Pyoderma gangrenosum is an inflammatory disease of unknown etiology that may be seen in association with several systemic diseases (Table 1). The disease process begins with the sudden appearance of painful pustules, papules, and nodules that rapidly spread, producing a large ulcer with a characteristic purplish undermined border (Figures 1 and 2). Pain in the lesion is a dominant symptom. Severe tenderness upon palpation is characteristic. The lesions, usually solitary but may be multiple, occur on all areas of the body. They are most prevalent, however, on the lower extremities.

In approximately 40% to 50% of cases, pyoderma gangrenosum is idiopathic. The most common underlying disease associated with pyoderma gangrenosum is ulcerative colitis, which has been reported in 30% to 60% of pyoderma gangrenosum cases. In most instances, the ulcerative colitis precedes the development of pyoderma gangrenosum, although it may be the initial presentation or, rarely, may signal an exacerbation. In ulcerative colitis cases, pyoderma gangrenosum generally has occurred in the absence of any evidence of clinical activity of the ulcerative colitis bowel disease. It should be noted that although ulcerative colitis is most frequently associated with pyoderma gangrenosum, only 1% to 5% of ulcerative colitis patients develop pyoderma gangrenosum.

**ETIOLOGY**

No etiology has been established, although a large body of evidence suggesting a possible vasculopathy has been accumulated. The fact that pyoderma gangrenosum has been seen in association with leukocytoclastic angitis and that it occurs with high frequency in Takayasu’s disease, suggests that vasculitis, possibly mediated by circulating immune complexes, may be present.

**CLINICAL FEATURES**

The principal clinical features of pyoderma gangrenosum are presented in Table 2. The disease is explosive in onset, usu-

### Table 1. Pyoderma Gangrenosum: Common Disease Associations

- Ulcerative colitis
- Crohn’s disease
- Seropositive and seronegative arthritis
- Myeloproliferative diseases
- Paraproteinemia
  - IgA myeloma

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Off-Label Product Discussion: The author has disclosed that this article references the following off-label/unapproved use of drugs or products: cyclosporine, thalidomide, and infliximab for pyoderma gangrenosum.
ally beginning as a painful pustular nodular lesion that rapidly expands and may enlarge to 5 to 10 cm within a 10-day period. The purplish undermined border is characteristic; sinus tract formation is also present. The base of the lesion is boggy and necrotic and may extend to the fat and fascial layers.

There is a second uncommon variant of pyoderma gangrenosum characterized by indolent slow growth of massive granulation tissue. Spontaneous regression and healing in one area and progression in another occurs. Both forms of pyoderma gangrenosum heal with thin atrophic cribriform scar formation.

Pyoderma gangrenosum is a diagnosis of exclusion. A recent review showed that misdiagnosis can occur frequently. Conditions that mimic pyoderma gangrenosum include vasculitis, arterial and venous insufficiencies, cutaneous blood vessel thrombosis secondary to the antiphospholipid syndrome, cutaneous malignancies, and polymicrobial and deep cutaneous fungal infections.

PATHERGY

Pathergy, an inflammatory reaction in the skin induced by trauma, is seen in approximately 40% of pyoderma gangrenosum patients. Clinically, this can be demonstrated by inserting a needle into the skin at a 30-degree angle. Over the ensuing 24 to 48 hours, a sterile pustule will develop at this site. Examples of pathergy are commonly detected at venesection sites on patients’ arms. These sites of pathergy may on occasion produce large tender nodules. Incision and drainage of these lesions promotes extension. These lesions are sterile.

TREATMENT

Until recently, the treatment of pyoderma gangrenosum has been problematic. Isolated chronic lesions can be successfully treated with the intralesional injection of triamcinolone (5-10 mg/cc). The slow infusion of 1 gm of methylprednisolone over a 4-hour period for 5 consecutive days has been shown to produce an immediate improvement in pain and to stop the spreading of the lesion. However, continued use of steroids to control the pyoderma gangrenosum is associated with many side effects.

Recently, pyoderma gangrenosum has been successfully treated with thalidomide. In the author’s experience, the lesions rapidly respond to this therapy. However, peripheral neuropathy (tingling sensations of the fingers and feet) may occur in as many as 30% of patients, even when low doses of 50 to 100 mg of thalidomide per day are administered.

Cyclosporine in a dosage of 4 mg per kilogram has also been associated with a very rapid response of the pyoderma gangrenosum lesions. It is important, however, to monitor the patient for signs of hirsutism, azthymia, and hypotension, which are associated with this drug.

Most recently, evidence has appeared in the literature indicating that infliximab (Remicade®) may be effective in the treatment of pyoderma gangrenosum associated with Crohn’s disease.

Table 2. Common Clinical Features of Pyoderma Gangrenosum

- Primary lesion, most commonly a pustule with an undermined purplish border, may be single or multiple
- Explosive onset generally heralded by pain; ulcers may reach 5 to 10 cm in 10 day period
- Pathergy; 40% of pyoderma gangrenosum patients develop lesions at sites of previous trauma (ie, venesection or surgical operative sites)
- Slow to heal; thin cribriform scar formation
References


