigue, and bone and joint pain.

Perioral dermatitis most commonly occurs in young women between 16 and 45 years of age. The characteristic eruption consists of pustular or papulovesicular lesions on an erythematous background.[12] The lesions are usually confined to the chin and nasolabial folds, with sparing around the vermilion border. GPD may be distinguished from perioral dermatitis by the presence of discrete yellow-brown papules rather than erythematous papules, the lack of pustules, and the presence of a perifollicular granulomatous infiltrate seen on examination of biopsy material.[5]

Treatment of GPD is based on anecdotal reports. Tetracycline has been recommended for GPD in children older than 8 years.[3,13] Recent studies have demonstrated that tetracyclines inhibit granuloma formation in vitro.[14] Tetracyclines are also effective treatment for rosacea and perioral dermatitis because of their anti-inflammatory properties.[15] According to anecdotal reports, topical tetracycline and topical clindamycin are sometimes effective therapy for perioral dermatitis and may be helpful in GPD when oral agents are undesirable. GPD also has responded to topical treatment with metronidazole gel in some reports.[3,16] Mild GPD may resolve spontaneously over several months without therapy. The use of oral steroids in this condition has not been reported, but in our patient, a tapering course of prednisone seemed justified because of the distressing nature of the eruption to the patient. Oral prednisone may produce anti-inflammatory

effects more quickly than tetracycline, but it is not necessary in most patients.

GPD represents a benign cutaneous inflammatory process that resolves without serious sequelae. The granulomatous inflammation is most likely a nonspecific reaction to a variety of insults. Topical corticosteroids may initiate or exacerbate the granulomatous reaction and should be strictly avoided. Practitioners should recognize and distinguish this condition from other granulomatous eruptions so that patients are appropriately managed.

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