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A perplexing case of superficial granulomatous pyoderma with sporotrichoid-like distribution

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Abstract

Superficial granulomatous pyoderma (SGP) is a rare pyoderma gangrenosum (PG) variant that differs from classic PG in that the ulcers tend to be more superficial, lack a rapidly advancing border, and are not typically associated with an underlying systemic disease. The ulcers are most commonly painless and located on the trunk, with a clean granulating base. They generally do not show undermining but may have a vegetative border. Lesions usually respond well to either topical or intralesional corticosteroids with complete healing. The classic histopathologic finding is a "three-layer granuloma" in the superficial dermis consisting of central neutrophilic inflammation and necrosis, a surrounding layer of histiocytes and multinucleated giant cells, and an outer most layer of plasma cells and eosinophils. Herein, we present a unique case of SGP with sporotrichoid-like distribution on the lower extremity.

Keywords:: superficial granulomatous pyoderma, pyoderma gangrenosum

Introduction

Superficial granulomatous pyoderma (SGP) is a rare variant of pyoderma gangrenosum (PG) that is clinically and histologically different from classic PG. Ulcers in SGP tend to be more superficial, lack a rapidly advancing border, and are not typically associated with an underlying systemic disease [1]. The ulcers are most commonly painless, located on the trunk. They have a clean granulating base without undermining and a vegetative border. Superficial granulomatous pyoderma lesions usually respond well to either topical or intralesional corticosteroids with complete healing [2]. Classically, the histopathology of SGP reveals a "three-layer granuloma" in the superficial dermis consisting of central neutrophilic inflammation and necrosis, a surrounding layer of histiocytes and multinucleated giant cells, and an outer most layer of plasma cells and eosinophils. [1] The variable clinical findings, in addition to its granulomatous histology, makes SGP a challenging diagnosis. Herein, we present a unique case of SGP with sporotrichoid-like distribution on the lower extremity, complicating an already rare diagnosis.

Case Synopsis

A 74-year-old man with diabetes mellitus type II, coronary artery disease, hypertension, and plaque psoriasis presented with a six-week history of an enlarging, tender red plaque with central ulceration on the right lower extremity and multiple red-pink nodules extending up the leg. He had no pertinent travel history but had frequently gardened in the six months prior to presentation. Before arrival to the clinic he had completed a 10-day course of doxycycline without improvement. The patient had no systemic symptoms and otherwise felt well.

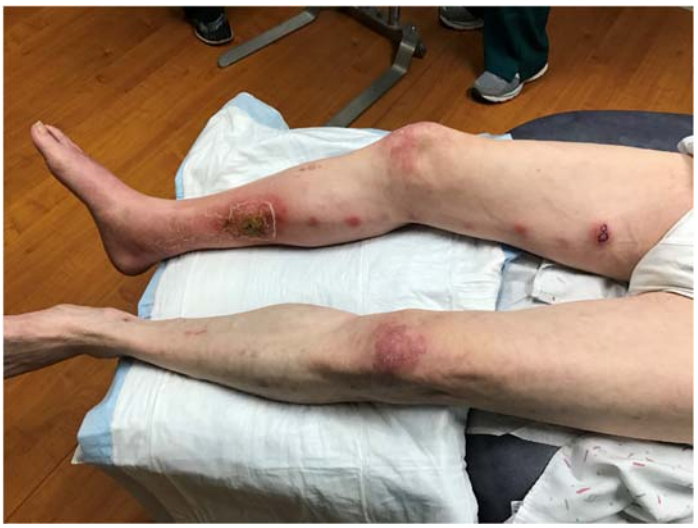

Figure 1. *Superficial granulomatous pyoderma presenting with a right lower extremity ulcerated plaque with multiple, tender nodules extending up the leg in a sporotrichoid-like distribution.*

Physical examination revealed a 10cm red plaque with overlying yellow crust and a 2cm×2cm central shallow ulcer on the right distal lower extremity. There were seven smaller, well-defined, red, firm, mobile nodules extending superiorly along the medial leg and thigh in a sporotrichoid-like distribution (**Figure 1**). Histopathology of the lesions demonstrated an acute and chronic suppurative and granulomatous infiltrate in the deep dermis extending down to subcutis. Over the ensuing months, several sterile biopsies were obtained for bacterial, mycobacterial, and fungal cultures, none of which grew a pathogenic organism. During this time the patient was started on a five-month course of oral itraconazole for presumed sporotrichosis. The specimens were initially sent to the University of Washington for broad-range polymerase chain reaction (PCR) testing, with two attempts that failed to identify a pathogen. The specimens were finally sent to the Centers for Disease Control and Prevention (CDC) for further work-up. Special stains were noncontributory, except for an immunohistochemical stain that was cross-reactive for *Mycobacterium* species, but paneubacteria 16S rRNA gene PCR was positive only for *Streptococcus salivarius* group. These were considered a false positive and contaminant, respectively. On further review of the pathology, a "three-layer" granulomatous inflammatory infiltrate was appreciated, with a central necrotic area with

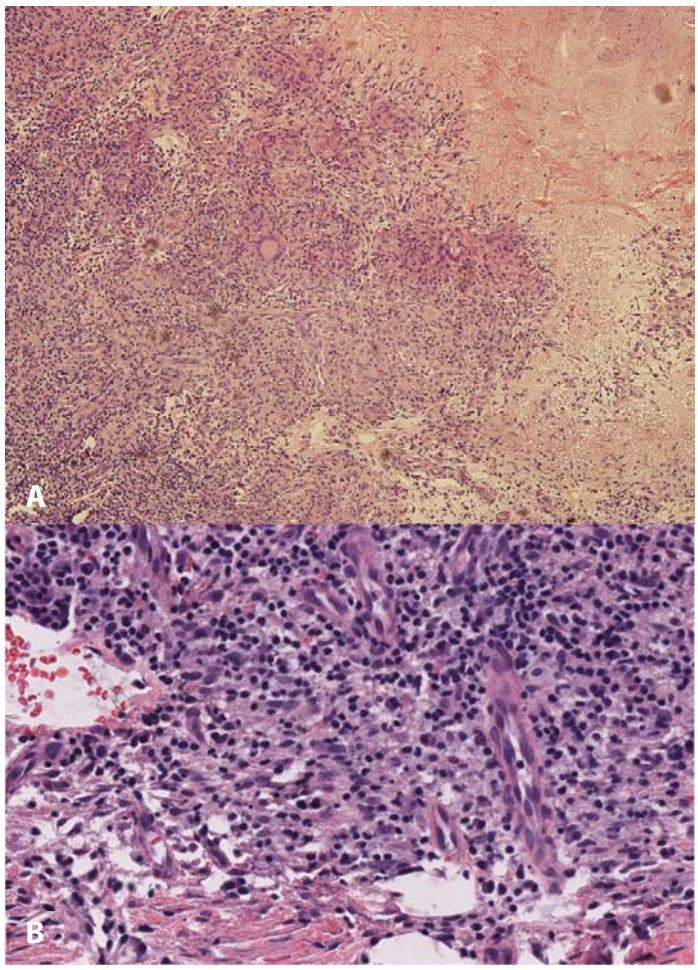

Figure 2. *Photomicrographs of the histopathologic features of superficial granulomatous pyoderma, demonstrating various features of the classic "three-layer" granuloma with H&E staining.* ***A)*** *Inflammation immediately adjacent to the necrotic zone is rich in histiocytes with giant cell formation, and neutrophils are present, 60×.* ***B)*** *At the edge of the infiltrate, plasma cells are seen, 300×.*

neutrophils, a second ring of histiocytes and giant cells, and an outermost ring of lymphocytes and plasma cells (**Figure 2**). The diagnosis of superficial granulomatous pyoderma (SGP) was made. Malignancy work-up including computed tomography scans of the chest, abdomen, and pelvis, urine analysis, serum electrophoresis, complete blood count, and prostate specific antigen within normal limits. Colonoscopy screening was performed in 2014 and demonstrated two polyps and a focal area of nonspecific inflammation in the sigmoid colon. The patient denied any symptoms concerning for colitis. The patient was started on potent topical corticosteroids and intralesional

triamcinolone injections (ILK) of the nodules extending up the lower leg. After 12-months of minimal change with aggressive wound care, five of which included administration of oral itraconazole, the ulceration healed after only four weeks of potent topical corticosteroids (**Figure 3**). The nodules extending up the leg also healed with ILK. The patient has not had any signs of recurrence after 16 months.

Case Discussion

This case of SGP is an important addition to the literature owing to its unique clinical presentation. The "classic" presentation of SGP is a single, indolent ulceration on the trunk arising after trauma to the skin [1]. The lesion is typically more superficial than classic PG, with a clean base, stable vegetative border, and the "three-layer granuloma" histologically [1]. Typically, SPG is not associated with any systemic disease and responds well to conservative treatments, like localized anti-inflammatory treatments and systemic antibiotics [1]. Since SPG is rare, atypical clinical presentations and histology can make this diagnosis more difficult, resulting in diagnostic delay and increased morbidity.

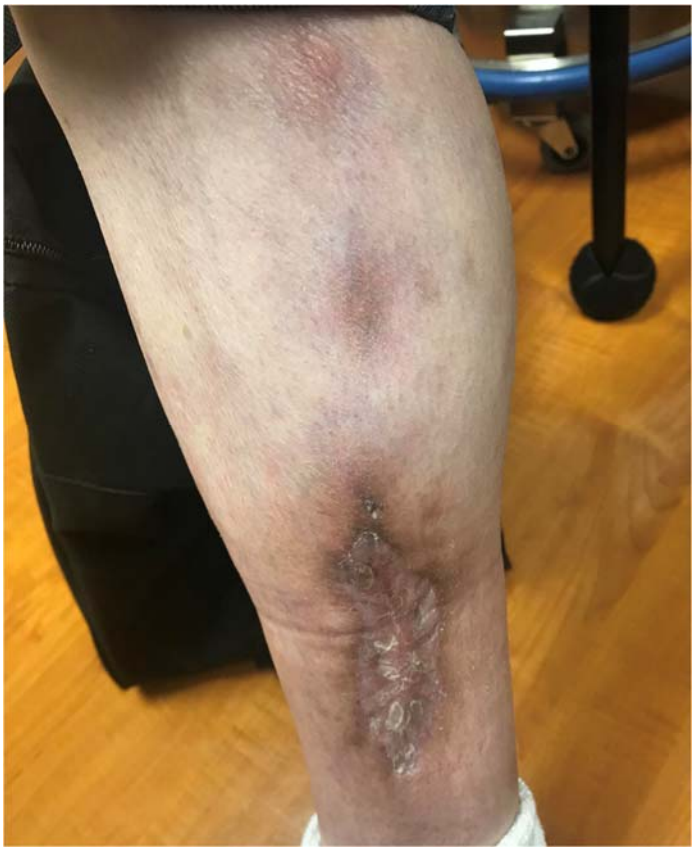

Figure 3. *Superficial granulomatous pyoderma lesion on right lower extremity after administration of topical and intralesional steroids.*

Although the classic location is on the trunk, only approximately 50% of lesions present in this location. Approximately a third present with lesions involving the extremities [3]. To our knowledge, the sporotrichoid-like distribution has not previously been reported in the literature and was another unusual feature that made the diagnosis in this case especially difficult. The most common underlying etiology of a sporotrichoid eruption is a lymphocutaneous infection, referred to as nodular lymphangitis. Numerous infectious agents can cause this condition, but the most common include *Sporothrix schenckii*, *Nocardia brasiliensis*, *Leishmania* species, and atypical mycobacterial species [4]. In this case, all infections had been ruled out as an underlying etiology, which is an important step in making this diagnosis. Superficial granulomatous pyoderma should be treated as a diagnosis of exclusion, eliminating the possibility of infection prior to treatment.

In most cases, the "three-layer granuloma" exhibited on histology does not have all the "classic" features present. Although plasma cells are found in the outermost layer in 78% of cases, eosinophils are only seen 43% of the time in this outermost layer. Also, only half of cases demonstrate pseudoepitheliomatous hyperplasia on pathology [3]. Therefore, the lack of "classic" histologic features of the "three-layer granuloma" should not exclude the diagnosis of SPG.

Conclusion

In summary, this case highlights the importance of keeping SPG in the differential diagnosis when evaluating ulcers with granulomatous inflammation, even in rare cases with sporotrichoid-like distribution. In addition, the absence of any one "classic" feature, clinically or histologically, should not lead to exclusion of this diagnosis. Finally, SPG is

a diagnosis of exclusion, so infectious causes should be ruled out prior to making this diagnosis.

Potential conflicts of interest

The authors declare no conflicts of interests.

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