

Cutaneous Polyarteritis Nodosa after Streptococcal Necrotizing Fasciitis

RONNIT HAMUY STEIN, M.D.¹, ROBERT G. PHELPS, M.D.^{1,2}, AND ALLEN N. SAPADIN, M.D.¹

Abstract

Polyarteritis nodosa (PAN) is a necrotizing arteritis of small and medium-sized vessels. It may present with hypertension and/or renal insufficiency. Peripheral neuropathy, myopathy, joint pains, testicular pain, and ischemic myalgias may also be seen. Gastrointestinal involvement may lead to gangrene of the bowel, peritonitis, perforation, intra-abdominal hemorrhage, and pancreatitis. The cutaneous manifestations include tender subcutaneous nodules grouped along the course of superficial arteries of the lower extremities, with or without an overlying livedo reticularis. Although multisystem involvement is characteristic, sometimes only one organ or system may be involved. Associations with viral hepatitis (both B and C) and streptococcal infection have been established for PAN. Recurrent strep infections of the upper respiratory tract, streptococcal glomerulonephritis and rheumatic fever have previously been linked to PAN. This report extends the spectrum of associated streptococcal infections to include necrotizing fasciitis.

Key Words: Polyarteritis nodosa, necrotizing fasciitis.

NECROTIZING FASCIITIS (NF) is a potentially lethal infection of the deep subcutaneous tissues that progresses to destroy fat and fascia (1). The majority of cases are polymicrobial in nature; coinfection with synergistic, facultative, aerobic and anaerobic gas-forming organisms is most common (2). When NF is caused by a single bacterial agent, group A beta hemolytic streptococcus (GAS) is the most common etiology (1). A current study reports 66% of patients with NF yielding GAS in microbiology swabs (3).

A 1997 study demonstrated that GAS-NF has been increasing in recent years (4). We describe a 35-year-old woman who developed cutaneous polyarteritis nodosa (PAN) six weeks after an episode of NF involving the same extremity. While previous studies have reported a predisposition to cutaneous PAN after streptococcal infection (5), its occurrence after GAS-NF has not been previously described.

Case Report

A 35-year-old woman was diagnosed with NF of the left lower extremity 5 years after cardiac

transplantation for viral cardiomyopathy. Medications at the time of the surgery included cyclosporine, prednisone and furosemide. Multiple cultures from the left thigh identified the etiologic agent as GAS. Treatment included aggressive antibiotic therapy and emergent fasciotomy. Six weeks later, the patient presented with a painful left calf associated with myalgias and arthralgias. Examination revealed tender subcutaneous nodules with overlying erythema on the distal portion of her left calf. The differential diagnosis included erythema nodosum and cutaneous PAN. Biopsy specimen revealed vasculitis of medium-sized arteries, consistent with cutaneous PAN. The erythema and nodules cleared with intralesional triamcinolone injections.

Six months later the patient developed cellulitis of the left thigh; multiple cultures identified the agent as GAS. The cellulitis cleared with a 5-day course of intravenous clindamycin and penicillin. Eight weeks later, the patient experienced excruciating pain in her left calf while standing. The pain had increased in intensity during the past month and was extending anteriorly. Examination revealed a recurrence of tender subcutaneous nodules on the left calf with an overlying livedoid erythema (Fig. 1). The area affected measured 8 x 12 cm. Medications included daily, low-dose prednisone (5 mg). Routine laboratory studies, including complete blood count and chemistry screen, were noncontributory. Rheumatoid fac-

From the Departments of ¹Dermatology and ²Pathology, Mount Sinai School of Medicine, New York, NY.

Address correspondence to Allen N. Sapadin, M.D., The Mount Sinai Medical Center, Box 1047, One East 100th Street, New York, NY 10029-6574.



Fig. 1. Cutaneous polyarteritis nodosa. Subcutaneous nodules on the left calf with an overlying livedoid erythema.

tor, hepatitis B and C, cryoglobulins, circulating immune complexes, and ANA were negative. Repeat skin biopsy specimen demonstrated fibrinoid necrosis of medium-sized dermal vessels (Fig. 2). The patient was treated with an increased dose of prednisone (60 mg/day) and improved dramatically within 48 hours. The prednisone was tapered within a two-week period and the patient has remained symptom-free since that time.

Discussion

Many cases of NF result from an injury that disrupts the integrity of the skin: minor cuts, burns, injection-drug use, surgical procedures, insect bites and varicella infection. Blunt trauma may also increase the risk for this infection. Diabetes, arterial insufficiency, and immunocompromised states may be predisposing factors (1). Both anaerobic and aerobic organisms have been implicated. Aerobic agents include group A *Streptococcus*, *Staphylococcus aureus*, *Escherichia coli*, and other enterobacteriaceae (6).

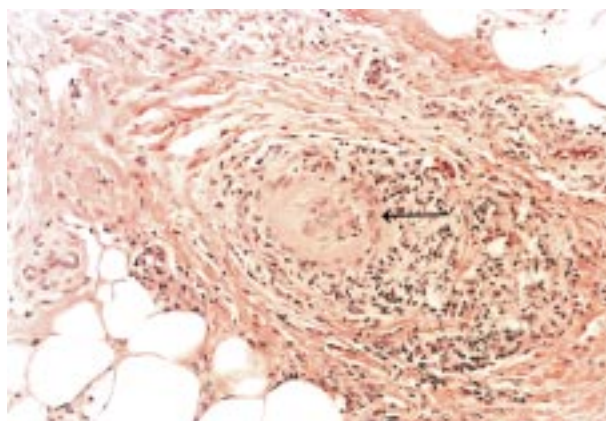


Fig. 2. Fibrinoid necrosis of medium-sized dermal vessel (arrow). (Hematoxylin-eosin stain; original magnification x 100.)

Cutaneous PAN is characterized by vascular inflammation and necrosis of small and medium-sized arteries; it has a tendency to be chronic or relapsing. Whereas the cutaneous form is limited to the skin, the systemic form of PAN additionally targets the kidneys, joints, and gastrointestinal tract (7). Renal disease and complications from hypertension account for the majority of deaths. Livedo reticularis and underlying tender, erythematous, subcutaneous nodules represent the classic presentation of cutaneous PAN (5, 7). Other cutaneous features associated with PAN include ulcers, ecchymoses, gangrene and lesions of the nailfolds and distal aspects of the digits. The lower extremities are preferentially affected (4). Arthralgias and myalgias may also be present (7).

Erythema nodosum is the most common entity with which PAN is confused. The differential diagnosis also includes other vasculitides such as Wegener's granulomatosis, hypersensitivity vasculitis, giant cell arteritis, and urticarial vasculitis (8). Diagnosis involves a high index of clinical suspicion and subsequent biopsy of the affected tissue. Laboratory testing is nonspecific and includes anemia, leukocytosis, altered renal function, urinary sediment abnormalities, and elevation in erythrocyte sedimentation rate. Rheumatoid factor, circulating immune complexes, and cryoglobulins may be present. ANCA tests are generally negative (8).

Although the pathogenesis of PAN is unknown, associations with viral hepatitis and streptococcal infection have been established (5). In 79 adult cases of cutaneous PAN, 7 patients had a history of recurrent streptococcal infection of the upper respiratory tract (5). Sheth et al. (7) describe four cases in children that were associated with previous streptococcal infection. Associations with rheumatic fever and streptococcal glomerulonephritis have also been described (7). This report extends the spectrum of streptococcal infections associated with cutaneous PAN to include NF.

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