Pyoderma gangrenosum of the tongue

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Introduction

Pyoderma gangrenosum is an uncommon neutrophilic dermatosis causing progressive, painful cutaneous ulceration. Head and neck involvement is rare, with only 20 cases of pyoderma gangrenosum with oral involvement reported. A 36-year-old white man presented with a 4-month history of oral ulceration involving the dorsum tongue. His medical history was significant for Crohn's disease. He was taking 6-mercaptopurine 25 mg daily and infliximab every 8 weeks at the time of presentation. On examination, there was a 4 cm × 2 cm soft superficial ulcer with ragged margins and a yellow base on the dorsum of the tongue. Blood monitoring identified anti-infliximab antibodies and low infliximab levels. Adalimumab 40 mg weekly subcutaneously was commenced. There was almost complete resolution at 3-month review. Oral pyoderma gangrenosum may present a diagnostic challenge. Early diagnosis, management and follow up are essential because of the associated morbidity. Treatment of the underlying systemic condition is an integral part of management.

Discussion

Systemic disorders are associated with approximately 80% of cases of pyoderma gangrenosum, notably inflammatory bowel disease, rheumatoid arthritis and seronegative arthritides, haematological malignancies and monoclonal gammopathy (Newell et al,

Case report

A 36-year-old white man presented with a 4-month history of a persistent solitary ulcer involving the dorsum tongue. There was a history of recurrent aphthous ulceration for approximately the last 3 years. There were no cutaneous lesions. His medical history was significant for Crohn's disease. He was being treated with 6-mercaptopurine 25 mg daily (commenced 1 year ago) and infliximab 5 mg/kg every 8 weeks (commenced 1 year ago). There were no gastrointestinal symptoms reported at the time of presentation. He was a former smoker.

On examination, there was a 4 cm × 2 cm soft well-demarcated superficial ulcer with ragged margins and a yellow base on the dorsum of the tongue (Figure 1). All other skin and mucosal surfaces were unaffected.

At presentation, blood monitoring revealed ferritin of 16 µg/litre (reference range 22–275 µg/litre). Full blood count, vitamin B12 and folate levels were within normal range. C-reactive protein levels and erythrocyte sedimentation rate were normal. Faecal calprotectin was 17 µg/g (reference range 0–49 µg/g). The thioguanine level was 356 pmol/8x10^8 red blood cells (reference range 235–450 pmol/8x10^8 red blood cells). Serum infliximab levels and anti-infliximab antibody levels were normal. Colonoscopy revealed a small linear ulcer at the previous surgical anastomosis site and two proximal aphthae.

An incisional biopsy showed part-ulcerated lingual mucosa covered by parakeratinised epithelium, which was variably hyperplastic at the ulcer periphery. The ulcer bed and surrounding superficial lamina propria contained a chronic, mixed inflammatory infiltrate. Periodic acid–Schiff staining was negative for fungal hyphae. There was no granulomatous inflammation (Figures 2a and b).

The patient was treated with Flixonase 400 mcg nasules used twice daily as a mouthwash, clobetasol 17-propionate 0.05% ointment, intralesional triamcinolone 40 mg/ml, tacrolimus 0.1% ointment and prednisolone 30 mg daily for 2 weeks, reducing by 5 mg every 3 days until discontinued, with minimal improvement. Blood monitoring 1 year after the initial presentation identified anti-infliximab antibodies and low infliximab levels. Adalimumab 40 mg weekly subcutaneously was commenced, in addition to continuing 6-mercaptopurine treatment. There was almost complete resolution of the ulcer at 3-month review (Figure 3).

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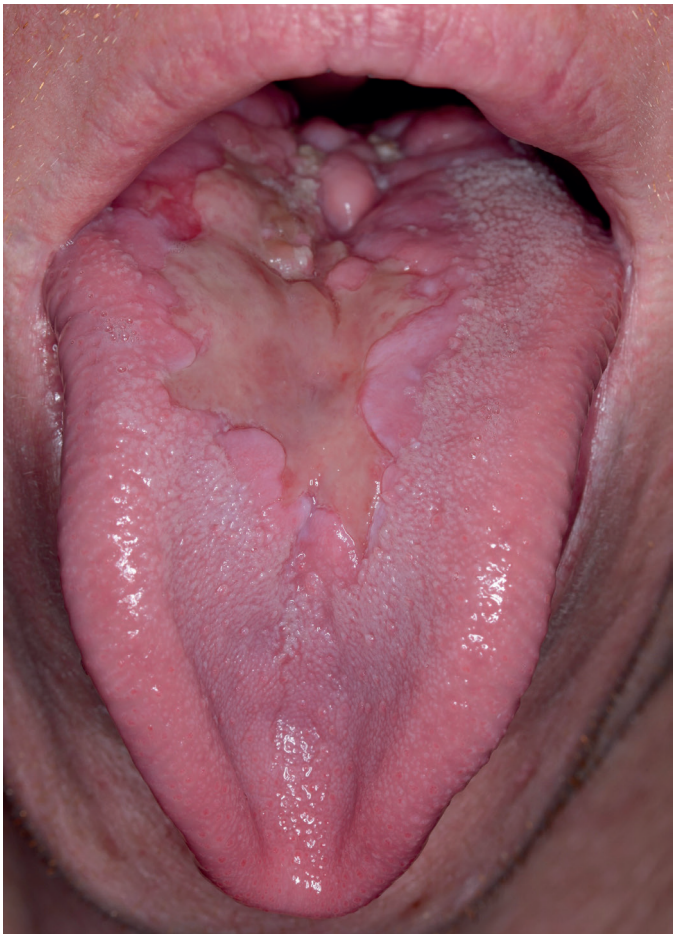

Figure 1. Extensive superficial ulcer with ragged margins and a yellow base on the dorsum of the tongue.

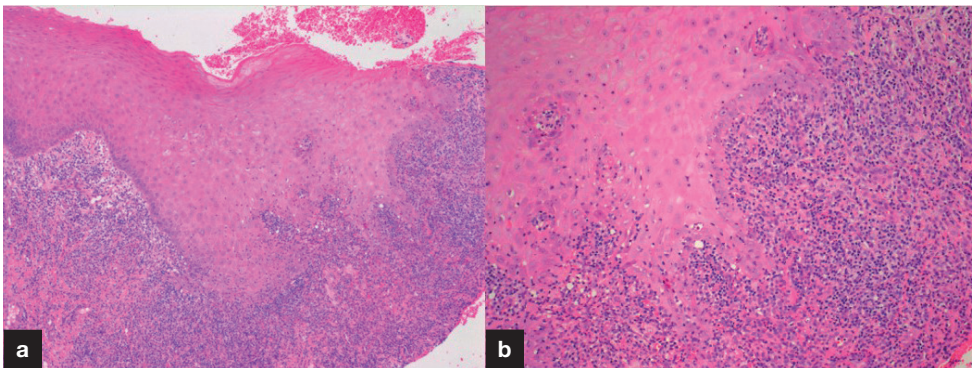

Figure 2. a. Part-ulcerated lingual mucosa covered by parakeratinised epithelium, which was variably hyperplastic at the ulcer periphery (haematoxylin and eosin × 5). b. The ulcer bed and surrounding superficial lamina propria contained a chronic, mixed inflammatory infiltrate (× 10).

2008). Lesions are often initiated by minor trauma (Bissonnette et al, 2017). Head and neck involvement is rare, with only 20 cases of oral pyoderma gangrenosum reported (Abela et al, 2007; Paramkusam et al, 2010; Curi et al, 2013; Bissonnette et al, 2017; Woo et al, 2017). Oral lesions in the absence of concomitant cutaneous involvement are found in 20% (Bissonnette et al, 2017), most frequently on the tongue, buccal mucosa and soft palate. Oral lesions present as red-coloured nodules or papulo-pustules that rupture to form ragged, irregular large ulcers. Bone loss and destruction of the periodontal support has been reported (Paramkusam et al, 2010).

There are no specific histopathological features on oral biopsies that point towards a diagnosis of pyoderma gangrenosum. Typical descriptions in the literature are ‘chronic

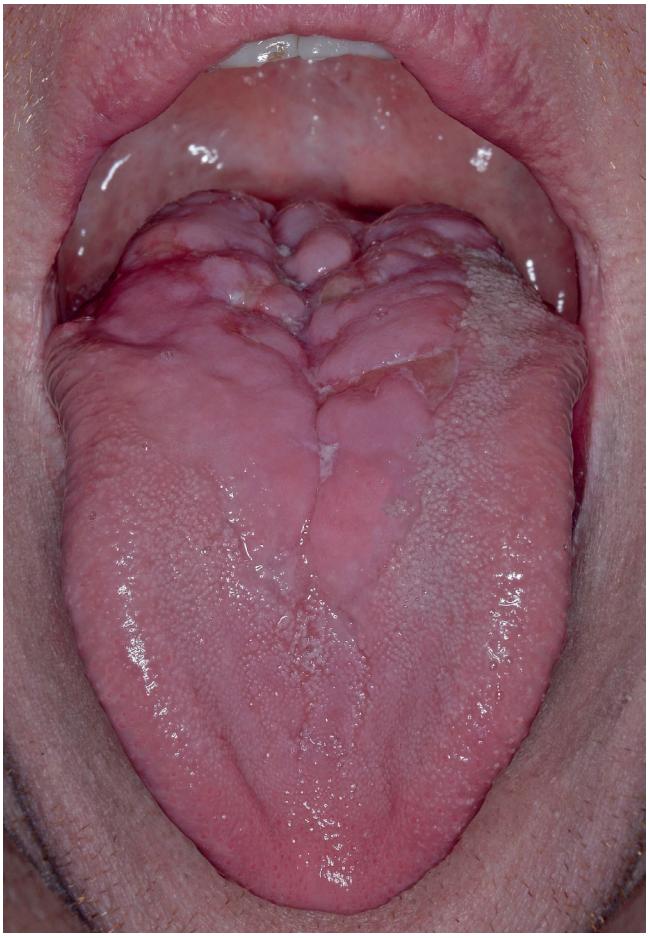

Figure 3. Three months post therapy.

inflammation', 'non-specific inflammation' and 'non-specific ulcer', as in the biopsy in this case (Setterfield et al, 2001; Bissonnette et al, 2017). The value of biopsy is partly in excluding more specific features such as granulomatous inflammation or vasculitis. Haematological investigations are useful to exclude infectious causes and to investigate haematological disorders. Erythrocyte sedimentation rate and C-reactive protein levels have been found to be elevated.

The differential diagnosis included mucosal tuberculosis, granulomatosis with polyangiitis, squamous cell carcinoma, major aphthous ulceration and deep fungal infections. While the other diagnoses were excluded based on histopathology, major aphthous stomatitis was excluded based on the clinical appearance, location and duration. The diagnosis of pyoderma gangrenosum was based on the characteristic morphology and evolution of the lesion, non-specific chronic inflammation on biopsy and presence of Crohn's disease. A point score system has been proposed to assist with the diagnosis of oral pyoderma gangrenosum (Bissonnette et al, 2017).

Treatment of the underlying systemic condition, if any, is an integral part of management. Systemic corticosteroids are most commonly used and constitute first-line therapy. Immunosuppressive agents such as ciclosporin, tacrolimus, azathioprine and cyclophosphamide have also been trialled to induce a prolonged remission period (Bissonnette et al, 2017). Infliximab and adalimumab have been suggested as second-line treatment for refractory multifocal lesions or cases of multi-organ involvement. When used in inflammatory bowel disease, the prevalence of anti-infliximab antibodies was 45.8% when episodic infusions of infliximab were given and 12.4% when maintenance infliximab was given (Lee et al, 2012). Local ulcer care is beneficial to enhance patient comfort and prevent secondary infection. Chlorhexidine 0.2% mouth rinse, clobetasol dipropionate 0.05% or tacrolimus 0.1% ointment can be used as adjuvant therapy. Recurrence of pyoderma gangrenosum has been documented in 10% of cases.

Learning points

- Pyoderma gangrenosum of the oral cavity is rare. Clinicians should be aware of this condition when a patient presents with a chronic ulcer with a background systemic disease.
- Biopsy is necessary to exclude other conditions that may present similarly including squamous cell carcinoma, major aphthous ulceration, mucosal tuberculosis, granulomatosis with polyangiitis, and deep fungal infections.
- Early diagnosis, management and follow up are essential because of the morbidity associated with these lesions and significant risk of relapse.

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