Pyoderma Gangrenosum in a Patient with Systemic Sclerosis

To the Editor:

Pyoderma gangrenosum (PG) is an ulcerative inflammatory noninfectious disease of the skin. Treatment is mainly empirical, consisting of a combination of local and systemic treatments, including corticosteroids and immunosuppressive drugs.1

In many cases, PG is associated with an underlying disease, most commonly inflammatory bowel disease, occasionally in the stromal area. PG occurs in several hematological and malignant diseases. PG has also been described in several rheumatic diseases: rheumatoid arthritis, spondyloarthropathies, systemic lupus erythematosus, Behçet’s disease, and sarcoidosis.2 Hod, et al3 reported a huge PG-like lesion as a presenting sign of antiphospholipid antibody syndrome (APS). In the literature we are aware of only 2 reports of PG lesions in patients with systemic sclerosis (SSc), one a patient who developed a lesion in a colonic stoma due to colon cancer4. It is not clear whether she developed the skin lesion because of the stoma or because of the SSc. A second report is of a patient with SSc who developed a scrotal pyoderma gangrenosum lesion5. The antiphospholipid antibody status of these patients is unknown.

We describe a patient with SSc who had a positive APS serology and who developed a PG skin lesion.

A 54-year-old woman was diagnosed with limited SSc 10 years before. Her main disease manifestations include severe Raynaud’s phenomenon, digital ulcers, dyspepsia, and sclerodactyly. Serological tests revealed a positive antinuclear antibody with an anticentromere pattern. There was a consistent finding of elevated antiphospholipid IgG (18 U/ml; normal values 0–9.9 U/ml) and IgM (87 U/ml; normal values 0–6.9 U/ml). Laboratory tests revealed a chronically mildly asymptomatic elevation of the creatine phosphokinase level. Treatment included diltiazem, enalapril, nitroderm patches, omeprazole, and aspirin.

In June 2009 she presented with a single 18.5 cm × 8.5 cm inflammatory skin ulcer on her left shin. The ulcer had a violaceous colored border and a necrotic central base. A skin biopsy demonstrated dermal edema and a dense diffuse infiltrate of neutrophils throughout the dermis and in the upper subcutaneous fat. These findings were compatible with the diagnosis of PG. She initially was treated topically with glucocorticoids and garamycin creams. Subsequently, she underwent surgical debridement with skin grafting. High-dose prednisone (1 mg/kg) was initiated, despite the fear of inducing an SSc renal crisis. Cyclosporine (2 mg/kg) was introduced as a corticosteroid-sparing agent. This regimen resulted in gradual improvement. About 5 months later, the ulcer healed and a scar remained. The prednisone dose was tapered and eventually stopped. Currently, she is being treated with cyclosporine monotherapy. There has been no recurrence of the lesion for about 2 years.

PG is an inflammatory lesion, rarely described in rheumatic diseases. Our patient dramatically improved while treated topically and systematically with high-dose prednisone and cyclosporine. Anticoagulation was not included in her treatment regimen. This treatment compares with that of the patient described by Hod, et al2, whose symptom of APS was PG and was thus treated with anticoagulation combined with immunosuppressive therapy. That our patient developed a PG skin lesion despite aspirin and that the lesion healed with antiinflammatory treatment without anticoagulation leads us to propose that the lesion was mainly related to SSc rather than APS status. Physicians should be aware of the possible association between SSc and PG and treat accordingly.

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J Rheumatol 2012;39:1; doi:10.3899/jrheum.110479