

Pyoderma Gangrenosum: A Case Report.

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ABSTRACT

Pyoderma gangrenosum – a rare, neutrophilic dermatosis – is associated with diseases, including inflammatory bowel disease, arthritis and haematologic disease. It is a condition of unknown etiology. It is the result of an exaggerated response to specific and non-specific stimuli. First-line treatment is with a steroid, usually prednisone. Here we report a case of 60-year-old female presented to us with Pyoderma gangrenosum.

Keywords: Pyoderma gangrenosum; neutrophilic dermatosis; prednisone.

INTRODUCTION

Pyoderma gangrenosum (PG) is a rare, non-infectious, ulcerating, neutrophilic dermatosis, that most frequently affects the lower extremities of adults aged 25-54 years. There are four main clinical types of PG: (i) ulcerative; (ii) pustular; (iii) bullous; and (iv) vegetative. One more subtype peristomal PG is also known. These different variants of PG can be linked to specific associated conditions in approximately 50% of patients. The most common associated diseases are inflammatory bowel disease (IBD), arthritis and haematologic disease.^[1-3]

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CASE REPORT

A 60-years-old woman, a diagnosed case of psoriatic arthritis, came to the medical outdoor with chief complaints of an enlarging, erythematous ulcer on her right lower extremity from 3 weeks. According to the patient, the lesion first started as a sore with slightly elevated bruising on the right lower leg. Over a period of two weeks, this area deteriorated into large, exuding wounds, 2 ulcers of 15x7 cm and 6x8 cm with irregular border and underlying violaceous edematous edges. [Figure 1]. Floor showed black necrotic tissue (eschar), purulent discharge, and atrophic scars were present at the site of old healed lesion. There was no history of trauma or insect bite. The systemic examination of the patient was normal. Her hemoglobin was 7.2 gm/dl, total leucocyte count was 14800/ mm³ with neutrophils 88%. There was an elevated C-reactive protein (CRP) of 126 mg/L, and erythrocyte sedimentation rate of 68 mm/hour. The ulcers were initially treated as an infected venous ulcer, and the patient was given topical mupirocin and oral cefuroxime axetil. Despite

treatment, the lesion progressed in size to 10 x 10 cm, with central necrosis and purplish overhanging borders. The ulcers were clinically suspected of Pyoderma gangrenosum. She was prescribed 60 mg of prednisone and 400 mg of cyclosporine daily. The punch biopsy taken from the edge of the right lower-leg ulcer showed necrosis and inflammatory dermal infiltrates composed of mature neutrophils, suggestive of Pyoderma gangrenosum. Within 2 weeks, the inflammation dramatically subsided, and prednisone was tapered to 20 mg daily. Within a month, the prednisone was stopped, but the cyclosporine was continued. The lesion gradually healed to leave a hyper-pigmented, atrophic scar.



Figure 1: Right leg of the patient showing 2 big ulcers of Pyoderma gangrenosum.

DISCUSSION

Pyoderma gangrenosum (PG) lesions are described as sharply outlined, painful, necrotic ulcers, creating purulent and hemorrhagic exudates, and with undermined, purplish borders and a surrounding zone of erythema. PG is a clinical diagnosis; there is no specific histological or serological marker for the condition that can alone provide the diagnosis.

The exact etiology of Pyoderma gangrenosum is not known.^[4] Many hypotheses have been proposed, but attention has focused mainly on immune abnormalities and alterations in cell-mediated immune response.^[5]

It is thought that Pyoderma gangrenosum may be the result of a hypersensitive reaction of the immune system due to an altered, exaggerated and uncontrolled inflammatory response to specific and non-specific stimuli, leading to a neutrophilic vasculitis, which is characterized by perivascular deposition of immunoglobulin M (IgM), C3 and fibrin. Neutrophils appear to play a key role in the pathogenesis of Pyoderma gangrenosum. This is supported by the fact that the disease responds to therapies that have antineutrophilic activity. The systemic diseases associated with PG are seen in approximately 50% of the overall cases of PG, but in patients with the ulcerative type of PG, they occur in >70% of patients.^[6] Inflammatory bowel disease is most common PG-associated disease, followed by arthritis, haematologic abnormalities, and haematologic malignancies. Other associations are rare; these include hidradenitis suppurativa, pyogenic arthritis-Pyoderma gangrenosum-acne syndrome, pulmonary disease, systemic lupus erythematosus, thyroid disease, solid organ malignancy, autoimmune hepatitis, and sarcoidosis. The most common malignant disease associated with PG is haematologic malignancy.^[3] This is seen in up to 7% of PG cases - predominantly acute myeloid leukemia, which has a 1- year mortality rate as high as 75%. In our case PG was associated with psoriatic arthritis which is a rare association.^[7] When Pyoderma gangrenosum is associated with systemic disease, the search for underlying systemic disease should be made and treatment of underlying disease should be started.^[8] Treatment of lesions usually involves systemic treatment, together with local therapies.^[9] Systemic treatments include steroids such as prednisone 40-120 mg/day until healing.^[10] Cyclosporine (6 mg/kg) is a common therapy used either alone or in combination with steroids.^[11]

CONCLUSION

This case describes an unusual clinical scenario that suggests underlying systemic disease should be considered in PG cases. The increasing incidence of Pyoderma gangrenosum means that presentations associated with systemic diseases may become more common and clinicians should be aware of it.

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