Pyoderma gangrenosum associated with rheumatoid arthritis mimicking vasculitis

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Abstract

Pyoderma gangrenosum is a rare neutrophilic dermatosis characterized by painful, rapidly progressing necrotic ulcers. Although PG has been reported in association with various autoimmune diseases, its coexistence with rheumatoid arthritis (RA) mimicking vasculitis is uncommon. We present a case report of a 75-year-old female with a longstanding history of RA who developed ulcerative skin lesions that initially presented as vasculitis. She presented with new-onset painful, ulcerative skin lesion on the lower extremity, accompanied by malaise. Physical examination revealed asymmetric, deep necrotic ulcers with undermined borders. Laboratory investigations showed elevated acute phase reactants and negative rheumatoid factor. Skin biopsy demonstrated neutrophilic infiltration, confirming the diagnosis of PG. Further investigations, including imaging studies and vasculitis-specific laboratory tests, were negative, ruling out vasculitis. The patient was managed with systemic corticosteroids and immunosuppressive therapy, resulting in significant improvement of skin lesions. This case highlights the importance of considering PG as a potential differential diagnosis in patients with RA presenting with ulcerative skin lesions resembling vasculitis.

Keywords: pyoderma gangrenosum; Rheumatoid arthritis; Vasculitis; Laboratory investigations

1. Introduction

Pyoderma gangrenosum is a chronic inflammatory dermatosis, often linked with non-infectious diseases including rheumatoid arthritis. Common in elderly patients, this rare skin ulceration is often misdiagnosed as vasculitis. Differentiating between PG and vasculitis in the setting of RA can be complex, as both conditions share similar clinical and histopathological features.

2. Case Presentation

A 75-year-old female with history of rheumatoid arthritis presented with recurrent left lower extremity wound without a formal diagnosis (figure 1). Patient's ulcer was responding to ciprofloxacin and prednisone until she finished both courses two weeks prior to admission. She had a biopsy a month prior to presentation and attempted to follow up with rheumatology but was unable to be evaluated in outpatient setting. She had noticed worsening of the ulcer and presented to emergency department for further evaluation.

On admission, she was noted to have 7/10 lower extremity pain and leukocytosis. Left foot x-ray showed increased density in the subcutaneous fat which may be due to cellulitis. Lower extremity MRI did not show evidence of osteomyelitis. Given patient's biopsy was suggestive of venous insufficiency, vascular surgery was consulted for further evaluation. Patient had venous and arterial LE US which did not show sonographic evidence of DVT, stenosis or occlusion. ABI was not measured as patient could not tolerate due to severe pain. Patient was evaluated by rheumatology and started on prednisone and sulfasalazine. ESR and CRP were elevated, LRINEC score 6. Wound cultures resulted in Staphylococcus aureus. She had repeat biopsy of her chronic wound which confirmed pyoderma
gangrenosum. Given concern for Pseudomonas gangrenosum, patient was started on dapsone and topical tacrolimus. Patient was instructed to follow up with infectious disease and rheumatology.

3. Discussion

Numerous factors are understood to play a role in the pathogenesis of pyoderma gangrenosum; autoimmune activation of T cells, aberrant neutrophil behavior, and genetic mutations have all been implicated (1). Therapy is likewise varied, ranging from wound care, topical and systemic corticosteroids, and biologics, and judicious treatment selection may depend on factors such as lesion surface area and patient comorbidities. Presently, a gold standard therapy for treating pyoderma gangrenosum remains elusive (2). Retrospective studies suggest that pyoderma gangrenosum presents a substantially elevated risk of mortality for patients, whether or not an associated, underlying condition exists (3). This risk – along with the potential complications of debility, pain, superinfection, and disfigurement, among others – underscores the importance of early diagnosis and treatment (4). As pyoderma gangrenosum is currently identified primarily by exclusion, updated guidelines such as those proposed by Maverakis et al. may help pave the way forward to improved outcomes.

Our patient’s lesion was not resistant to treatment as she responded well to prednisone and sulfasalazine. More extensive disease is usually treated with systemic steroids and cytotoxic agents with good results (5). Autoinflammatory conditions, such as rheumatoid arthritis, can present with recurrent attacks of non-infectious inflammation at target sites (skin in our patient) without elevated levels of circulating autoantibodies (6). Our patient’s misdiagnosis of vasculitis was the culprit of her recurrent attacks as the lesion presents similarly to Pyoderma gangrenosum. It is not uncommon for the diagnosis of Pyoderma gangrenosum to be missed, as this lead to repeat surgical debridement of the patient’s left leg lesion. Our case highlights the need to exclude other causes that can present with similar lesions in a patient with Pyoderma gangrenosum.

Figure 1 Full thickness wound demonstrated on the left lower extremity.

4. Conclusion

The association between pyoderma gangrenosum and rheumatoid arthritis is well-documented, but the exact pathogenesis remains unclear. It is hypothesized that immune dysregulation and an underlying systemic inflammatory state contribute to the development of both conditions. The diagnosis of PG associated with RA requires a high index of suspicion, as it can often be mistaken for other cutaneous vasculitides. Further research is warranted to enhance our understanding of the pathophysiology, diagnostic modalities, and therapeutic options for this challenging condition.

Compliance with ethical standards

Disclosure of conflict of interest

The above listed authors have no conflicts of interest to declare.
Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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