

SAPHO Syndrome

Diana A. Rivers DO

Lehigh Valley Health Network, Diana.Rivers@lvhn.org

Megan A. Jones DO

Lehigh Valley Health Network, Megan.Jones@lvhn.org

Joshua M. Levin MD

Lehigh Valley Health Network, Joshua_M.Levin@lvhn.org

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SAPHO Syndrome

Diana A. Rivers, DO, Megan A. Jones, DO and Joshua Levin, MD
Lehigh Valley Health Network, Allentown, Pa.

HISTORY OF PRESENT ILLNESS: 53 year-old Caucasian male presents with non-healing, painful ulcers with purulent drainage for several months. Additionally, he had a history of chronic discharge from the right medial thigh following his right hip arthroplasty which improved with clobetasol. He followed with orthopedics post-operatively and received multiple courses of antibiotics, despite several negative tissue cultures. The right lower leg ulcer improved with intralesional triamcinolone, topical corticosteroids and collagenase. However, new ulcers continued to appear involving the left cheek and left dorsal hand. These new lesions responded well to intralesional corticosteroids and topical clobetasol.

MEDICAL HISTORY/SURGICAL HISTORY: Acne, pyoderma gangrenosum, rheumatoid arthritis, osteoarthritis, atrial fibrillation, hypercholesterolemia, adrenal insufficiency, diverticulitis, right hip arthroplasty

MEDICATIONS: Prednisone, tretinoin, aspirin, ibuprofen, acetaminophen, simvastatin, tramadol, clobetasol propionate, ergocalciferol, folic acid, digoxin

PREVIOUS TREATMENTS: Clobetasol propionate, silver sulfadiazine, collagenase

PHYSICAL EXAMINATION: Right lower leg, 4x3 cm ulcer with cribriform surface at the periphery, fibrin coat, and mild surrounding erythema. Hands with swelling and joint deformity. Left neck with a 1 cm erythematous nodule with purulent discharge. Right medial leg with a large, 8x7 cm pink, boggy area of skin with three small draining fistulas. Single, 2x2 cm ulcer with yellow, fibrinous material.

LABORATORY DATA: Multiple aerobic bacterial wound cultures: right leg (4/24/18), left neck (4/24/18), right leg (5/29/18), left dorsal hand (8/2/18), right hip (9/11/18) – all negative. Aerobic bacterial wound culture (4/24/18) right leg: *Stenotrophomonas maltophilia*.

Fungal culture (2/27/18) right lower leg – negative.

Tissue cultures: aerobic and anaerobic bacterial, fungal, AFB (2/7/18) right trochanteric bursa- negative. Aerobic bacterial and mycobacterial (2/27/18) Left lower leg – negative.

BIOPSY: *Health Network Laboratories* (S18-8148, 2/27/18) Right lower leg: "ulceration with dense, neutrophilic inflammation. A few blood vessels show patchy fibrinoid change, slight fibroplasia and mural thickening and focal neutrophilic inflammation. PAS, GMS, Fite stains negative."



Figure 1: Right medial thigh with chronic draining fistulas following right hip arthroplasty.



Figure 2: Right lower leg ulcer with a cribriform surface at the periphery, fibrin coat, violaceous border and erythematous halo. Consistent with pyoderma gangrenosum (PG).

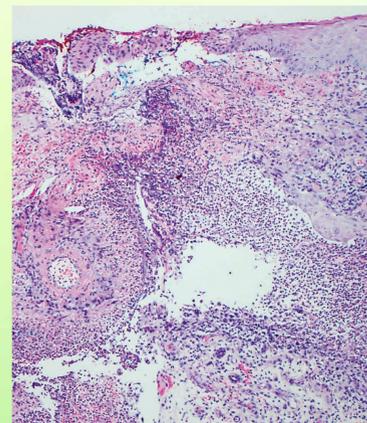


Figure 3: H&E, Right lower leg (10x): Ulceration with dense, neutrophilic inflammation.

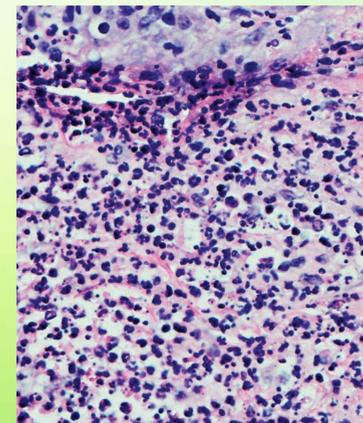


Figure 4: H&E, Right lower leg (60x): Dense, neutrophilic infiltrate.

DISCUSSION: Synovitis, acne, pustulosis, hyperostosis, and osteitis (SAPHO) syndrome is a rare entity that was first defined in 1987 by Chamot, et al. Incidence is less than 1:10,000 and mean age of initial symptom onset is 29 years old. This syndrome is manifested by a combination of inflammatory cutaneous disorders and musculoskeletal findings. The pathogenesis of this inflammatory condition is poorly understood.

Diagnosis of SAPHO syndrome is based on clinical and radiographic findings. Although multiple diagnostic criteria have been proposed, none have been validated. Cutaneous and osteoarticular manifestations have an insidious onset and may occur years apart which leads to delayed diagnosis and management. The most common cutaneous manifestations include palmoplantar pustulosis, acne conglobata, acne fulminans, hidradenitis suppurativa, pustular psoriasis, pyoderma gangrenosum, psoriasis vulgaris, and Sweet syndrome. Osteoarticular manifestations (osteitis, hyperostosis) cause bone pain, swelling, and restricted mobility. The sternum, clavicles, and sternocostoclavicular joints are affected in 90% cases. Spondylodiscitis, osteosclerosis, and paravertebral ossifications of the spine occur in 30% of cases. Additionally, 33% of patients have metadiaphysis of the femur, tibia, and/or fibula. The average number of bone lesions per patient in a review of 120 cases was 1.92. Synovitis affects the sacroiliac, sternoclavicular joints, hips, or knees in up to 20% of patients.

During acute flares, C-reactive protein and erythrocyte sedimentation rate may be elevated. Occasionally, the alkaline phosphatase level may also be increased. Radiographically, osteolysis, sclerosis, and hyperostosis are seen. Technetium-99 scans may detect early or sub-clinical osteoarticular lesions. Histopathologically, bone biopsies show sterile osteomyelitis which is indistinguishable from infective osteomyelitis. Skin biopsies show neutrophilic pseudo-abscesses in the acute stage, mononuclear cells with occasional non-caseating granulomas in the intermediate stage, or mononuclear cells, fibrosis and new bone formation in the chronic stage.

Due to the rarity of this condition, treatment algorithms are not established. Retrospective studies and case series support non-steroidal anti-inflammatory drugs (NSAIDs) as first line. Bisphosphonates, corticosteroids, and methotrexate are considered second line agents. In patients who fail these therapies, biologic agents have been used with success. In a recent review of 66 cases treated with biologics, TNF blockers had the best response rate in osteoarticular and skin disease. Other biologics reported to be efficacious include IL-1 inhibitors, IL-17 inhibitors, and IL-23 inhibitors.

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