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Case report

Pyoderma gangrenosum of the eyelid associated with inflammatory bowel disease

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ABSTRACT

Purpose: Pyoderma gangrenosum (PG) of the eyelid can be difficult to diagnosis and may mimic other, more common pathologies, thereby delaying proper treatment and management. PG may be associated with systemic disorders that have significant comorbidities.

Observations: The authors present two cases of pyoderma gangrenosum of the eyelid associated with inflammatory bowel disease.

Conclusions and importance: This case series highlights the importance of early recognition of eyelid pyoderma gangrenosum to avoid local and systemic comorbidities with timely and appropriate management.

1. Introduction

Pyoderma gangrenosum (PG) is an inflammatory, ulcerating condition characterized by skin lesions with a predilection for the lower extremities and abdomen.[1] Typically, there are sterile pustules and ulcers that spread within the dermis and lead to necrosis of the epidermis.[2] The pathophysiology is believed to be related to an exaggerated recruitment of neutrophils to the dermis that can eventually replace the dermal layer in late stages.[3] Diagnosis is made by first excluding other possible causes of skin ulceration followed by a detailed history and supportive histopathology and immunohistochemistry.[2] PG is associated with systemic disorders such as myeloproliferative disorders, diabetes, rheumatoid arthritis, granulomatous hepatitis and inflammatory bowel disease in about half of the cases and therefore, when diagnosed, an extensive workup must be performed.[2,4]

2. Findings

2.1. Case 1

A 63 year old male with a history of recurrent ulcerations and presumed cellulitis on bilateral lower extremities presented to the emergency room with right upper eyelid edema and erythema that progressed rapidly over one week. Despite systemic and topical antibiotics, the eyelid became necrotic (Fig. 1A). A new lesion on the chin prompted biopsy of both locations, which demonstrated sheets of neutrophils in the dermis suggestive of pyoderma gangrenosum (Fig. 1B,C,D). He underwent systemic workup to evaluate for malignancy, vasculitic processes, and rheumatologic conditions. A positron emission tomography/computed tomography scan revealed colonic thickening which was further investigated with a colonoscopy. The colonoscopy revealed colitis with ulceration and neutrophilic inflammation consistent with Crohn's disease (Fig. 1E). Upon treatment with systemic steroids, there was brisk improvement in his eyelid condition (Fig. 1F). He has since been transitioned to infliximab, an anti-TNF agent, with good response and continues to do well at 6 months follow up.

2.2. Case 2

A 66-year-old male with a history of ulcerative colitis and ankylosing spondylitis presented with lacrimal gland inflammation for 1 year. A negative workup prompted transcutaneous incisional biopsy of the gland which showed nonspecific inflammation. Seven weeks after that biopsy he developed a full thickness defect in his eyelid (Fig. 2A), and a biopsy of the skin in this area was consistent with pyoderma gangrenosum. The eyelid inflammation and defect healed over 6 weeks with systemic steroids (Fig. 2B), and he was transitioned to adalimumab

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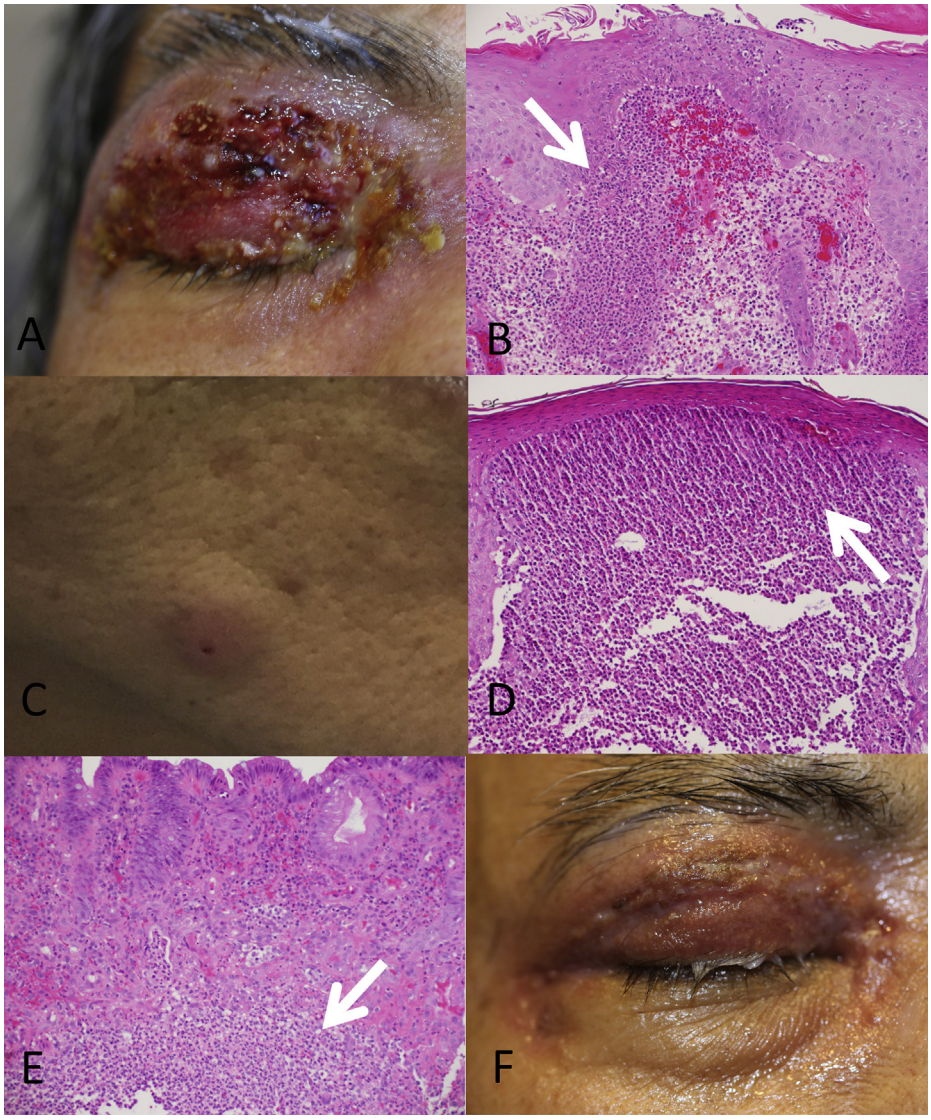

Fig. 1. *A*– Initial clinical presentation with right eyelid ulceration and necrosis. *B* – Histopathology of eyelid ulceration (20x) demonstrates collection of neutrophils within the dermis (arrow) and epidermal acanthosis. *C*– Clinical photograph of the indurated chin lesion. *D*– Histopathology of chin lesion (20x) reveals dense sheets of neutrophils in the dermis (arrow). *E*– H&E stain of transverse colon biopsy (20x) shows active colitis with ulceration, neutrophilic infiltration (arrow) and architectural changes. *F*– Clinical improvement in the eyelid after initiation of steroids.

weekly followed by rituximab every 6 months with sustained response.

3. Discussion

Pyoderma gangrenosum lesions of the eyelid are rare and are often misdiagnosed as infection, chalazion or tumor at initial presentation.[3,4,6] Incorrect diagnosis and management can lead to significant morbidity especially when there is a delay in the diagnosis of systemic illnesses.[2] Furthermore, PG is associated with significant pathergy and erroneous treatment can inadvertently make the condition worse.[1] Our second case demonstrates the pathergy associated with this condition as the patient necessitated steroid therapy for the skin incision to heal. The second case is unique in that the lesion was lateral and traditionally eyelid lesions have been reported to be central or medial. This may be related to the location of the incision site.

Case reports of PG involving the ocular adnexa have been associated with rheumatoid arthritis, rhinosinusitis, inflammatory bowel disease, and diabetes.[1,4–6] A single case of eyelid PG has been reported in association with ulcerative colitis and none have been reported with Crohn's disease.[4] There has been a single case of orbital pyoderma gangrenosum in association with Crohn's disease[7] but to date our case is the first reported eyelid manifestation. Non-eyelid PG is present in 0.5–5% of patients with ulcerative colitis and it coincides with flares.[4]

When evaluating an aggressive process of the eyelid, it is important to take a detailed review of systems and to consider biopsy early when the response to antibiotic therapy is not as expected. Patients often have a history of previous skin lesions as in the example of our first case.[2] Prompt diagnosis improves the prognosis in this condition and allows identification of comorbid systemic disorders.

Patient consent

Written informed consent was obtained from patients for publication of the case report as well as imaging.

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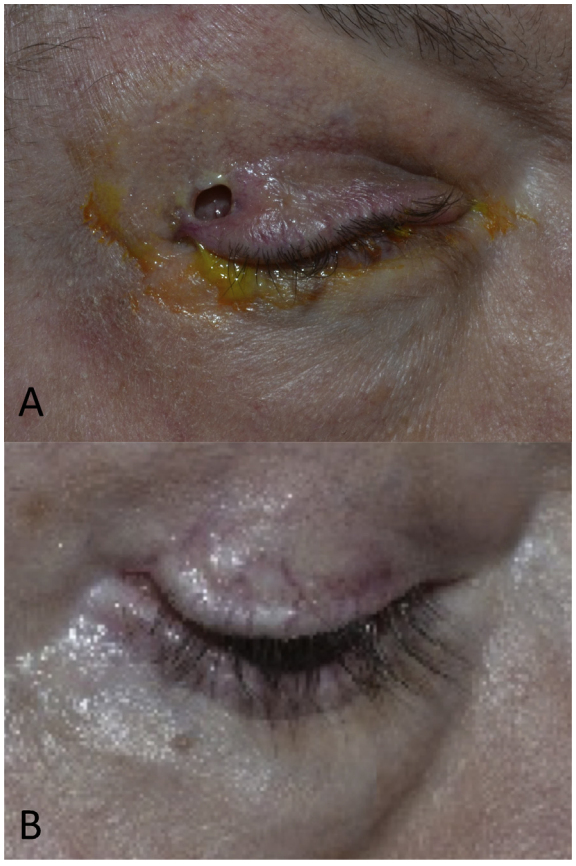

Fig. 2. *A* – Full thickness defect of the right upper eyelid after lacrimal gland biopsy. *B* – Well healed defect after initiation of systemic steroids.

Authorship

The authors attest that they meet the current ICMJE criteria for Authorship.

Author contributions

Larissa Habib: responsible for writing the manuscript.

Natalie Wolkow: responsible for helping with the photo editing of the pathology slides and the clinical photos.

Liza Cohen: resident responsible for primarily caring for the patients as an inpatient and editing of manuscript.

Lina Ma: pathologist responsible for describing and diagnosing the pathology.

Michael Yoon: attending supervising the clinical plan and treatment of the second patient.

Nahyoung Grace Lee: senior author responsible for and formal analysis and conceptualization.

Declaration of competing interest

The authors have no financial disclosures.

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