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Pyoderma Gangrenosum at Uncommon Site in a Pediatric Patient: Case Report and Review of Literature

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Authors' contributions

This work was carried out in collaboration among all authors. Authors ZM and SS designed the study, performed the statistical analysis, wrote the protocol and wrote the first draft of the manuscript. Authors BB, MM, NI, LB and KS managed the analyses of the study. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

A 12-year-old girl presented with three ulcerated lesions on her face that were red, swollen, had advancing borders, and undermined. Despite treatment with intravenous and topical antibiotics, the ulcers did not improve, and cultures showed no growth of organisms. The patient did not have any underlying systemic disease, and a skin biopsy revealed neutrophilic dermatitis. Oral prednisone was started, which resulted in the healing of the lesions. The skin biopsy and positive response to corticosteroids confirmed the diagnosis of Pyoderma Gangrenosum (PG). PG is a rare inflammatory skin condition, and facial lesions are a rare presentation. Early initiation of immunosuppressive therapy is crucial for complete healing and to minimize the significant psychological impact.

Keywords: Face; pediatric; pyoderma gangrenosum.

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1. INTRODUCTION

“Pyoderma gangrenosum (PG) is a type of inflammatory skin disease characterized by the presence of sterile neutrophils. It was first reported by Brunsting et al. in 1930 and occurs mainly in adults” [1]. “PG is known for its locoregional aggressiveness and tendency to recur chronically.PG is a rare cause of cutaneous ulceration in children, accounting for 4% of cases worldwide” [2]. “There are five subtypes of pyoderma gangrenosum (PG) based on clinical presentation, namely peristomal, pustular, bullous, vegetative, or classic ulcerative form” [3,4]. “PG commonly affects the lower extremities but can also appear on other parts of the body, such as the trunk, upper extremities, head, and neck”[5,6]. Here, we report an atypical presentation of a PG of the face in a child

2. PRESENTATION OF THE CASE

A 12-year-old child, with no previous medical history, consulted for the spontaneous appearance of three painful ulcerating lesions on the face that had been evolving for 1 month. Despite treatment with antibiotics, there was no response. The girl had no systemic, gastrointestinal, or arthritic symptoms.

The clinical examination revealed two ulcers on both cheeks, one measuring 7x4 cm and the other 5x3 cm. The ulcers were well-defined with soft, erythematous raised edges. The bottom of the ulcers was fibrinous and topped with hemorrhagic crusts that bled on contact and were very painful. There was also an ulceration measuring 3x2 cm in the right temporal region with the same appearance. The rest of the examination was unremarkable (Figs. 1a,1b).

Fig. 1a. Two ulcerating-bourgeoning lesions of the right hemiface

Fig. 1b. One ulcerating lesion on the left hemiface

There was no evidence of acetowhite organisms or fungal infection on acetowhite staining and periodic acid Schiff staining. In addition, bacterial, fungal, and tuberculosis cultures from secretions, biopsy tissue, and blood were all negative. The patient's biological workup was normal and no systemic disease was found.

Histopathological examination of the edge of the ulcers showed an ulcerated epidermal coating with an underlying dermis containing a polymorphic inflammatory infiltrate rich in neutrophils (Fig. 2). There was no evidence of infection or malignancy, suggesting Pyoderma Gangrenosum.

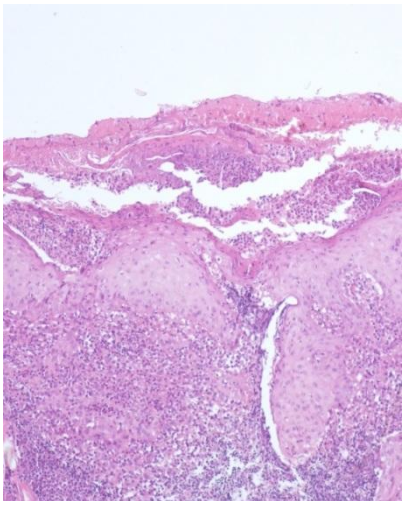

Fig. 2. Skin biopsy of the erythematous margins of the ulcer showing neutrophilic infiltration of the dermis (hematoxylin and eosin, ×200)

The patient received 1 mg/kg/day of prednisone, tapered off every 2 weeks. The ulcer size reduced with cribriform atrophic scars, indicating significant improvement. Almost complete healing was achieved after 6 months (Figs. 3a,3b)and After two years of observation, there were no new skin lesions observed. The patient's positive response to the therapy further supported the diagnosis of Pyoderma Gangrenosum (PG).

Fig. 3a, 3b. Lesions completely resolved with cribriform scarring and post-inflammatory hyperpigmentation after prednisone therapy (Right hemiface, left hemiface)

The patient did not have any gastrointestinal, musculoskeletal, or any other systemic complaints.

3. DISCUSSION

“Pyoderma Gangrenosum (PG) is a rare skin condition characterized by neutrophilic inflammation. It typically presents as a painful nodule, plaque, or pustule that expands and breaks down to form an enlarging ulcer with raised, undermined, and violaceous borders, surrounded by a zone of redness. PG lesions that heal can have a characteristic cribriform (perforated) appearance” [7]. “PG is estimated to affect up to 10 cases per million people per year, and represents up to 3% of chronic leg ulcer cases, but may occur anywhere on the body. PG has been reported in people of all ages; most cases, however, present in the second to sixth decades of life, with a possible female predominance” [8]. “A Delphi exercise identified one major criterion for Pyoderma Gangrenosum (PG): a biopsy of the ulcer edge demonstrating neutrophilic infiltrate. Additionally, eight minor criteria were established, which included exclusion of infection, pathology, a history of inflammatory bowel disease or inflammatory arthritis, the history of papules, pustules, or vesicles ulcerating within four days of appearing, peripheral erythema, undermining border, and tenderness at the ulceration site, multiple ulcerations, at least one on an anterior lower leg, cribriform or "wrinkled paper" scar(s) at healed ulcer sites, and decreased ulcer size within one month of initiating immunosuppressive medication(s). Our patient meets these criteria, and the analysis showed that four out of the eight minor criteria maximized discrimination, yielding a sensitivity and specificity of 86% and 90%, respectively”[9]. “Pediatric PG is easily overlooked and misdiagnosed due to its infrequent incidence and atypical involvement areas such as the head or face. The face is not the preferred location for Pyoderma Gangrenosum. In one of the largest case series of PG, only 7.8% occurred on the head or neck. The leg was the most frequent site of PG (77.7%), followed by the trunk (11.7%), and the upper extremities (8.7%)” [10]. “Pereira and colleagues found that the most common patients had a single lesion (62.5%), while multiple (more than three) lesions occurred only in 16.7% of the cases” [11]. “In a literature review of facial PG, few pediatric cases have been reported. Hilary Haimes and colleagues reported two pediatric cases of facial PG preceding the diagnosis of inflammatory bowel disease” [12]. “A similar location was recently described by Chen J et al in a 3-year-old girl. This girl presented with recurrent multiple painful ulcerative lesions for 10 months. The ulcer on the forehead rapidly progressed and involved both cheeks and nose” [13]. “Another article reported the case of a 3-year-old girl who developed PG on the left upper

eyelid without prior trauma or associated pathologies" [14]. In our case, the diagnosis of Pyoderma Gangrenosum (PG) was considered after ruling out other potential diagnoses due to negative mycological and bacteriological examinations, and the presence of suggestive histological findings. In addition, successful healing of the lesions with systemic corticosteroid therapy confirmed the diagnosis of PG. "Prompt diagnosis and treatment are essential to avoid severe scarring that can affect aesthetic and mental health. Notably, systemic comorbidities, such as inflammatory bowel disease, occurred in approximately 50% of PG cases in children" [13]. Regardless of the absence of symptoms of systemic comorbidities in our patient, further long-term multidisciplinary follow-up is necessary.

4. CONCLUSION

In summary, pediatric pyoderma gangrenosum can occur on any part of the body, including the face. A comprehensive evaluation is crucial in order to diagnose other potential causes and to assess any comorbidities related to the condition. Once the diagnosis is established, aggressive immunosuppression should be started to promote complete healing.

CONSENT

All authors declare that 'written informed consent was obtained from the patient (for publication of this case report and accompanying images).

ETHICAL APPROVAL

All authors hereby declare that all experiments have been examined and approved by the appropriate ethics committee and have therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

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COMPETING INTERESTS

Authors have declared that no competing interests exist.

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