CASE REPORT

Disfiguring facial pyoderma vegetans with an excellent outcome

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SUMMARY

We describe a case of disfiguring facial pyoderma vegetans in order to highlight the challenges in managing this rare skin condition and review the literature.

A 54-year-old woman presented to dermatology clinic with a 3-month history of a left-sided facial lesion, which had been treated as an infected sebaceous cyst. The lesion had dramatically increased in size in the weeks prior to presentation. There was a history of Crohn's disease and ileal adenocarcinoma, both of which were in remission. A clinical diagnosis of pyoderma vegetans was made and the patient responded well to immunosuppressive therapy with oral ciclosporin. Carbon dioxide (CO₂) laser resurfacing to residual scarring contributed to an excellent cosmetic result.

BACKGROUND

Pyoderma vegetans (PV) is a rare inflammatory skin disease, presenting with large, persistent, vesiculopustular exudative plaques with well-defined borders.¹ Reported here is a case of PV, which illustrates the challenges of diagnosis and management of a rare dermatological condition alongside the need to balance risks and potential benefits of treatment for patients with complex healthcare needs.

Our patient experienced a rapid progression of her facial lesion prior to referral, thus highlighting the importance of clinical re-evaluation in apparently straightforward cases behaving atypically. The significant psychological burden resulting from skin disease is also evidenced.

Unusually, this case of PV occurred in a woman with inactive Crohn's disease and against a background of previous ileal adenocarcinoma. Treatment

Figure 2 Micrograph of skin biopsy demonstrating neutrophils in an eccrine duct. Magnification x20.

of PV is often difficult, with delayed diagnosis and unsatisfactory outcomes reported in the literature.¹ Despite the relative risks associated with treatment, our patient achieved an excellent outcome.

CASE PRESENTATION

A 54-year-old woman presented with a 3-month history of a large, left-sided facial lesion (*figure 1*). It had been treated in primary care as an 'infected sebaceous cyst' but had failed to respond to multiple courses of oral antibiotics and had rapidly enlarged in the weeks before presentation. She had a 20-year history of Crohn's disease that had been quiescent for a number of years, ileal adenocarcinoma 7 years previously, congestive cardiac failure and severe chronic obstructive pulmonary disease, which restricted her mobility. There was a family history of inflammatory bowel disease.

She lived with her husband and was a carer for their disabled child. She described periods of low mood and had become self-conscious about her facial disfigurement to the extent that she was virtually housebound.

On examination, there was a large, tumid plaque of coalescing papulopustular lesions on the left cheek. The raised, vegetative surface had a cobblestone-like appearance and a well-circumscribed border (*figure 1*). The surrounding facial skin was normal.

INVESTIGATIONS

A transient peripheral blood eosinophilia ($0.5 \times 10^9/L$) was noted at the time of presentation.

An incisional skin biopsy was performed and showed a severe inflammatory process with abscess formation. The epidermis was acanthotic,

Figure 1 Skin appearances at presentation to the dermatology clinic.

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Figure 3 Treatment of pyoderma vegetans with topical therapies resulted in an encouraging clinical response.

spongiotic and featured exocytosis of lymphocytes. There were numerous intraepithelial neutrophils. Eosinophilic spongiosis was not a feature. Dense mixed—predominantly acute—inflammatory changes were abundant within the dermis and there were sinus tracts lined with histiocytes. Well-defined granulomas were not identified. Relatively scanty intradermal eosinophils were present. There were intraepithelial abscesses within eccrine glands. There was no evidence of vasculitis or malignancy. Stains for fungi, bacteria and mycobacteria were negative (figure 2).

Skin swabs taken for bacteriology cultured normal skin flora and coliforms.

A chest, abdomen and pelvis CT scan, performed to exclude recurrence of ileal adenocarcinoma or Crohn's disease, was normal with the exception of renal artery stenosis as an incidental finding. Renal function was normal.

Based on clinical features and histological findings, a diagnosis of PV was made.

DIFFERENTIAL DIAGNOSIS

- ▶ Pyoderma vegetans
- ▶ Cutaneous Crohn's disease
- ▶ Botryomycosis
- ▶ Pyoderma gangrenosum
- ▶ Pemphigus vegetans

Figure 4 Following treatment with ciclosporin, the lesion of pyoderma vegetans had resolved leaving an atrophic macular scarred area on the patient's left cheek, which had cobblestone lesions peripherally and an island of verrucous tissue centrally. There was no evidence of residual inflammation within the skin.

TREATMENT

Our patient was initially treated with oral prednisolone 30 mg/day and lymecycline 408 mg/day in addition to topical therapy with betamethasone with clioquinol. The clinical response to treatment was encouraging. Following histology review, her topical treatment was intensified to 0.3% tacrolimus in clobetasol 17-propionate ointment, which resulted in additional improvement (figure 3).

When our investigations confirmed that there was no recurrence of ileal adenocarcinoma or Crohn's disease and that despite renal artery stenosis her renal function was normal, oral ciclosporin at a dose of 4 mg/kg/day was started. There was an excellent response to treatment and the ciclosporin dose was gradually reduced to 2 mg/kg. Remission was achieved after 8 months and ciclosporin was stopped.

However, despite resolution of PV, our patient was left with significant skin texture change, warty excrescences, scarring and telangiectasia that were a focus of considerable distress for her (figure 4).

Although we were concerned that her PV could Koebnerise (skin disease recurring at the site of trauma), due to the impact of the patient's facial appearance on her quality of life, a referral to the dermatology laser team was made. Following assessment, carbon dioxide (CO₂) laser resurfacing was considered, and after a successful test patch, she underwent resurfacing of the area of residual scarring on her left cheek (figure 5).

Figure 5 Skin appearances at discharge demonstrating a good result to a combination of topical, systemic and laser treatments.

OUTCOME AND FOLLOW-UP

Following an excellent cosmetic result from laser resurfacing, we observed a significant improvement in our patient's mood. There was no evidence of Koebnerisation or recurrent disease activity during follow-up for 4 years after initial presentation, at which time point she was discharged.

DISCUSSION

PV is a rare, chronic vegetative skin disease first described in 1898 by Hallopeau.² PV most commonly affects intertriginous areas, the face, oral mucosa and scalp^{3–5} where erythematous cutaneous papulopustular lesions rapidly coalesce into raised vegetative plaques with well-circumscribed borders. The condition usually presents between 20 and 50 years of age and has a 3:1 male predominance.^{4,6} The pathogenesis and aetiology are incompletely understood, although bacterial colonisation in immunosuppressive states has been described.^{1,3} PV has been most frequently associated with ulcerative colitis (UC),^{5–16} and disease activity is an influential factor with presentation of PV often occurring during flares of UC.^{5,12} In contrast, PV is less commonly associated with Crohn's disease^{11,17–23} where it can occur during phases of low or absent Crohn's disease activity.²⁴ PV can be associated with lymphoproliferative disorders, alcoholism, HIV and immunosuppressive therapy.¹ It has also been described in immunocompetent and apparently healthy individuals.³ The presentation of our case was unusual in several respects, namely a female patient with quiescent Crohn's disease. In addition, although the oral variant of PV has been associated with adenocarcinoma in one case report,⁸ to the best of our knowledge, our case is the first to describe cutaneous PV presenting in a patient with a previous history of adenocarcinoma.

Diagnosis of PV can be challenging and it usually depends on differentiating it from other chronic, vegetative dermatoses such as cutaneous Crohn's disease and botryomycosis. Cutaneous Crohn's disease can cause erythema, ulceration and cobblestoning, commonly of the face or oral cavity.¹² Botryomycosis results from bacterial infection usually with *Staphylococcus aureus* and presents variably—commonly with ulceration, nodulation and fissuring, which is not associated with inflammatory bowel disease.²⁵ Immunobullous disorders, such as pemphigus vegetans, are characterised by deposits of IgG and complement on direct immunofluorescence.²⁶ PV may also closely resemble pyoderma gangrenosum (PG), an ulcerating neutrophilic dermatosis associated with inflammatory bowel disease in 30% of cases.²⁷ There are no diagnostic histological features for PG, but a perivascular lymphocytic infiltrate and vasculitic changes to the ulcer edge are commonly seen.²⁷

There is considerable debate in the literature whether PV and PG are variants of the same disease although there are also well-documented differences, which suggest that they are distinct entities per se. In contrast to PV, cases of PG do not appear to closely correlate with inflammatory bowel disease activity.²⁸ Large case series have reported that eosinophilia on histopathology, and to a lesser extent peripheral eosinophilia, may be helpful in distinguishing PV from PG.²⁸ Notably, our patient had a transient peripheral blood eosinophilia at the time of presentation, which in conjunction with the pattern of clinical features supported the diagnosis of PV. We also observed no Koebnerisation of our patient's lesion following CO₂ laser resurfacing, which, in contrast to PG, is in keeping with other reports in the literature where multiple biopsies have been taken from lesions of PV without extension or induction of new lesion formation.

PV is a rare entity that may or may not be a variant of PG, pemphigus vegetans or pyodermitis vegetans. In addition, nomenclature has been unclear; pyostomatitis vegetans has been used to describe oral lesions and pyodermitis–pyostomatitis vegetans termed a separate entity.³ As with many rare disorders, a clear therapeutic strategy is not well defined. In the literature, PV has most commonly been treated with steroids, either topical, oral or both,^{1,4,8–11,15,16,19,21,23,29,30} and immunosuppressive treatments, such as ciclosporin, have shown promising results.^{4,17} Oral tetracyclines have demonstrated utility in the management of PG due to their inhibition of inflammation and dermal destruction, and these were therefore successfully incorporated into the management plan of our patient.³¹ Anti-tissue necrosis factor biologic agents have been used successfully,^{1,17,32} and in UC-associated PV, colectomy can lead to remission.^{8,15,33,34}

Our case highlights the importance of careful clinical and histological evaluation in an apparently straightforward case and the requirement for clinicians to consider the patient holistically when designing a management plan. Our case also illustrates successful treatment outcome as a consequence of teamwork across a network of specialist dermatology services including medical dermatology, dermatopathology and laser dermatology.

Learning points

- This case highlights the importance of careful clinical, supported by histological, evaluation in apparently straightforward cases. Be prepared to reconsider an initial diagnosis if there is no improvement.
- The risks of treatment must be carefully balanced against the potential benefits of treatment on a case-by-case basis.
- The psychological impact of dermatological diseases cannot be overestimated. Many common dermatological diseases, although not life threatening per se, can be devastating and life ruining for sufferers.
- Management of dermatological conditions are complex and present diagnostic and treatment challenges to the clinician.

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