Granulomatous Periorificial Dermatitis in a Young Woman

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Granulomatous periorificial dermatitis is a skin disease presenting predominantly in prepubertal black-skinned children. It is characterized by tiny, monomorphous, papular eruptions around the mouth, nose and eyes. Extra-facial lesions in children had also been reported. These lesions are self-limited and resolved without scarring in most cases. We report a 28-year-old woman presented with pink to normal skin colored, discrete and coalescing papules ranging from 1 to 3 mm in diameter over the face, nape and bilateral forearms of 8 months duration. A biopsy on the right nasolabial fold showed dermal granuloma formation around hair follicles, composed of lymphocytes, epithelioid histiocytes and occasional multinucleated giant cells. Oral doxycycline and topical metronidazole gel were given and the skin lesions cleared after 2 months treatment. (Dermatol Sinica 24: 38-41, 2006)

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INTRODUCTION

In 1970, Gianotti et al. first described five children, ranging from 2 to 7 years old, who had asymptomatic, distinctive normal skin colored "micronodular" eruption surrounding the mouth. Biopsy specimens demonstrated a lymphohistiocytic infiltration with occasional giant cells. In the literature, the condition has been variably called Gianotti-type perioral dermatitis, sarcoidlike granulomatous dermatitis, facial Afro-Caribbean childhood eruption (FACE), and childhood granulomatous perioral dermatitis.

CASE REPORT

A 28-year-old healthy lady complained of asymptomatic, papular skin eruption on the face, nape and bilateral forearms at our outpatient clinic in June, 2004. She claimed that the asymptomatic papular skin rash on the face happened 4 years ago and disappeared spontaneously about a year later. Similar skin lesions were noted again on her face about 8 months ago with progression to the nape and bilateral forearms gradually. Topical clindamycin, topical steroid, and oral Chinese herbal drugs were given but in vain. She denied any topical or oral medications prior to the onset of the skin rashes. Physical examination revealed multiple skin papules over the paranasal, perioral, periocular areas, glabella, nape and bilateral forearms (Fig. 1). There were many 1- to 3-mm, pink to normal skin colored papules scattered discretely or grouped and most concentrated on the paranasal area with extension to the nostrils and involvement of the vermilion borders. Neither comedo nor telangiectasia was noted. Laboratory data, chest X ray and KOH examination were all normal without specific findings. A skin biopsy specimen from the right nasolabial fold showed mild spongiosis, basal vacuolization and lym-
phocyte exocytosis of inter- and intra-follicular epidermis. Dermal granuloma formation around the hair follicles was noted and composed of lymphocytes, epithelioid histiocytes and occasional multinucleated giant cells. Mild dermal fibroplasia, interstitial lymphohistiocytic infiltration and vascular dilatation were noted (Fig. 2). Acid-fast, periodic acid-Schiff, and Giemsa stains revealed no evidence of specific microorganisms. Granulomatous periorificial dermatitis was diagnosed. Stop using all of her medications was suggested. Oral doxycycline, 100 mg twice a day combined with topical 0.75% metronidazole gel was given for 3 weeks. Improvement was noted. Metronidazole gel was continued for further 3 weeks and all the skin lesions disappeared without scarring and pigmentation 2 months later. No recurrence was noted after 6 months follow-up.

DISCUSSION

Granulomatous periorificial dermatitis (GPD) is an uncommon skin disease occurred predominantly in prepubertal children. Black children were affected much more commonly than children of other racial backgrounds. Both genders are equally affected. Arguably, it has been proposed that GPD is a localized variant of rosacea. However, some authors suggested that "granulomatous rosacea" does not associate with persistent facial erythema, flushing, and is a misnomer in rosacea group. They suggest a new name "granulomatous facial dermatitis" to categorize diseases including GPD, facial Afro-Caribbean childhood eruption (FACE), and lupus miliaris disseminatus faciei.

In the literature, GPD is often considered as a granulomatous variant of perioral dermatitis. The classic perioral dermatitis consists of papular, pustular or papulovesicular lesions on an erythematous background. The skin lesions are usually confined to the chin and nasolabial folds and spare around the vermilion border. In perioral dermatitis, most of the histopathology reports have shown mild, nonspecific, subacute inflammation with variable perifollicular or perivascular lymphohistiocytic infiltration and occasional papillary edema. However, some cases with granulomatous changes have been reported and were designated as GPD. GPD is characterized by a monomorphous, papular eruption occurring around the mouth, nose and eyes. The primary lesion is a discrete 1-3 mm dome-shaped flesh-colored, yellow-brown, or red papule. Slight scaling of the lesions or surrounding erythema may occur. Extrafacial lesions had been reported involving the trunk, extremities, labia majora and could be generalized. Extrafacial involvement do not appear to adversely affect the duration, response to therapy, or risk of extracutaneous manifestation. Whether classic type GPD or GPD with extrafacial involvement, in most cases the lesions healed without sequelae and only occasionally the disease resolved with pinpoint atrophic scars or altered pigmentation. Results of routine laboratory studies and chest radiographs are usually normal. Most of the GPD occurred in children. In the literature, GPD had been described in a young woman with a persistent eruption around the mouth and chin. She was unresponsive to conventional therapies, and oral isotretinoin was given. After 20 weeks treatment, the lesions cleared and left pitted, atrophic scars. Histological findings of GPD reveal upper dermal and perifollicular granuloma formation. The granulomas consist of epithelioid histiocytes, lymphocytes and occasional multinucleated giant cells. Focal epidermal spongiosis is also described. In addition, special stains should be done to rule out possible infectious causes such as lupus vulgaris, atypical mycobacterial infection, and late secondary or tertiary syphilis. The presence of lymphocytic inflammation can help distinguish GPD from the "naked" granulomas in cutaneous sarcoidosis that typically lack inflammatory cells. The etiology of GPD is unknown. Topical corticosteroid may induce or exacerbate both GPD and perioral dermatitis. Some suggested it to be an unusual granulomatous inflammatory response to allergens. The initial allergen may cause inflammation and a focal disruption of the follicular wall, inciting a granulomatous reac-
tion.9 Suspect inciting subject including essential oils in bubble gum, formaldehyde, cosmetic preparations, and antiseptic solutions.6,12 The clinical differential diagnosis includes trichoepithelioma, cutaneous sarcoidosis, fungal or mycobacterial infection. However, these disorders could be differentiated by the detailed history, review of systems, thorough physical examination, laboratory studies, and special stains or tissue cultures of biopsy specimens.

Although the new name "granulomatous facial dermatitis" was suggested to categorize diseases including GPD and lupus miliaris disseminatus faciei (LMDF),5 clinically, LMDF presents as multiple red to yellow-brown papules, nodules affecting central area of the face with a remarkable preference for the eyelids.13 Histopathological examination shows tuberculoid granulomas usually with central caseation necrosis. As the name LMDF suggests, it was originally presumed to be an expression of cutaneous tuberculosis or a tuberculid due to its histopathological similarity. However, all other facts including absence of bacilli in the lesions, not detectable of Mycobacterium tuberculosis by polymerase chain reaction were all against with the hypothesis of tuberculosis.13,14 The lesions of LMDF are often disappeared spontaneously with ice-picking scars.14 The prognosis of GPD is good and spontaneous resolution usually occurs by a few months to 3 years after onset.7 Although it is asymptomatic, patients seek for medical help due to disfiguring cosmetically. At first, topical corticosteroid should be strictly avoided. The mainstay of treatment includes administration of oral macrolides or tetracycline, alone or in combination with topical erythromycin, metronidazole, or sulfur-based lotion would hasten resolution in most patients.6,7,9-10 Successful treatment of GPD by oral doxycycline and topical metronidazole gel in this adult patient is encouraging, although spontaneous improvement cannot be ruled out. In our experience, practitioners should recognize this condition from other facial eruptions and therefore the patients are appropriately managed.

REFERENCES