Granulomatous Periorificial Dermatitis in an Adult
A case report with review of literature

Abstract: Granulomatous periorificial dermatitis (GPD) is a benign, self-limiting eruption that is considered a clinical variant of periorificial dermatitis, also known as perioral dermatitis. It presents primarily in prepubertal children as monomorphic scaly papules over perioral, paranasal and periorbital areas of the face with rare occurrence in adults. We report a 36-year-old Omani male patient who presented to the Dermatology Clinic at Bahla Polyclinic, Bahla, Oman, in 2018 with a papular eruption over his face for the previous six months. Based on clinical and histopathological findings the patient was diagnosed with GPD with sarcoid-like histology. He was treated effectively with oral doxycycline and topical metronidazole. This report provides a review of the literature on GPD and summarises all reported cases in adults to date.

Keywords: Perioral Dermatitis; Dermatitis; Granulomas; Case Report; Oman.

Case report
A 36-year-old Omani male patient presented to the Dermatology Clinic at Bahla Polyclinic, Bahla, Oman, in 2018 with a papular eruption over his face for the previous six months. He reported no itching, burning sensation or facial redness. He had no known comorbidities and denied a history of fever, shortness of breath or other systemic complaints. In addition, he denied having used any topical or oral medications prior to the eruption. There was no recent history of travel and no abnormal environmental exposure. He was prescribed topical mometason creamy for two months and tacrolimus (0.1%) ointment for three months with partial response and recurrence once the treatment was discontinued.

Following the reoccurrence of the papular eruption, examination showed monomorphic scaly erythematous papules localised to the perioral, paranasal and peri-orbital areas of the face [Figure 1]. The vermilion border

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was involved. There was no facial erythema or telangiectasia. There were no other skin lesions and other orifices were not involved. The rest of the physical examination was unremarkable. The differential diagnoses included periorificial dermatitis, GR, cutaneous sarcoidosis and lupus miliaris disseminatus faciei (LMDF).

Laboratory investigations, including angiotensin converting enzyme (ACE) levels, were normal. The chest X-ray was also normal. A punch biopsy was taken from the area with erythematous papules and sent for histological investigation.

Histopathological examination of an erythematous papule showed non-caseating naked granulomas containing histiocytes, multinucleated giant cell of Langhans type and focally surrounded lymphocytes [Figure 2]. Periadnexal and perivascular lymphocytic infiltrate was also present. Stains for fungi (i.e. periodic acid–Schiff) and acid-fast bacilli (i.e. Ziehl-Neelsen and Wade-Fite) were negative.

As a result of the clinical and histopathological findings, the patient was diagnosed with GPD. The patient was treated with oral doxycycline (100 mg) once daily and topical metronidazole cream twice daily. He showed marked improvement after six weeks with complete resolution of the lesions without scarring after 12 weeks, after which treatments were stopped [Figure 3]. He had no recurrence on follow-up visits after three and six months.

The patient gave consent for his images and clinical information to be reported in a journal. The authors explained that while the patient’s name would not be published, complete anonymity could not be guaranteed.

**Discussion**

GPD is a well-recognised entity that affects commonly dark-skinned prepubertal children. While topical steroids are considered to be the most important and frequently reported pathogenic factor, other reported factors include cosmetic products, physical factors and microorganisms.1,8–11 GPD is a controversial disease as it shares many similarities with other granulomatous disorders such as GR and cutaneous sarcoidosis. GDP is distinguished from cutaneous sarcoidosis by the absence of systemic involvement and a self-limiting nature. Antony et al. reported a case of GPD that could be a variant of sarcoidosis with raised ACE levels and a chronic nature.8 GR usually shows similar histology to GPD but it mainly affects the central face and may show classic signs of telangiectasia, oedema and erythema.7 LMDF is distinguishable from GPD as it has a tendency to affect periorbital areas only, a presence of caseation on histology and resolution with scarring. Misago et al. reported a case of CGPD with similar features to LMDF suggesting that the term ‘facial idiopathic granulomas with regressive evolution’ should include both CGPD and LMDF.12 Since GPD sometimes presents with eczematous features, seborrhoeic dermatitis is also an accepted differential diagnosis, but the latter usually involves other areas such as the eyebrows and scalp with a dramatic response to topical steroids and different histologic features without granulomas. Dermatoscopy may show additional features that aid in diagnosis.13 Table 1 summarises the differential diagnoses with
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Table 1: Differential diagnosis of granulomatous papules on the face \(^{1,13-17}\)

| Disease          | Typical patient characteristics | Clinical features                                           | Main dermoscopic features                                      | Histopathological findings                                      | Prognosis                        |
|------------------|---------------------------------|-------------------------------------------------------------|----------------------------------------------------------------|----------------------------------------------------------------|----------------------------------|
| GPD              | Prepubertal children             | Monomorphic, skin-colored to yellow-brown or red papules confined to the periorificial areas of the face | Not described                                                 | Dermal non-caseating granulomas                                  | Spontaneous resolution without scarring |
| GR               | Middle-aged women                | Yellow-brown or pink papules on the cheeks, perioral or perioral skin | Linear reddish or purple vessels arranged in a polygonal network (vascular polygons) | Epithelioid granulomas adjacent to hair follicles; Casation in 10% of the cases | Chronic nature                  |
| LMDF             | Young adults                     | Reddish-yellow or yellowish-brown papules on the central face and eyelids | Follicular keratotic plugs and vascular structures             | Casating granulomas                                              | Spontaneous resolution with scarring |

GPD = granulomatous periorificial dermatitis; GR = granulomatous rosacea; LMDF = lupus miliaris disseminatus faciei.

Table 2: Summary of reported cases of granulomatous periorificial dermatitis in adults \(^{1-5}\)

| Author and year of publication | Age in years | Gender | Clinical presentation | Histopathological findings | Treatment (duration) |
|--------------------------------|--------------|--------|-----------------------|---------------------------|---------------------|
| Chintagunta et al. \(^{1}\)    | 34           | Female | Well-defined erythematous to pigmented plaques associated with scaling involving the perioral, paranasal and glabella | Granulomatous inflammation in the dermis composed of lymphocytes, histiocytes, epithelioid cells and multinucleated giant cells | Oral doxycycline 100 mg OD + pimecrolimus 1% cream BID (3 months) |
| Vincenzi et al. \(^{2}\)       | 19           | Female | Numerous, flesh-colored micropapular lesions involving the perioral and perinasal areas associated with a mild diffuse erythema and slight vesiculation | Numerous well-formed granulomas containing occasional multinucleated giant cells in the dermis | Oral clarithromycin 250 mg OD (6 weeks) then 125 mg OD (8 weeks) |
| Vincenzi et al. \(^{3}\)       | 25           | Female | Numerous red micropapules involving the nasal folds and the perioral regions | Perifollicular non-caseating epithelioid cell granulomas in the dermis with some multinucleated giant cells and a variable number of lymphocytes and histiocytes in perivascular and perifollicular areas | Oral clarithromycin 250 mg OD (10 days) then 250 mg on alternate days (20 days) |
| Tambe et al. \(^{4}\)          | 30           | Female | Multiple erythematous, scaly papules and plaques on the supra orbital, peri-orbital, perioral and perinasal area | Perifollicular and perivasculcar granulomatous inflammatory infiltrate composed of lymphocytes, epithelioid cells and giant cells | Oral isotretinoin 20 mg OD + metronidazole cream (3 weeks) |
| Li et al. \(^{5}\)             | 28           | Female | Pink to normal skin-colored, discrete and coalescing papules ranging from 1–3 mm in diameter over the face, nape and bilateral forearms | Dermal granuloma formation around hair follicles, composed of lymphocytes, epithelioid histiocytes and occasional multinucleated giant cells | Oral doxycycline 100 mg OD + topical metronidazole gel (2 months) |
| Loai and Huang \(^{6}\)        | 24           | Female | Multiple, discrete, red to brown papules on erythematous base on the perioral and perioral areas | Granulomatous infiltration composed of lymphocytes, histiocytes, epithelioid cells and multinucleated giant cells, without caseation in the dermis | Oral minocycline 50 mg bid + tacrolimus ointment 0.03% BID (50 days) |
| Present case                | 36           | Male   | Monomorphic scalp erythematous papules localised to the perioral, paranasal and periorbital areas of the face | Non-caseating naked granulomas containing histiocytes, multinucleated giant cell of Langhans type and focally surrounded lymphocytes | Oral doxycycline 100 mg OD + metronidazole cream (3 months) |

OD = once daily; BID = twice daily.

clinical and dermoscopic features of granulomatous papules on the face.

GPD has a self-limiting nature, therefore treatment is not necessary. However, many topical and systemic treatments have been reported to hasten clearance. \(^{12,16,17}\) Topical treatments include metronidazole, erythromycin or pimecrolimus. \(^{16,17}\) Systemic treatments mainly include tetracycline antibiotics such as tetracycline and doxycycline; oral erythromycin and clarithromycin are also effective. \(^{2,7,18}\)

To date, there are a total of six reported cases of GPD in adults [Table 2]. \(^{1-5}\) The present case is the only case in a male. In two cases, the lesions were erythematous plaques and the remainder had erythematous papules as in the present case. All cases showed dermal non-caseating granulomas upon histopathological examination. One case was treated with oral isotretinoin while the others were treated with oral antibiotics. All cases showed complete resolution without recurrence.
Conclusion

GPD is a well-recognised entity that may affect adults and should be differentiated from GR, cutaneous sarcoidosis and other granulomatous disorders of the face by clinicopathological correlation to minimise systemic treatment use.

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