Atypical Case of Pyoderma Gangrenosum with Associated Secondary Bacterial Infection

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Introduction

Pyoderma gangrenosum is a rare condition that's characterized by the presence of large, necrotic ulcer(s) with red or violaceous borders that are typically surrounded by a halo of erythema. This condition is most associated with ulcerative colitis and, less often, Crohn's disease. However, 30-50% of cases are idiopathic and are thought to constitute a form of neutrophil-dominant autoimmune reaction. Histology often is nonspecific, most commonly manifesting as neutrophilic infiltrates in the dermis.

Case Description

This is the case of an otherwise healthy 30-year-old female who presented to the clinic with a 1-year history of intermittent right-sided jaw pain, swelling, and associated overlying skin lesion. Physical exam revealed a 3 x 4 cm, firm mass originating at the right mandible as well as a 2 x 3 cm, hyperkeratotic, pigmented external skin lesion (Figure 1). CT scan revealed erosion of the intraoral mass into the mandible and external cheek skin along with several prominent level 1 lymph nodes, raising concern for a malignancy. Initial biopsy was negative for neoplasia and demonstrated non-specific evidence of inflammation. Acute enlargement of the skin lesion after an injury prompted further inpatient workup (Figure 2A). CBC, CMP, ANCA panel, and Quaniferon Gold were all normal. However, CRP/ESR were elevated, and initial wound cultures were positive for *Staphylococcus aureus.* Follow-up biopsy showed abundant inflammatory cells, including lymphocytes, histiocytes, plasma cells, and neutrophils. This was highly suggestive of pyoderma gangrenosum. While outstanding microbiology studies were negative at time of discharge, there was still concern for *Actinomyces* infection, as the causative organism can be particularly difficult to grow. After appropriate treatment for both PG and associated bacterial infection, the facial lesion and intraoral mass resolved at one month follow-up (Figure 2B).

Figures

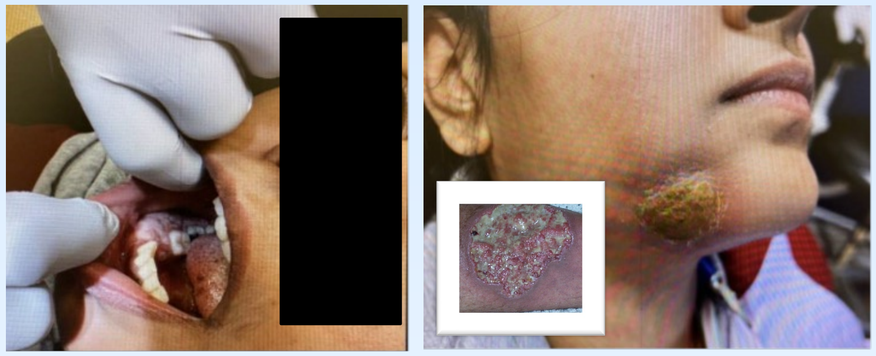

Figure 1: (1A: Left) Intraoral exam revealing 3 x 4 cm firm, flesh-colored mass originating at the right mandible, near teeth #25-29. (1B: Right) External skin exam revealing 2 x 3 cm, hyperkeratotic, pigmented lesion. (Inset) Representative image of a typical case of pyoderma gangrenosum.

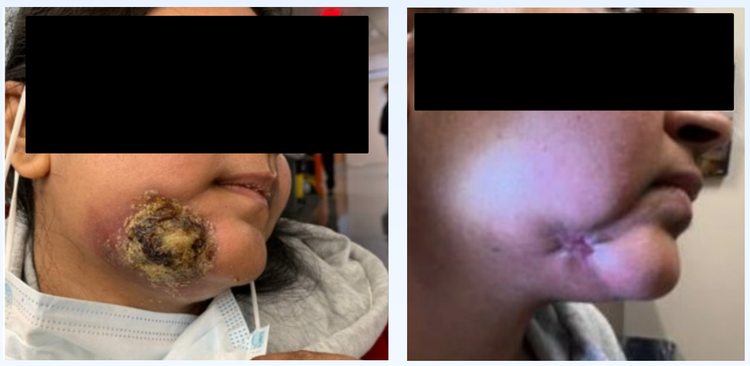

Figure 2: (2A: Left) Photograph showing worsening of the skin lesion after physical injury. (2B – Right) Resolution of the external skin lesion at 1 month follow-up.

References

1. Ruocco E, Sangiuliano S, Gravina AG, Miranda A, Nicoletti G. Pyoderma gangrenosum: an updated review. J Eur Acad Dermatol Venereol. 2009 Sep;23(9):1008-17. doi: 10.1111/j.1468-3083.2009.03199.x. Epub 2009 Mar 11. PMID: 19470075.
2. Wollina, U. Pyoderma gangrenosum – a review. Orphanet J Rare Dis 2, 19 (2007). https://doi.org/10.1186/1750-1172-2-19
3. Hayes, Robert C, and Anne Curtis. "Pyoderma gangrenosum with a contiguous erosion of the distal ulna." Journal of cutaneous medicine and surgery vol. 8,3 (2004): 162-5. doi:10.1007/s10227-004-0108-6
4. Gavioli, Elizabeth M. PharmD; Casias, Michael PharmD; Ngo, Linda PharmD. Pyoderma Gangrenosum and Superimposed Infection: A Case Report. Advances in Skin & Wound Care 33(6):p 1-3, June 2020. | DOI: 10.1097/01.ASW.0000661796.90753.f3

Discussion

- Pyoderma gangrenosum most commonly presents on the lower extremities, although it has been reported in other sites, including the face. Pyoderma gangrenosum typically begins as a deep nodule or superficial pustule that becomes necrotic and ulcerates. This case represents an atypical form of pyoderma gangrenosum, as it did not initially present as an ulcer, but a solid mass that extended into the mandible and external cheek skin. It is quite rare for pyoderma gangrenosum to spread to adjacent bone.
- Approximately 20% of cases exhibit a Koebner phenomenon, where accidental trauma can cause additional lesions. In this case, the lesion had a waxing and waning course, worsening with external or intraoral trauma.
- Pyoderma gangrenosum is also classically associated with other inflammatory pathologies, especially Crohn's disease and ulcerative colitis.
- Pyoderma gangrenosum can take one of two clinical courses, with either rapid development and spread of lesions or slow and indolent progression. The latter clinical history combined with lymphadenopathy, which is generally atypical in this condition, initially increased clinical suspicion for a neoplasm.
- Diagnosis of pyoderma gangrenosum is contingent on ruling out other causes of skin ulceration, as the histopathology of this condition is not definitive and often changes with different stages of the disease.
- There are a limited number of case reports of pyoderma gangrenosum associated with superinfection. However, in all these cases, patients developed secondary infections after being treated with immunosuppressive medications. This patient's chronic clinical course and slight improvement on antimicrobials prior to starting immunosuppressive medications suggests associated superinfection.

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