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Recurrent aseptic abscesses resulting in superficial pyoderma gangrenosum-like ulcers in a patient with granulomatosis with polyangiitis

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Abstract

Patients with granulomatosis with polyangiitis occasionally present with cutaneous manifestations, which are important clues for the early diagnosis. Although pyoderma gangrenosum-like ulcers are rarely observed, a unique case with unusual clinical features is presented herein. A 75-year-old woman with positive proteinase 3-antineutrophil cytoplasmic antibody (PR3-ANCA) repeatedly developed aseptic abscesses on the abdomen, buttock, lower legs, and forearms. Histopathological features of biopsy taken from ulcers showed necrotizing granulomatous inflammation with multinucleated giant cells without leukocytoclastic vasculitis. The present case was initially diagnosed as limited granulomatosis with polyangiitis without renal and lung involvement. However, two years later, she died of cerebral hemorrhage.

Keywords: aseptic abscess, polyangiitis, pyoderma gangrenosum, Wegener granulomatosis

Introduction

Cutaneous manifestation is observed in up to 50 % in granulomatosis with polyangiitis (GPA), [1], which histopathologically shows leukocytoclastic vasculitis, granulomatous vasculitis, and extravascular granuloma. Among the specific skin manifestations, palpable purpura is the most common and other lesions such as subcutaneous nodules, papules, bloody vesicles, ulcers, livedo, and gingival hyperplasia are also observed.

Case Synopsis

A 75-year-old woman was admitted to our hospital, under suspicion of GPA based on findings of refractory sinusitis, polyarthralgia, and positive proteinase 3-antineutrophil cytoplasmic antibody (PR3-ANCA). The patient had a past history of diabetes mellitus but detailed examination did not reveal either lung or kidney involvement. Moreover, neither asthma nor peripheral blood eosinophilia was observed. The patient recurrently developed several asymptomatic reddish abscess-like nodules on the abdomen, buttock, lower legs, and forearms (**Figure 1A**). During the course, some of the nodules spontaneously ruptured, leaving superficial ulcerative plaques and cribriform scars (**Figure 1B**). Bacterial and mycobacterial cultures were repeatedly performed, all of which were sterile.

Histopathological examination showed a nodular mixed inflammatory infiltrate with abscess formation in the dermis and subcutaneous tissues (**Figure 2A**). Higher magnification revealed granulomatous inflammation infiltration of neutrophils,

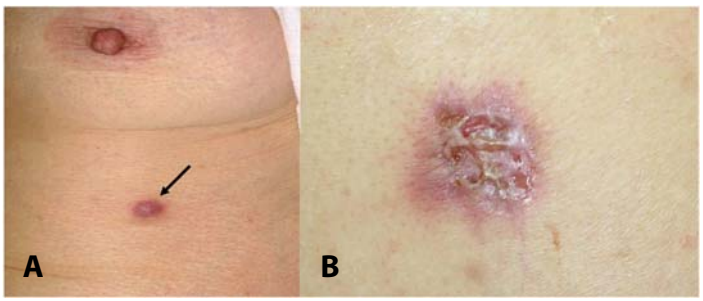

Figure 1. ***A)*** *Clinical appearance showing abscess-like nodules on the abdomen.* ***B)*** *Some nodules leave superficial ulcerations after rupture, forming cribriform scars.*

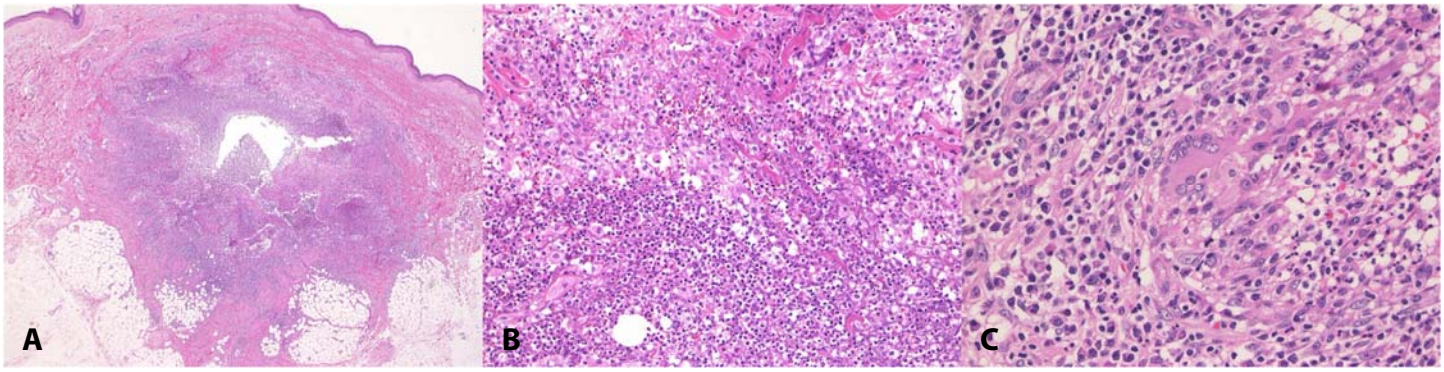

Figure 2. *H&E histopathology **A)** shows a nodular mixed inflammatory infiltrate with abscess formation in the dermis and subcutis, 40×. **B)** Higher magnification shows red blood cell extravasation, infiltrates of a number of neutrophils and histiocytes, and degenerated collagen, 200×. **C)** Multinucleated giant cells were scattered in the dermis, 400×.*

mononuclear cells, and histiocytes and some red blood cells (**Figure 2B**). Degenerated basophilic collagen and multi-nucleated giant cells were also observed (**Figure 2C**). Leukocytoclastic vasculitis was not observed. Ziehl-Neelsen stain was negative. Although the patient was treated with oral prednisolone, she died of cerebral hemorrhage two years later. Autopsy was not performed.

Case Discussion

Pyoderma gangrenosum (PG)-like ulcers are rarely observed in GPA, which histopathologically shows hemorrhagic necrotizing granulomatous inflammation with giant cells and collagen degeneration [2-4]. It has been suggested that cases presenting with purpura show leukocytoclastic vasculitis and are associated with active disease with renal and pulmonary involvement, whereas cases presenting with PG-like ulcers do not show vasculitis but granulomatous inflammation and are slowly progressive with less frequent involvement of internal organs [1,2]. The present case was initially diagnosed as limited GPA without renal and lung involvement.

Among cases of PG-like ulcers, several patients develop severe, large, and deep lesions with advanced necrosis and destruction of the surrounding tissues, predominantly involving the head and neck area, sometimes termed malignant pyoderma [5]. In the present case, neither facial nor cervical involvement were observed. Multiple abscesses or cellulitis-like nodules appeared, which then ruptured and resulted in superficial PG-like ulcers, leaving cribriform scars. Histopathological features showed necrotizing granulomatous inflammation with multinucleated giant cells but without leukocytoclastic vasculitis. Cases presenting with such clinical appearances and courses are rare. Although the differentiation between PG and PG-like ulcers in GPA is sometimes difficult [6], we excluded PG based on findings such as unique clinical course (ulceration following rupture of abscess-like nodules), necrotizing granulomatous inflammation containing a number of histiocytes, and the presence of PR3-ANCA.

Conclusion

Our unique case suggests that PG-like lesions vary between severe aggressive large, deep ulcers and superficial small ulcers with cribriform scars. Even if GPA cases are considered indolent at the time of diagnosis, careful follow-up is necessary.

Potential conflicts of interest

The authors declare no conflicts of interest.

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