

Pyoderma Gangrenosum Complicated with Multiple Cavitory Pulmonary Abscesses: A Case Report and Review of the Literature

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Abstract

Pyoderma Gangrenosum (PG) is an ulcerating, idiopathic, neutrophilic, non-infective inflammatory disease that primarily affects the skin. Pulmonary manifestations are uncommon. The most important differential diagnoses are pneumonia, lung cancer, lung abscess, and Wegener's granulomatosis. Here, we present a case with fever, cough, and cutaneous lesion, which was initially misdiagnosed as pneumonia. A CT scan showed irregular cavitated consolidation in both lower lungs. Histopathological skin findings were non-specific. The skin and lung lesions rapidly improved with corticosteroid treatment. This case demonstrates the challenges in the diagnosis of PG with lung involvement.

Keywords: Lung; Pyoderma gangrenosum; Cavitation

Introduction

Pyoderma Gangrenosum (PG) is an ulcerative, neutrophilic, non-infective dermatological disease of low frequency (10 per 1,000,000 individuals) and unknown etiology. Although it can be associated with systemic disease, such as inflammatory bowel disease, rheumatoid arthritis, and hematological disorder [1,2], pulmonary involvement is rare. Pulmonary manifestations can be asymptomatic or present with continuous fever, dyspnea, cough, and hemoptysis. Thoracic radiography and Computed Tomography (CT) can show cavitation and other etiologies such as lung infection. To avoid misdiagnosis of pulmonary manifestations of PG, as well as other pulmonary diseases, in-depth study is necessary.

Case Presentation

A 22-year-old woman was admitted to our hospital following four days of fever and cough as well as the development of cutaneous lesions. The peak temperature was 39.8°C and the cough was mostly dry, with no sputum. The lesions began as small papules less than 1 cm on the right hand and back (Figure 1). She had no chest pain, dyspnea, or hemoptysis. She did not have arthralgia, abdominal pain and diarrhea. She denied recent travel, weight loss, and alcohol and tobacco use. Her medical history was unremarkable. Her temperature was 39.6°C, and her blood pressure, heart rate, respiratory rate, and oxygen saturation were normal. Abdominal examinations were also normal. Blood tests showed a white blood cell count of $19.22 \times 10^9/L$ (normal range $3.5-9.5 \times 10^9/L$), procalcitonin level of 11.31 ng/L (normal range <0.05 ng/L), and C-reactive protein level of 117.52 mg/L (normal range, <10 mg/L). Antinuclear and antineutrophil envelope antibody spectra were negative. Bacterial, mycobacterial, and fungal blood cultures and sputum were sterile.

A CT scan of the chest revealed irregular cavitated consolidation in both lower lungs (Figure 2). Despite consultations among expert respiratory physicians, the diagnosis was uncertain. Infectious disease was suspected. The patient was started on empirical broad-spectrum antibiotics following diagnosis of pneumonia. However, no improvement was noted in either fever or cough, and her condition deteriorated. The cutaneous lesions spread and progressed rapidly, developing into large blisters on the legs, right hand, and back



Figure 1: Lesions began as small papules less than 1 cm on back.

(Figure 3). They rapidly increased in size, began to ulcerate, and were extremely painful. Cutaneous lesion cultures were sterile. The patient underwent skin biopsy. Skin biopsy from the edge of an ulcer showed focal ulceration of the epidermis associated with fibrin purulent surface exudate. Superficial dermis was markedly edematous. At deeper levels, moderate numbers of neutrophils were found in the dermis. There was no tissue necrosis or vasculitis. Stains for bacterial and fungal organisms were negative. By a process of exclusion, the final diagnosis was PG at multiple sites, with dominant involvement of lungs and skin. The patient was treated with intravenous hydro prednisone 20 mg for 5 d, followed by oral prednisone 40 mg once a day. The dose of prednisone was then slowly tapered off. After four weeks, follow-up CT scan showed regression of the lungs (Figure 4). The skin lesions healed but left scars (Figure 5).

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Figure 2: CT of the chest showing irregular cavitated consolidation in both lower lungs.



Figure 3: Cutaneous lesions spread and progressed rapidly, developing into large blisters.



Figure 4: CT scan showing lung regression.



Figure 5: Healed skin lesions, with scarring.

Discussion

Our case presents a patient who complained of fever and cough. Blood tests showed higher than normal white blood cell count, procalcitonin level, and C-reactive protein level. Further CT scan showed irregular cavitated consolidation in both lower lungs. Histopathological skin findings were non-specific. The main manifestation was predominant neutrophil infiltration. By a process of exclusion, a diagnosis of PG was made. This disease is an ulcerating, idiopathic, neutrophilic, non-infective inflammatory skin disorder of low frequency. A previous epidemiological study estimated an incidence of PG in a standardized European population of six per million per year [3]; however, the incidence in China has not been accurately determined. In addition, although its etiology remains unknown, PG is associated with underlying systemic diseases in half of the cases, including inflammatory bowel disease, rheumatoid arthritis, hematologic disease, and malignancy [1,2]. Our patient was examined without identification of any underlying disease.

Classic PG mainly affects the skin, with pulmonary involvement rare. Skin lesions often begin as pustules, vesicles, or nodules that progress into painful ulcers or erosions with raised borders. The pulmonary symptoms can present with fever, cough, chest pain, and hemoptysis. Lung involvement can occur simultaneously or from a few weeks up to several years after the diagnosis of cutaneous PG. Our patient had pulmonary involvement nearly simultaneously with the appearance of skin lesions. Diagnosis of PG featuring the lung is challenging. Bacterial, mycobacterial, and fungal cultures of blood and sputum are negative [4-6]. Thoracic radiography and CT can show infiltrates, which are often cavitating. Other manifestations also include pleural effusion. The most important differential diagnoses are pneumonia, lung cancer, lung abscess, and Wegener's granulomatosis. The lung cavitated consolidations in our patient were extracutaneous manifestations of PG. Because it is a rapid response to steroid treatment.

Because of the rarity of PG, the choice of therapy is mostly based on case studies. Corticosteroids are usually the first line of therapy [7-9]. High dose prednisolone is currently the preferred treatment. In addition to corticosteroids, other immune-modulating agents are often used in combination with steroids, such as cyclosporine, dapsone, and infliximab [10]. Our patient was treated with corticosteroids and produced a good response, with the follow-up CT scan four weeks later showing lung regression. The skin lesions healed but left scars (Table 1).

Clinical features	The skin: presents most commonly as an extremely painful erythematous lesion which rapidly progresses to a blistered or necrotic ulcer. The lung: continuous fever, dyspnea, cough, chest pain and hemoptysis.
Radiological Characteristics	It includes multiple pulmonary nodules, lung abscesses, pleural effusion, cavitory consolidations, unilateral lung shadows and interstitial pneumonitis.
Pathological Characteristics	Little is known about the pathogenesis. Neutrophil dysfunction including altered chemotaxis is thought to be a main feature
Differential Diagnosis	Other causes of cutaneous ulcers should be considered. These include arterial and venous disease, haematological causes (sickle cell disease, cryoglobulinaemia, anti-phospholipid syndrome), vascular occlusion, vasculitis, infections, calciphylaxis, drug-induced ulceration, primary or metastatic tumours, hypertension (Martorell ulcer) Other pulmonary diseases: Lung infection, lung cancer or Wegener's granulomatosis
Underlying Systemic Diseases	Inflammatory bowel disease, hematologic disorders, arthritis and inherited autoinflammatory syndromes
Treatment	It is immune modulation generally by the use of prednisolone, azathioprine, or ciclosporine

Table 1: A systemic review of pulmonary manifestations of pyoderma gangrenosum with respect to manifestations, differential diagnosis and treatment.

Conclusion

PG is an ulcerating, idiopathic, neutrophilic, non-infective inflammatory disease that primarily affects the skin. Pulmonary involvement is rare. Moreover, early diagnosis of PG with lung involvement is challenging. A diagnosis of PG may be considered when the cause of seeming inexplicable, simultaneous clinical manifestations.

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