

Pyoderma gangrenosum developed from aggravated pemphigus foliaceus after dog bite

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Dear Editor,

Pyoderma gangrenosum (PG) is a rare neutrophilic dermatosis that causes the development of painful ulcers on the skin, initially starting with nodules or abscesses, and quickly progressing at the peripheral regions into ulcers.¹ The exact cause is not yet known, and 20-30% of patients are said to experience a pathergy phenomenon in which lesions occur in the sites of previous trauma.² In addition, pyoderma gangrenosum is accompanied by systemic diseases, such as Crohn's disease or inflammatory colitis, rheumatoid arthritis, and hyperthyroidism.³ Although numerous reports indicate that pyoderma gangrenosum is associated with systemic dis-

eases,⁴ case reports on the association of pyoderma gangrenosum with skin diseases, especially blistering disorders, are very rare.

A 55-year-old man, diagnosed with pemphigus foliaceus (PF) at another hospital, had been controlling lesions by taking a low dose of oral steroids (methylprednisolone 6 mg/day) for seven years. He had a laceration resulting from being bitten in the left leg by a Jindo dog. After stopping steroids with debridement and antibiotics administrations, he presented multiple flaccid bullous lesions such as pemphigus recurred on the trunk, and then many painful erythematous and violaceous bullae, erosions, and ulcers were found to be distributed across the whole body, and blistering lesions were accompanied by erosive lesions or plaques with crusts. The lesion of the left leg that developed after being bitten by a dog was also accompanied by a purple border and ulcers (Figure 1).

Microbial culture tests, including a blood culture, bacterial and fungal cultures of the lesion of the left leg revealed negative results. A skin biopsy was performed on samples from the boundary of the ulcer on the left leg, as well as a blistering lesion on the right arm. In all the biopsies, there are diffuse infiltration of neutrophils in dermis (Figure 2), whereas no specific findings were observed in Grocott's methenamine silver and Periodic acid-Schiff stains. A direct immunofluorescence performed on the right arm also showed no specific findings.

Through the above clinical and histopathological manifestations, we diagnosed the patient with pyoderma gangrenosum developed from aggravated pemphigus foliaceus. Methylprednisolone aceponate was applied topically, and steroids were injected into the skin lesions. Furthermore, a combined systemic treatment with the oral steroids (1 mg/kg/day), Dapsone 100 mg and Azathioprin 100 mg was administered. Although the number of blistering lesions decreased and we observed improvement of the patient's overall condition, he died of aggravated pneumonia, septic shock, and disseminated intravascular coagulation due to a weakened immune system.

There are no clear diagnostic criteria for this disease to date, but Su *et al.* have proposed a potential diagnostic criterion, which states that for the diagnosis of PG, two main criteria and two or more sub criteria must be met.⁵ The main criteria include the observation of clear, irregular, purple ulcer boundaries; rapid progression of painful ulcers; and a lack of other potential factors that may cause ulcers. There are four sub-criteria: pathergy or cribriform scarring, systemic disease associated with PG, histological findings with neutrophil infiltration in the dermis, and response to systemic steroid treatment. Therefore, clinical examinations and histological tests can be useful to diagnose this disease, and it is important to identify extensive neutrophil infiltration in the dermis through histological tests at the boundaries of ulcers. In some cases, non-specific findings such as vasculitis and various inflammatory cells other than neutrophils may be observed, and histological tests, special stains, and microbial culture tests may be performed rule out the infectious disease.⁶ In this case report, two main criteria were met: the clinical pattern of ulcers, and a lack of

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other factors as the potential cause of ulcers. Furthermore, the patient showed a response to steroid treatment, and results of histological and special stains verified the diagnosis of PG. Microbial tests including bacterial and fungal cultures and a blood culture

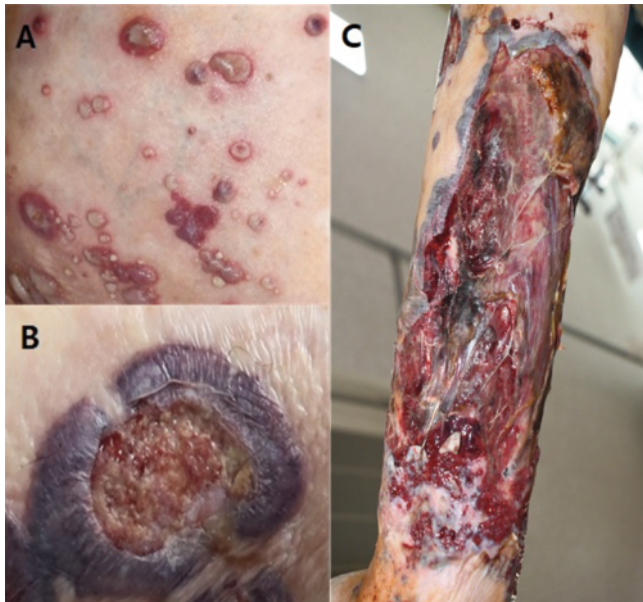


Figure 1. A) Multiple various-sized primary lesions of flaccid blisters on the trunk. B) A close-up view of painful lesions with peripheral violaceous undermined edges and central ulcers that originated from bullous lesions on whole body. C) Pyoderma gangrenosum showing the characteristic central ulcers and peripheral violaceous border after pathergy phenomenon occurred in the left leg due to dog bite trauma and debridement.

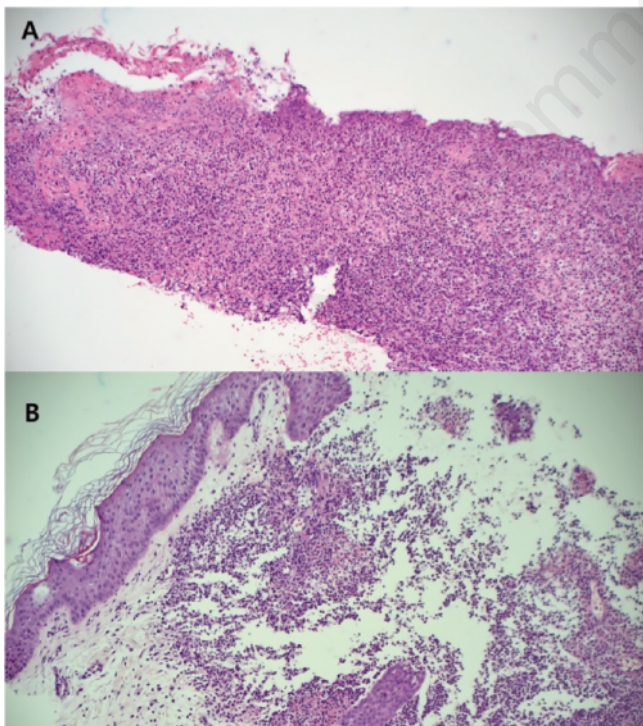


Figure 2. A) Epidermal necrosis and massive neutrophil infiltration in the dermis on the left leg (H&E, X100). B) Dermal edema and diffuse inflammatory cell infiltration composed of neutrophils in the dermis on right arm (H&E, X100).

test showed negative results, which contributed to the diagnosis of PG.

Pathergy phenomenon is the appearance of skin lesions in a prior trauma area, and pathergy may occur with pyoderma gangrenosum.⁵ Physicians often overlook pathergy phenomenon, so they misdiagnose it as an infectious disease and decide to perform debridement. Pathergy occur after debridement, and lesions may be further exacerbated.²

Treatment of PG should be combined with the treatment of accompanying systemic diseases. Daily disinfection with physiological saline solution to reduce pain and prevent secondary bacterial infections can be helpful, along with the use of topical antibiotics. In addition to topical application of strong steroids, intralesional triamcinolone injections can be effective, and systemic steroids are most commonly used.⁴

We present a rare and interesting case of a 55-year-old patient diagnosed with PF who developed PG through aggravated lesions upon discontinuation of steroids. We comment that clinicians should be aware of the potential for PG to develop previous PF patients who sustain an injury. As in this case, a biopsy must be performed for the accurate diagnosis, and it should be treated correctly according to the diagnosis. The authors report this rare and interesting case in the context of a literature review.

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