Pyoderma Gangrenosum Mimicking Necrotizing Fasciitis on Magnetic Resonance Imaging: A Case Report and Literature Review

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Patient: Female, 67-year-old
Final Diagnosis: Pyoderma gangrenosum
Symptoms: Purpura
Medication: —
Clinical Procedure: —
Specialty: Hematology

Objective: Unknown etiology

Background: Pyoderma gangrenosum (PG) is a sterile neutrophilic dermatosis that can be associated with systemic diseases, such as ulcerative colitis, polyarthritis, diabetes mellitus, myelodysplastic syndrome, and/or myeloid leukemia, and is often misdiagnosed as a necrotizing infection. Few reports have described imaging studies of PG; however, necrotizing fasciitis (NF) exhibits distinct imaging characteristics. If deep fascial involvement is not demonstrated on magnetic resonance imaging (MRI), NF is excluded.

Case Report: We present a case of PG mimicking NF on MRI in a 67-year-old woman with acute myeloblastic leukemia. After undergoing a second cycle of decitabine therapy, she was admitted for pain in her lower left leg. The condition was initially misdiagnosed as NF because MRI findings demonstrated signal intensity in the fascia. MRI revealed fasciitis that exhibited linear fluid signal intensity in the fascia of the lower left leg. Despite broad-spectrum antibiotics, the lesion rapidly progressed to a swollen hemorrhagic patch with bullae and an ulcer. Skin biopsy results ultimately led to the diagnosis of PG, based on histopathological findings. The patient was treated with intravenous steroids and regular wound dressing. The skin lesion on the lower left leg exhibited a good response.

Conclusions: Despite the presence of a lesion that invaded the fascia on MRI, our patient was diagnosed with PG following a skin biopsy and completely recovered with steroid treatment. To distinguish PG from NF, it is more important to identify the characteristic clinical features than to rely solely on imaging findings.

Keywords: Magnetic Resonance Imaging • Fasciitis • Pyoderma Gangrenosum

Abbreviations: PG – pyoderma gangrenosum; NF – necrotizing fasciitis; MRI – magnetic resonance imaging

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Background

Pyoderma gangrenosum (PG) is an aseptic neutrophilic dermatosis that can be related to systemic diseases, such as ulcerative colitis, polyarthritis, diabetes mellitus, and hematologic malignancy. The pathogenesis of PG, however, is poorly understood. Rapidly progressing PG is often misdiagnosed as a necrotizing infection requiring urgent surgical intervention. The surgical debridement of lesions complicates the initial pathogenic phenomenon, accelerating the necrotic process [1,2]. Failure to reevaluate the diagnosis after repeated attempts at the procedure for nonhealing ulcers leads to exacerbation of the disease and increases the infectious risk, with devastating patient morbidity. It would be useful if there were radiological findings that distinguished PG from necrotizing infection; however, there are few reports of such cases to date. We report a case in which the magnetic resonance imaging (MRI) findings showed a high possibility of necrotizing fasciitis (NF), but was ultimately confirmed as PG. The Institutional Review Board of Gyeongsang National University Hospital approved this retrospective case study and waived the requirement for informed consent.

Case Report

The purpose of this case report was to identify any imaging findings that can distinguish between PG and NF. A 67-year-old woman who visited the hospital with symptoms of exertional dyspnea and fatigue was diagnosed with acute myeloblastic leukemia with mutated NTM1 with FLT3-ITD, a high allelic ratio known as an intermediate to poor prognostic factor. She denied any previous medical history and smoking, alcohol, and substance abuse. After undergoing a second cycle of decitabine therapy (20 mg/m² per day for 5 days), the patient was admitted for pain in her lower left leg. Physical examination revealed erythematous swollen purpuric macules on the lower limbs. This was accompanied by tenderness and a burning sensation in the lower left leg. The patient had a fever with a maximum temperature of 39°C for 3 days. Blood pressure, pulse rate, and respiratory rate were 120/60 mmHg, 100 beats per min, and 16 breaths per min, respectively. Laboratory investigations revealed the following: leukocyte count, 2.21×10⁹/L (reference range 4.0-10.0); neutrophils, 1.08×10⁹/L (range 1.5-7.5); hemoglobin, 7.8 g/dL (range 12.0-16.0); hematocrit, 23.0% (range 36-48); platelets, 89×10⁹/L (130-400); C-reactive protein level, 85.7 mg/L (range 0-5); D-dimer level, 1.99 mcg/mL (range 0-0.5); and fluorescent antinuclear antibody titer, negative. On the day of MRI, which was performed 3 days after hospitalization, the patient’s skin lesions had progressed to an erythematous swollen hemorrhagic patch with bullae on the lower left leg area. MRI revealed fasciitis that exhibited linear fluid signal intensity in the fascia of the left lower leg and a diffuse reticular high signal intensity in the subcutaneous fat layer of the same region of the leg (Figure 1). Despite intravenous administration of broad-spectrum antibiotics (cefazolin 2 g twice daily for 10 days), the patient’s condition did not improve, and extensive necrotic ulceration involving the lower left leg developed. The patient had to undergo surgical debridement of the necrotic tissue and was discharged with a healing ulcer.
a day for 3 days followed by Maxipime 2 g 3 times a day for 3 days, vancomycin 1 g twice a day, and clindamycin 600 mg 3 times a day for 6 days), the lesion rapidly progressed to a swollen hemorrhagic patch with bullae and a large ulcer with a maximum diameter of 1 cm. Five days after hospitalization, purplish plaques with central flaccid bullae measuring a maximum of 5 cm in diameter (Figure 2) were observed, which subsequently became erosive. A skin biopsy of the lesion was done and revealed ulcer and diffuse infiltration of neutrophils in the dermis (Figure 3A). Some blood vessels showed vasculopathy with fibrinoid necrosis, but there was no occlusive vascular thrombotic change (Figure 3B). Gomori methenamine silver stain showed no fungal organisms. Skin culture yielded no microorganisms. Considering these results, the patient was diagnosed with PG, based on histopathological findings. Intravenous methylprednisolone (methylprednisolone sodium succinate, 50 mg for 2 weeks and tapering afterward) was administered for 1 month in total, and regular wound dressing was performed. There were no adverse effects of steroid use in the patient. The skin lesion of the left lower leg exhibited a good response to the treatment. The hemorrhagic patch on the leg disappeared completely. The patient was discharged after 1 month of methylprednisolone administration.

**Discussion**

Necrotizing fasciitis (NF) is usually characterized by persistent severe pain, development of blisters due to vessel occlusion in the fascia and muscularis layers, necrosis and ecchymosis of the affected skin, gas in the soft tissues detected by either palpation or imaging studies, edema which can extend beyond the erythematous borders, sensory paralysis of the skin, symptoms of systemic toxicity (including fever, leukocytosis, delirium, and renal failure), and lastly, by rapid disease progression despite administration of appropriate antimicrobial therapy [3]. Additionally, NF exhibits distinct imaging characteristics. Gas from subcutaneous tissue can be observed on simple radiography [4], and computed tomography (CT) reveals edema, hypertrophy, and contrast enhancement in the fascia [4,5]. MRI is the most recommended modality for identifying NF, demonstrating 90% to 100% sensitivity and 50% to 85% specificity [6]; therefore, we immediately resorted to performing an MRI without CT or ultrasound, given the advancing rapid deterioration of the patient’s condition. Low signal intensity in soft tissues on T1-weighted imaging and high signal intensity in the soft tissue/fascia in T2-highlighted images are accompanied by contrast enhancement [5,6]. The criterion standard for diagnosis of NF is identification of the fascial involvement.
| Author    | Age/sex | Initial diagnosis                  | MRI finding                                                                 | Histology                                                                 | Treatment and outcome                                                                                           |
|-----------|---------|-----------------------------------|------------------------------------------------------------------------------|--------------------------------------------------------------------------|-----------------------------------------------------------------------------------------------------------------|
| Lee [14]  | 47/M    | Diabetic foot infection           | Cellulitis with myositis but no assertive abscess pocket                      | Neutrophil infiltrate in whole field with absence of squamous carcinoma or other malignancy | – 2-week course of prednisolone injections  
– After 9 months of follow-up, no evidence of new necrosis or deterioration was found, and the foot was in good condition |
| Yoshida [15] | 67/M | Pyoderma gangrenosum and sterile osteomyelitis | Low intensity with T1-weighted phase and high intensity with T2-weighted phase in the sternum and the overlying fat and skin tissues, indicating severe inflammation | Epidermal ulceration with prominent infiltration of neutrophils | – Steroid pulse therapy  
– Only partially effective, and patient died of respiratory failure |
| Fonder [16] | 38/F | Pyoderma gangrenosum               | Superficial T2 bright signal compatible with inflammation involving the subcutaneous tissue down to the level of the deep fascia | Necrosis, a mixed inflammatory infiltrate, and a small-vessel leukocytoclastic vasculitis | – Resistant to multiple treatment regimens. Adalimumab was then initiated in combination with cyclosporine, prednisone, and sulfasalazine  
– The ulcer and gastrointestinal disease responded rapidly |
| Jain [17]  | 70/F    | Cellulitis                         | Wound present medially in the distal calf with adjacent skin thickening along with circumferential subcutaneous edema | Severe acute cellulitis, abscess formation, and granulation tissue        | – Oral prednisone 100 mg and high-potency topical steroids  
– After a couple of days, there was a marked reduction in erythema and ulcer showed signs of healing |
| Milind [18] | 49/M | Disseminated tuberculosis          | Small fluid collection adjacent to right ischial tuberosity suggesting an infective etiology | Perivascular lymphocytic infiltration with endothelial swelling and focal neutrophilic abscess | – Two pulses of dexamethasone cyclophosphamide with intervening low-dose cyclophosphamide  
– Finally, complete healing occurred |
| Husain [19] | 55/M | Ulcerative colitis                | A complex appearing, peripherally enhancing fluid collection within the subcutaneous tissues overlying the lateral aspect of the distal fibular metaphysis, associated with extensive subcutaneous edema extending along the lateral part of the distal leg and ankle in conjunction with mild cortical thickening and/or chronic periostitis of the distal tibia and fibula | Diffuse neutrophilic infiltrates throughout the dermis | – Steroid treatment was administered and proper wound care was performed  
– The wound showed good signs of healing in subsequent encounters |
### Table 1 continued. Literature review of magnetic resonance imaging and histology of pyoderma gangrenosum.

| Author  | Age/sex | Initial diagnosis    | MRI finding                                                                 | Histology                                                                 | Treatment and outcome                                                                 |
|---------|---------|----------------------|----------------------------------------------------------------------------|----------------------------------------------------------------------------|---------------------------------------------------------------------------------------|
| Park [this case] | 67/F    | Necrotizing fasciitis | High signal intensity in the left lower leg fascia and subcutaneous fat layer | Diffuse acute inflammation in the dermis with an ulcer and neutrophilic infiltration is apparent in the histopathological examination of the skin lesion | – Intravenous methylprednisolone was administered and regular wound dressing was performed
|          |         |                      |                                                                            |                                                                            | – The skin lesion of the left lower leg exhibited a good response                     |

MRI – magnetic resonance imaging.

at surgery [7] and histology that reveals minimal changes in the epidermis, lymphohistiocytic infiltrate in the dermis, suppuration, necrosis of the superficial fascia, blood vessel thrombosis, and edema in the fascial planes [8].

Few imaging studies have investigated PG (Table 1). The necrotic tissue in a PG lesion can be a diagnostic pitfall, and blood and tissue culture investigations are occasionally positive in PG. Inflammatory markers can be very high in PG, and pyrexia is not a NF-specific feature [9]. PG is generally known to invade to the dermis. NF can be excluded by MRI if deep fascial involvement is not demonstrated. However, our patient had a lesion that invaded the fascia on MRI findings, which was the cause of the initial misdiagnosis of NF. Limitations of this imaging modality primarily include sensitivity greater than specificity and time constraints within which to confirm the diagnosis [10]. Therefore, skin biopsy for histological and microbiological analyses is important to narrow the differential diagnosis [11].

Massive neutrophilic infiltrations in the absence of vasculitis and granuloma formation are typical characteristics of PG [12]. Although PG is a neutrophilic skin lesion, it can occur in neutropenic conditions, such as autoimmune neutropenia and leukocyte adhesion defect type 1 [13]. Therefore, investigations should be performed to rule out other clinical conditions; however, preemptive action should be taken, as patients’ skin lesions deteriorate rapidly. Generally, systemic administration of corticosteroids is the definitive treatment for PG. However, when septic shock ensues as a complication of NF, antimicrobials and vasopressors are usually administered as adjuncts along with the steroids. Notably, early use of steroids is effective in the treatment of NF and PG. The distinct feature of NF is a clinical feature characterized by rapidly worsening vital signs, such as decreasing blood pressure and tachycardia, which is similar to that of septic shock but with dermatological findings. Thus, in actual clinical practice, the utility of MRI findings for diagnosing NF is limited, as the disease is mainly characterized by the aforementioned clinical findings. However, in the presented case, the lesion seemed to have worsened in days, with few vital changes, suggesting that an NF diagnosis could be ruled out clinically.

### Conclusions

Despite the presence of a lesion that invaded the fascia on MRI, our patient was diagnosed with PG and was able to completely recover with steroid treatment. Therefore, PG should be suspected in any patient with sterile, necrotic lesions that are nonresponsive to prolonged antibiotic therapy. To distinguish PG from NF, it is more important to identify the characteristic clinical features than to rely solely on imaging findings or the response to steroid therapy. Taking this into consideration, MRI should not be used as the sole diagnostic tool to distinguish between PG and NF.

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### Declaration of Figures’ Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.
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