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Granulomatous Periorificial Dermatitis: A Variant of Granulomatous Rosacea in Children?

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Abstract

Background—We report a case of granulomatous periorificial dermatitis (GPD). A 13-year-old Jamaican boy presented with pink to flesh-colored perioral and periocular papules that erupted during a summer visit to Jamaica. The child was initially diagnosed with sarcoidosis and treated with oral steroids, but the eruption recurred 2 years later. He was referred to Dermatology. Biopsy of one of the facial papules revealed a dense granulomatous infiltrate with surrounding prominent lymphocytes. A diagnosis of GPD was made.

Objective and Conclusion—Also known as facial Afro-Caribbean childhood eruption (FACE), GPD occurs in prepubertal children. It can be confused with sarcoidosis, infection, and granulomatous rosacea but contains lymphocytes around the granulomas and lacks the systemic involvement seen in sarcoidosis. Cultures are invariably negative. The histologic features of GPD and granulomatous rosacea can be identical, and certain treatments may exacerbate the condition, highlighting the importance of clinical correlation. The correct diagnosis is important to minimize treatment as GPD is ultimately self-limited.

Granulomatous periorificial dermatitis (GPD) can mimic granulomatous rosacea and sarcoidosis. Gianotti and colleagues described five children with a granulomatous facial reaction in 1970.¹ Since then, several similar cases have been reported.^{2–13} The characteristic features are flesh-colored, pink, and yellow papules around the mouth, nose, and eyes in preadolescent children. The trunk and genitalia can also be involved. Classically described in children of Afro-Caribbean descent, GPD has also been called facial Afro-Caribbean childhood eruption (FACE), as well as Gianotti-type perioral dermatitis, granulomatous perioral dermatitis, and sarcoid-like granulomatous dermatitis.

The microscopic appearance of GPD can be identical to granulomatous rosacea, showing a superficial and deep dermal, noncaseating granulomatous infiltrate with macrophages and lymphocytes around hair follicles. We illustrate this entity in a preadolescent Jamaican boy who was also applying potent topical corticosteroids to a long-standing papular eruption around his mouth and nose. GPD is ultimately self-limited but may be exacerbated by treatments such as potent topical steroids, as in this case, highlighting the importance of a correct diagnosis.

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Case Report

A 13-year-old Jamaican boy presented with periorificial papules on the face that erupted during a summer visit to Jamaica 2 years earlier (Figure 1), where he was diagnosed with sarcoidosis after a biopsy of one of the papules. He initially responded to oral corticosteroids, but the eruption recurred two summers later, again while in Jamaica. The lesions were asymptomatic. He had been using betamethasone dipropionate 0.05% cream twice daily on his facial eruption with no response for several months when he first presented for evaluation. We immediately stopped this therapy.

His past medical history was significant for acne, eczema, asthma, and multiple food allergies.

Physical examination revealed multiple, nonscaly, round to oval, 1 to 4 mm, pink to flesh-colored papules tightly clustered around the mouth and, to a lesser extent, around the eyes and nose.

The involved areas had slight underlying hyperpigmentation. The rest of the skin examination and mucous membranes were uninvolved. There was no palpable lymphadenopathy.

Laboratory studies, including an erythrocyte sedimentation rate, a complete blood count, liver function tests, angiotensin-converting enzyme level, and 1,25-dihydroxyvitamin D, were all within normal limits. A computed tomographic scan of the chest showed no evidence of sarcoidosis.

Perioral and nasal fold skin biopsies revealed a dermal process with dense granulomatous infiltrate at all levels of the dermis (Figure 2) and around hair follicle epithelium (see Figure 2B). The infiltrate consisted of epithelioid cell granulomas with numerous surrounding lymphocytes and histiocytes. Scattered multinucleated giant cells were present. Polaroscopy was negative for refractile foreign material. Gomori methenamine silver and acid-fast bacilli (AFB) stains were negative (not shown). Bacterial, fungal, and AFB tissue cultures obtained from another facial skin biopsy were all negative.

Treatment consisted of erythromycin 400 mg four times a day for 6 weeks along with pimecrolimus 1% cream to relieve pruritus. Topical steroids were discontinued. The papular eruption cleared in approximately 2 months and has stayed clear, with occasional mild flares, requiring short-term reinstitution of pimecrolimus 1% cream.

Discussion

The histology of GPD closely resembles and can be identical to that seen in granulomatous rosacea. Both show noncaseating granulomas with a lymphohistiocytic infiltrate and a prominent perifollicular component. In fact, some authors view GPDs and granulomatous rosacea as variants of the same disease in different age and ethnic groups. It has been suggested that some cases of GPD are early rosacea, and the patient later goes on to develop more classic characteristics of rosacea.² However, the clinical picture suggests otherwise. The majority of cases of GPD have been self-limited, with no sequelae. Additionally, granulomatous rosacea is rarely seen in childhood. Telangiectasias, pustules, and flushing clinically, as well as ectatic vessels and pustules histologically, are not seen. Thus, the few cases of GPD reported to progress to rosacea may have been misdiagnosed, actually representing early granulomatous rosacea.

The differential diagnosis and distinguishing features are listed in Table 1 and were reviewed by Ahmed.¹⁴ Contact dermatitis can be evaluated with patch testing.¹⁵ Sarcoidosis differs clinically and histologically. Clinically, a limited perioral distribution and absence of other findings for sarcoidosis (abnormal laboratory tests or chest radiograph) would be unusual in sarcoidosis. Histologically, sarcoidosis is characterized by “naked granulomas,” without the inflammatory cells around and within the granulomas noted in this case. Infection (lupus vulgaris and tinea incognito) can be ruled out with histology and appropriate cultures. Lastly, the history is critical to determining an etiology; for example, children using inhalers can develop a rosacea-like facial dermatitis because of chronic exposure to corticosteroids.¹⁶

GPD may be on the spectrum of granulomatous rosacea, but based on the clinical behavior, it appears to be a distinct entity. The close resemblance to granulomatous rosacea, exacerbation by topical steroids, and self-limited course illustrate the importance of clinical correlation of granulomatous eruptions in children.

Some authors have suggested that the granulomas may be reactive to the lymphocytic infiltrate disrupting the hair follicles, similar to acne rosacea.^{5,7,8} This hypothesis is based on the observation that no granulomas were seen in some specimens. Otherwise, the pathophysiology is unknown.

The treatments of GPD and granulomatous rosacea are similar. Although the eruption is usually self-limited, treatment with tetracyclines, metronidazole, and erythromycin seems to be the most effective.^{4,17,18} Topical corticosteroid use is contraindicated as it may exacerbate both GPD and rosacea. The use of potent fluorinated topical steroids is thought by some to be an etiologic factor for the development of GPD.¹² However, in most reports, GPD actually develops prior to the use of potent topical steroids. Topical tacrolimus is effective, as demonstrated in this case.¹⁹

The etiology of GPD is not known, and it is unclear why the eruption typically occurs in children of Afro-Caribbean descent. GPD has been associated with essential oils in bubble gum, antiseptic solution, formaldehyde, and black synthetic mesh. In our case, the patient did chew gum sold in Jamaica but not in the United States. The eruption occurred both summers in Jamaica, supporting a potential etiologic role for environmental factors.

In summary, a history of exposures and topical corticosteroid use, ethnic background, and negative studies for infection and systemic disease such as sarcoidosis are helpful in the diagnosis of this rare disorder in children.

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References

1. Gianotti F, Ermacora E, Benelli MG, Caputo R. “Perioral dermatitis” in children and adults. *G Ital Dermatol Minerva Dermatol*. 1971; 46:132. [PubMed: 5109387]
2. Savin JA, Alexander S, Marks R. A rosacea-like eruption of children. *Br J Dermatol*. 1972; 87:425–9. [PubMed: 4265054]
3. Georgouras K, Kocsard E. Micropapular sarcoidal facial eruption in a child: Gianotti-type perioral dermatitis. *Acta Derm Venereol*. 1978; 58:433–6. [PubMed: 82354]

4. Falk ES. Sarcoid-like granulomatous periocular dermatitis treated with tetracycline. *Acta Derm Venereol.* 1985; 65:270–2. [PubMed: 2411091]
5. Frieden IJ, Prose NS, Fletcher V, Turner ML. Granulomatous perioral dermatitis in children. *Arch Dermatol.* 1989; 125:369–73. [PubMed: 2923443]
6. Smitt JH, Das PK, Van Ginkel JW. Granulomatous perioral dermatitis (facial Afro-Caribbean childhood eruption [FACE]). *Br J Dermatol.* 1991; 125:399. [PubMed: 1954141]
7. Knautz MA, Leshner JL Jr. Childhood granulomatous periorificial dermatitis. *Pediatr Dermatol.* 1996; 13:131–4. [PubMed: 9122070]
8. Williams HC. Childhood granulomatous periorificial dermatitis. *Pediatr Dermatol.* 1996; 13:515. [PubMed: 8987067]
9. Urbatsch AJ, Frieden I, Williams ML, et al. Extrafacial and generalized granulomatous periorificial dermatitis. *Arch Dermatol.* 2002; 138:1354–8. [PubMed: 12374542]
10. Tarm K, Creel NB, Krivda SJ, Turiansky GW. Granulomatous periorificial dermatitis. *Cutis.* 2004; 73:399–402. [PubMed: 15224784]
11. Misago N, Nakafusa J, Narisawa Y. Childhood granulomatous periorificial dermatitis: lupus miliaris disseminatus faciei in children? *J Eur Acad Dermatol Venereol.* 2005; 19:470–3. [PubMed: 15987296]
12. Nguyen V, Eichenfield LF. Periorificial dermatitis in children and adolescents. *J Am Acad Dermatol.* 2006; 55:781–5. [PubMed: 17052482]
13. Hussain W, Daly BM. Granulomatous periorificial dermatitis in an 11-year-old boy: dramatic response to tacrolimus. *J Eur Acad Dermatol Venereol.* 2007; 21:137–9. [PubMed: 17207200]
14. Ahmed I. Clinicopathologic challenge. Childhood granulomatous peri-orificial dermatitis with extra-facial lesions. *Int J Dermatol.* 2007; 46:143–5. [PubMed: 17269963]
15. Nedorost ST. Medical Pearl: the evaluation of perioral dermatitis: use of an extended patch test series. *J Am Acad Dermatol.* 2007; 56 (5 Suppl):S100–2. [PubMed: 17434026]
16. Dubus JC, Marguet C, Deschildre A, et al. Local side-effects of inhaled corticosteroids in asthmatic children: influence of drug, dose, age, and device. *Allergy.* 2001; 56:944–8. [PubMed: 11576072]
17. Fisher AA. Periocular dermatitis akin to the perioral variety. *J Am Acad Dermatol.* 1986; 15 (4 Pt 1):642–4. [PubMed: 2945840]
18. Miller SR, Shalita AR. Topical metronidazole gel (0. 75%) for the treatment of perioral dermatitis in children. *J Am Acad Dermatol.* 1994; 31 (5 Pt 2):847–8. [PubMed: 7962733]
19. Rodriguez-Martin M, Saez-Rodriguez M, Carnerero-Rodriguez A, et al. Treatment of perioral dermatitis with topical pimecrolimus. *J Am Acad Dermatol.* 2007; 56:529–30. [PubMed: 17317501]

Figure 1.

A 13-year-old boy with multiple, nonscaly, round to oval, 1 to 4 mm, pink to flesh-colored papules tightly clustered around the mouth and, to a lesser extent, around the eyes and nose.

Figure 2.

A, Low-power view showing granulomatous infiltrates at all levels of the dermis. *B*, Granulomas surround vessels and adnexal structures (*arrow*). *C*, The infiltrates consist of giant cells, histiocytes, and numerous lymphocytes. *A–C*, Hematoxylineosin stain; original magnifications: *A*, $\times 10$; *B*, $\times 20$; *C*, $\times 40$.

Table 1**Papulonodular Disorders on the Faces of Children**

Disorder	Clinical	Pathology
Granulomatous periorificial dermatitis	Papules and nodules on the faces of prepubertal children	Periadnexal inflammation of lymphocytes and plasma cells with noncaseating granulomas Multinucleated giant cells common
Sarcoidosis	“Apple jelly” firm red-brown papules around eyes, ears, cheeks, and nose Lacrimal gland hyperplasia Uncommon in children	“Naked” epithelioid noncaseating granulomas with scant inflammatory cells Multinucleated giant cells uncommon
Granulomatous rosacea	Erythematous papules and pustules around eyes and on cheeks Uncommon in children	Periadnexal inflammation of lymphocytes and plasma cells with noncaseating granulomas Multinucleated giant cells common
Perioral dermatitis	Erythematous papules and pustules resembling rosacea mainly around the mouth	Perivascular and perifollicular inflammation of lymphocytes with giant cells Dermal edema
Lupus miliaris disseminata faciei	Red-brown papules symmetrically on eyelids, periorbital area, nose, and lips Develops rapidly and scars Uncommon in children	Periadnexal caseating granulomas and prominent fibrosis
Lupus vulgaris	Erythematous nodular lesions around the nose, cheeks, ears, and neck Ulceration can lead to disfiguring scarring Can begin in childhood	Epithelioid granulomas and nonspecific inflammatory infiltrates with scant caseation Variably positive mycobacterial stains
Perioral contact dermatitis	Erythematous papules and plaques around the mouth	Spongiotic dermatitis with Langerhans cell microgranulomas Eosinophils in allergic contact dermatitis Epidermal necrosis in irritant contact dermatitis
Tinea incognito	Erythematous papules and pustules on any part of the face Annular lesions	Spongiotic dermatitis with dermatophyte hyphae in stratum corneum