A case of vegetative pyoderma gangrenosum

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ABSTRACT

A 55-year-old Chinese man presented with a 17-month history of pustules, papules, nodules, plaques, and ulcers involving most of his body, including the scalp, face, neck, trunk, and extremities. Several superficial ulcers with vegetative exophytic margins and plaques were also present. Cribriform sinuses were on the plaque surfaces and ulcer bases. C-reactive protein level was 120.07 mg/dL, with the erythrocyte sedimentation rate at 95 mm/h. Repeated smears and cultures were negative. Forearm incisional biopsy showed pseudoepitheliomatous hyperplasia and diffuse neutrophil infiltration with microabscesses. Vegetative pyoderma gangrenosum was diagnosed. Lesions markedly abated after systemic corticosteroid therapy.

Introduction

Vegetative pyoderma gangrenosum (PG), also termed superficial granulomatous PG, is a poorly understood entity with features similar to, but a clinical presentation different from, that of classic PG.1,2 In 1988, Wilson-Jones and Winkelmann3 described 25 patients with superficial granulomatous pyoderma that frequently healed without systemic corticosteroid therapy. These lesions characteristically evolved in an indolent fashion into a granulomatous or pyodermatous vegetative lesion and exhibited associated sinuses and cribiform scarring, most commonly on the trunk. We present a case of vegetative PG involving almost the whole body that was successfully treated with methylprednisolone.

Case Report

A 55-year-old Chinese man presented with a 17-month history of pustules, papules, nodules, plaques, and ulcers involving almost the entire body. He was admitted to our hospital in June 2012. The primary lesions were erythematous papules and pustules that first appeared on his back and then gradually involved other parts of the body. Some lesions gradually increased in size to form nodules, vegetative plaques, and superficial ulcers. The lesions were not painful. A putative diagnosis of deep mycosis was made prior to hospital admission. He was treated with the systemic antifungal agents terbinafine and itraconazole, but the lesions did not respond. A pustule also developed at the injection site. The patient had no history of trauma, insect bites, or inflammatory bowel disease. Initial examination revealed multiple pustules (Figure 1A), papules, nodules, plaques, and ulcers (Figure 1B–D) involving the scalp, face, neck, trunk, and extremities, some of which were surrounded by a halo of erythema. Several superficial ulcers and fluctuant, erythematous plaques that measured 2–12 cm in diameter were seen on the chest, right forearm, and left lower extremity (Figure 1B–D). The ulcers had vegetative exophytic margins. Some ulcers demonstrated clean bases but no undermining. Cribriform sinuses were seen on the surfaces of plaques and the bases of ulcers, from which bloody pus discharged under pressure. The systemic examinations were normal.

Laboratory evaluations revealed a high C-reactive protein level (120.07 mg/dL) and erythrocyte sedimentation rate (95 mm/h). The patient’s antibody screen, including evaluation of antineutrophilic...
The results of the following laboratory tests were within normal limits: rheumatoid factor, antinuclear antibody, C3, C4, serum immunoglobulins, serum protein electrophoresis, stool examination for parasites and occult blood, abdominal ultrasonography, human immunodeficiency, hepatitis B virus, hepatitis C virus, and rapid plasma reagin. Chest radiography and a tuberculin test failed to show any signs of tuberculosis. Staphylococcus aureus and Bacillus proteus were cultured from the lesions only once. Subsequent smears and cultures for bacteria and fungi in samples obtained from several ulcers and plaques were negative. A biopsy specimen was obtained from the right forearm. Routine hematoxylin–eosin staining of the tissue specimen showed parakeratosis, acanthosis, pseudoepitheliomatous hyperplasia, and diffuse neutrophilic infiltration with microabcess formation in the dermis. Palisading histiocytes and multinuclear giant cells were seen around microabcesses (Figure 2). Periodic acid Schiff and acid-fast bacilli staining produced negative results.

Neoplasia and infectious diseases were ruled out, and the patient’s condition was diagnosed as vegetative PG. This diagnosis was based on the clinical appearance of the vegetative nodular and plaque lesions with superficial ulcers and by the neutrophilic abscess formation without vasculitis revealed during the pathology examination. He was treated with methylprednisolone at an initial dosage of 60 mg/d for 15 days, with no improvement. The methylprednisolone dosage was then increased to 80 mg/d for 10 days, and the lesions markedly improved. The dosage was then tapered to 60 mg/d over 14 days, and the lesions markedly diminished and gradually healed. After several reductions of the methylprednisolone dose over another 2 months, a high degree of cribiform scarring developed at the sites of the previous lesions (Figure 1E). The steroid was continued but in tapering dosages. Eight months ago, the prednisone dose was tapered from 25 mg/d to 20 mg/d. However, after about 2 weeks at this final dosage, the patient presented with similar lesions on the trunk, so the prednisone dosage was increased to 40 mg/d for 1 month. The lesions regressed. The prednisone was continued but again at tapering doses until it reached 25 mg/d 1 month ago. To date, the patient has been followed-up for 2 years with no other underlying disease found.

Discussion

Vegetative PG, also termed superficial granulomatous pyoderma, is a recently recognized variant of PG with unique clinical, histological, and therapeutic characteristics. Characteristically, these lesions begin as a single furunculoid purple abscess, nodule, or plaque. They are most commonly present on the trunk. Over time, they evolve in an indolent fashion into a granulomatous or pyoderma-tous vegetative lesion with associated sinuses and cribiform scarring. Superficial, focal abscesses surrounded by peripheral palisading histiocytes and foreign body giant cells may be seen on histological examination. Cultures for deep fungal infection, mycobacterium, and atypical mycobacterium are negative. Vegetative PG is responsive to simple therapeutic modalities. It is not associated with any underlying systemic diseases.

Our patient exhibited some unusual characteristics compared with previously described patients with vegetative PG. First, the lesions comprised multiple slowly enlarging pustules, papules, nodules, plaques, and ulcers. In particular, several nodular lesions and plaques 2–12 cm in diameter were seen on the trunk and left lower extremity. The plaques’ appearance mimicked that of neoplastic lesions such as malignant lymphoma, but there was no histopathological evidence of neoplasia. The plaques in our case
were similar to those in previous reports, but in our case they were more proliferous. Second, the lesions involved almost the entire body, including the scalp, face, neck, trunk, and extremities. Lesions of vegetative PG most commonly involve the trunk, although other locations such as the face, extremities, and scrotum have been described. To the best of our knowledge, no previous reports have described lesions involving the entire body. Third, the patient’s therapeutic response was unusual. Most cases of vegetative PG are relatively benign and respond to conservative therapy, including topical and intralesional steroids. Although some are refractory, these lesions uncommonly require oral corticosteroids and other immunosuppressive agents. Several reports have described patients with vegetative PG that required more aggressive systemic immunosuppressive agents. Several reports have described patients with vegetative PG that required more aggressive systemic immunosuppressive agents. These cases were responsive to intravenous immunoglobulin, cyclosporine, or infliximab. The lesions in our case markedly abated with intravenous methylprednisolone at a dosage of 80 mg/d. They had not responded to a 15-day course of methylprednisolone at 60 mg/d.

To diagnose vegetative PG with confidence, we excluded infections with similar clinical appearances. We performed excretion and tissue cultures for fungus. The culture results were negative. Chest radiography and a tuberculin test failed to show any signs of tuberculosis. As periodic acid-Schiff and acid-fast bacilli staining also gave negative results, we excluded deep fungal infections and tuberculosis. In patients with impaired immunity, blastomycosis-like pyoderma may result from an abnormal response to bacterial invasion of the epidermis. We evaluated several smears and cultures for bacteria. *S. aureus* and *B. proteus* were cultured only once, so we considered them contaminating bacteria. The patient received ofloxacin and fosfomycin treatment early in the course of his treatment, but his condition only worsened. The antibiotic treatment was discontinued when vegetative PG was diagnosed. Methylprednisolone treatment was commenced, and the lesions markedly diminished. Also, the patient’s systemic examination proved normal, and he had no other diseases. We therefore excluded blastomycosis-like pyoderma.

In contrast to classic PG, vegetative PG is not routinely associated with underlying disorders. Nonetheless, several of the previously described patients had coexisting diseases: chronic lymphocytic leukemia, polymyalgia, immunoglobulin A paraproteinemia, sarcoidosis, possible rheumatoid arthritis, and definite rheumatoid arthritis in association with Sjögren syndrome. Our patient had coexisting anemia, which might have been associated with dietary restrictions and chronic wasting disease. The most recent routine blood test showed a normal red blood cell count and hemoglobin level.

In summary, we have presented a case of vegetative PG, a variant of PG, with unusual clinical characteristics. The PG was successfully treated with systemic corticosteroid therapy.

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**References**