One SAPHO-sticated Syndrome: Hidradenitis Suppurativa in the Setting Of SAPHO Syndrome

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Learning Objectives

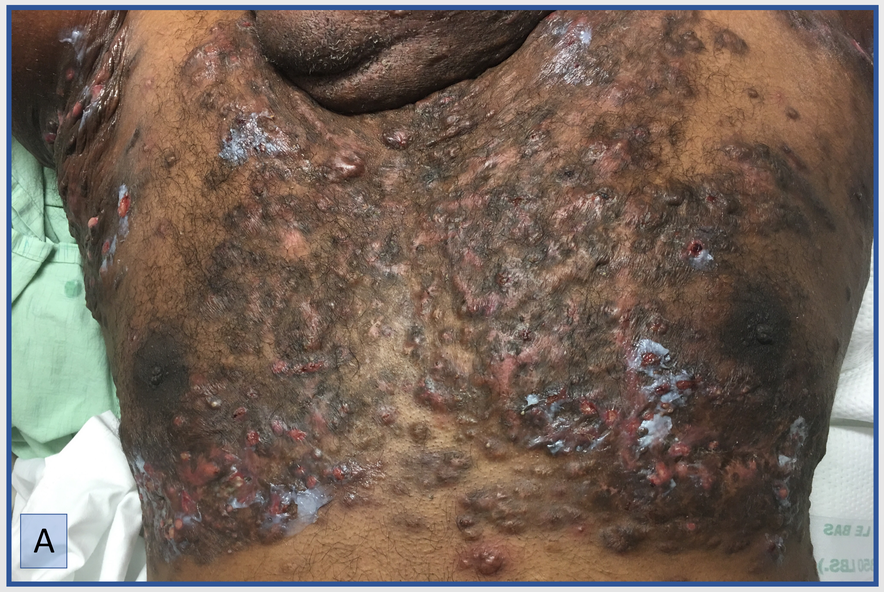
1. Recognize the clinical features of severe hidradenitis suppurativa and SAPHO syndrome
2. Evaluate and manage hidradenitis suppurativa in the setting of SAPHO syndrome

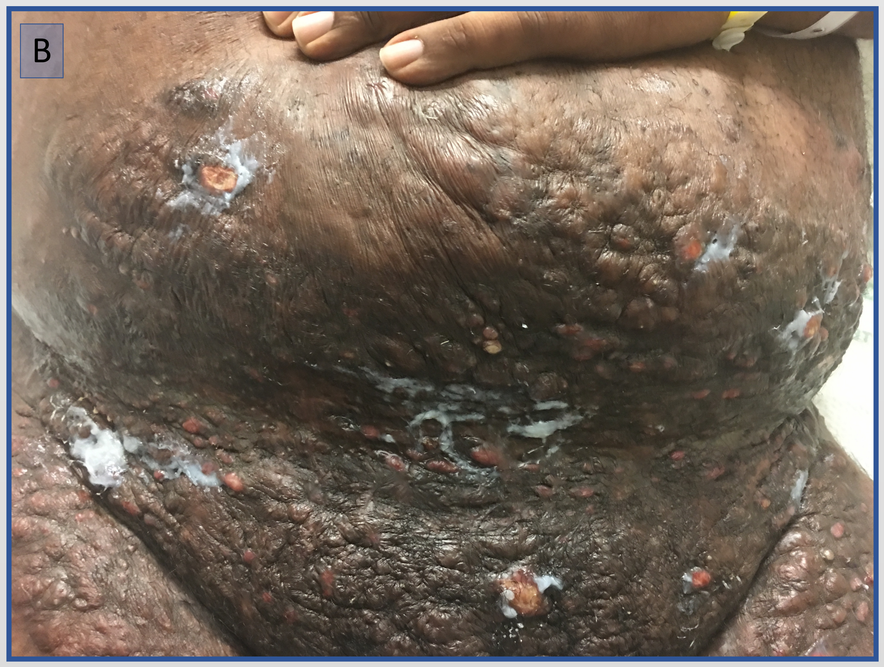
Case Presentation

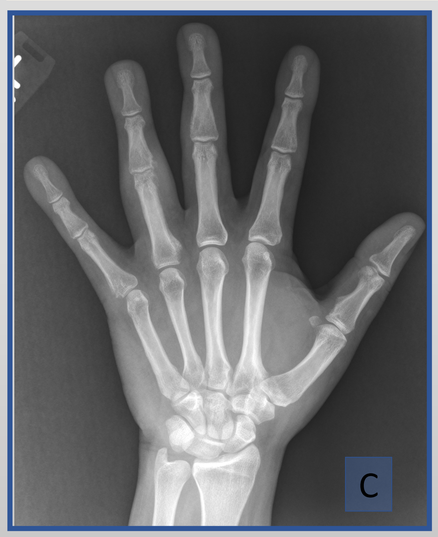
History of Present Illness:
A 36-year-old African American man with a ten year history of hidradenitis suppurativa (HS), hypertension, and morbid obesity presented with a severe HS flare and diffuse joint pain. His current flare began as nodules surrounding hair follicles that evolved into erosions with pus. He endorsed subjective fevers, chills, fatigue, and 10/10 pain in the affected areas. He also has worsening pain and swelling of the third and fourth MCP joints. He has been treated with topical clindamycin, oral doxycycline, and intralesional triamcinolone acetonide. He recently started using adalimumab injections.

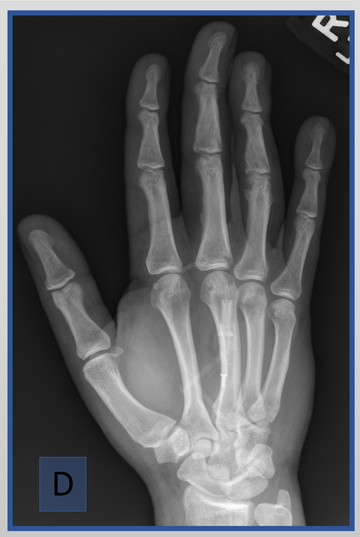
Physical Exam:
Temp: 38.7 ºC
Skin: nodules and sinus tracks draining pus, diffuse thick scarring over chest, axillae, occiput, lower abdomen and pannus, groin and inner thighs.

Studies:
White Blood Cell Count: 20.3 (4.5 – 11.0 K/MM3)
ESR: 86 (<=15mm/HR)
X-rays: Right and left hand showed erosions and periosteal reaction consistent with inflammatory arthropathy.









A. Axilla & Thorax
B. Lower Abdomen & Pannus
C. Left hand x-ray with periosteal reaction at the 4th PIP
D. Right hand x-ray with periosteal reaction at the 3rd & 4th phalynx

Discussion

HS occurs in intertriginous areas and at sites of skin friction. Presents initially as painful, deep-seated, inflammatory nodules. Recurrent disease involves clusters of open comedones, abscesses, sinus tracts, and scarring.

HS, along with acne conglobate and acne fulminans, affects 25% of patients with SAPHO (synovitis, acne, pustulosis, hyperostosis, and osteomyelitis) syndrome.

SAPHO syndrome consists of various aseptic neutrophilic dermatoses associated with aseptic osteoarticular lesions.

The most common **skin lesions** include palmoplantar pustulosis and severe acne. **Osteoarticular** manifestations include osteitis, hyperostosis synovitis, arthropathy and enthesopathy.

HS Management: follows the Hurley Staging system and includes topical or oral clindamycin, oral tetracyclines, adalimumab, infliximab, intralesional corticosteroids, topical resorcinol, and punch debridement of lesions.

SAPHO management: mostly symptom control, medications include NSAIDs, antimicrobial therapy, colchicine, corticosteroids, bisphosphonates, and TNF-alpha inhibitors.

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