

Clinical Images: Marked Inflammation in a Patient with Cervical Vertebral SAPHO Complicated by Vertebral Body Collapse and Severe Kyphosis

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A 61 year old Caucasian man, diagnosed with palmar plantar psoriasis for 20 years, presented with continuous and inflammatory pain involving the chest, lower back, and neck lasting over 1 year, with no other symptoms.

This work was presented at a radiology–rheumatology meeting focusing on the contribution of imaging to the understanding of the pathogenesis and treatment decisions in musculoskeletal rheumatic diseases that took place in December 2016 at the Sheba Medical Center, Tel Hashomer, Israel

Physical examination revealed severe limitation of neck movement, particularly in extension and chest expansion. Laboratory studies disclosed mildly elevated C-reactive protein.

Chest and cervical spine computed tomography scans showed sternomanubrial sclerosis and mild kyphosis (C6-7) with narrowing of disc space. Cervical spine sagittal gadolinium-enhanced magnetic resonance imaging (MRI) showed marked edema and enhancement at the C6-7 vertebrae, vertebral collapse with spinal cord compression, and pronounced soft tissue involvement [Figure 1].

Microbiological workup of muscle and cervical disc biopsy (culture, pan bacterial polymerase chain reaction, periodic acid-Schiff, Ziehl-Neelsen, and Giemsa stains) and

serology for *Bartonella* and *Brucella* were negative. Pathology showed chronic inflammation and focal regenerative changes.

Based on a working diagnosis of synovitis, acne, pustulosis, hyperostosis, and osteitis (SAPHO) syndrome, three monthly infusions of pamidronate were given. However, cervical pain worsened and inflammatory markers increased. Repeated MRIs (sagittal T2 image) showed worsening of vertebral collapse and kyphosis, with severe cord compression [Figure 2]. Major neurosurgery was conducted with anterior and posterior fixation of C6-7. Pathology showed chronic inflammation and fibrosis.

Following surgery, treatment with infliximab along with low-dose methotrexate was initiated, resulting in clinical and laboratory improvement.



Figure 1. Cervical spine sagittal gadolinium-enhanced magnetic resonance imaging (MRI) showing marked edema and enhancement at the C6-7 vertebrae, vertebral collapse with spinal cord compression, and pronounced soft tissue involvement

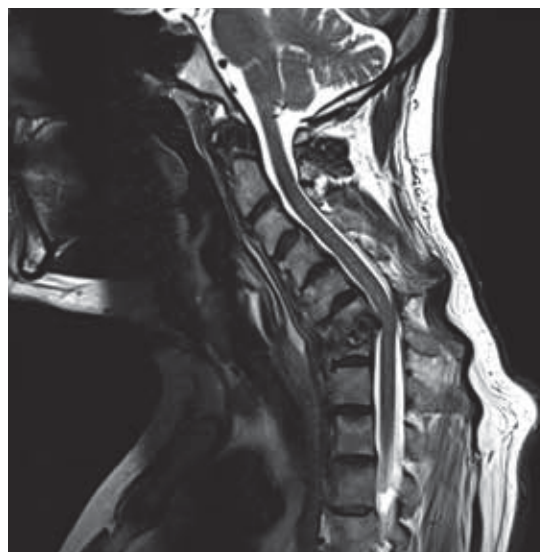


Figure 2. Repeated magnetic resonance images (sagittal T2 image) showing worsening of vertebral collapse and kyphosis with severe cord compression

Spinal involvement in SAPHO occurs in up to 50% of cases, with the cervical spine least commonly involved [1]. In all described cases, prevertebral soft tissue swelling is less than 1 cm in thickness [2]. Our patient showed a rare case of SAPHO syndrome manifested by marked inflammatory involvement of the cervical spine, pronounced prevertebral soft tissue swelling, and rapid deterioration resulting in vertebral body collapse and cer-

vical kyphosis requiring major surgery. This case accentuates the wide clinical spectrum and potential severity of SAPHO syndrome. Early, aggressive anti-inflammatory treatment with tumor necrosis factor-alpha inhibitors may be warranted in such cases.

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References

1. Depasquale R, Kumar N, Lalam RK, et al. SAPHO: What radiologists should know. *Clin Radiol* 2012; 67 (3): 195-206.
2. Laredo JD, Vuillemin-Bodaghi V, Boutry N, Cotten A, Parlier-Cuau C. SAPHO syndrome: MR appearance of vertebral involvement. *Radiology* 2007; 242 (3): 825-31.

Capsule

Bacterial toxin acetylates lysine residues

A toxin produced by the bacterium that causes cholera has a catalytic activity that contributes to its effects on the cytoskeleton of host cells. Zhou et al. determined the protein structure of the Rho guanosine triphosphatase (GTPase) inactivation domain of the toxin from *Vibrio cholerae* and found it to be similar to that of a human fatty acyltransferase. Indeed, the toxin peptide could catalyze fatty acylation of

lysine residues of Rho-family GTPases, which regulate the actin cytoskeleton. Such covalent modification of lysine residues in mammalian proteins had been noted before, but the enzymes responsible were not known.

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Eitan Israeli

Capsule

Lifetime risk of primary total hip replacement surgery for osteoarthritis from 2003 to 2013: a multinational analysis using national registry data

Akerman and co-authors tried to compare the lifetime risk of total hip replacement (THR) surgery for osteoarthritis between countries and over time. In 2003, lifetime risk of THR ranged from 8.7% (Denmark) to 15.9% (Norway) for females and from 6.3% (Denmark) to 8.6% (Finland) for males. With the exception of females in Norway (where lifetime risk started and remained high), lifetime risk of THR increased significantly for both genders in all countries from 2003 to 2013. In 2013, lifetime risk of THR was as high as 1 in 7 women in Norway, and 1 in 10 men in Finland. Females consistently demonstrated the highest lifetime risk of THR at both time points. Notably,

lifetime risk for females in Norway was approximately double the risk for males in 2003 (females 15.9%, 95% confidence interval [95%CI] 15.6–16.1; males 6.9%, 95%CI 6.7–7.1), and 2013 (females 16.0%, 95%CI 15.8–16.3; males 8.3%, 95%CI 8.1–8.5). Using representative, population-based data, this study found statistically significant increases in the lifetime risk of THR in five countries over a 10 year period, and substantial between-gender differences. These multinational risk estimates can inform resource planning for osteoarthritis service delivery.

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Eitan Israeli

Capsule

A revealing repertoire for systemic sclerosis

Systemic sclerosis (SSc) is an autoimmune disease associated with fibrosis and serious complications including pulmonary arterial hypertension (PAH). Abnormal B cell responses have been associated with SSc pathogenesis. de Bourcy et al. analyzed immunoglobulin heavy chain transcripts of SSc-PAH patients enrolled in a clinical study of B cell depletion. SSc-PAH was associated with several B cell development anomalies, particularly underuse of the IGHV2-5 segment and B cell

homeostasis abnormalities. Depletion temporarily reversed these anomalous SSc-PAH disease signatures, and the rate of naïve B cell replenishment could be estimated from baseline measurements. These results define antibody signatures associated with SSc-PAH and reveal how B cell depletion shapes the antibody repertoire during reconstitution.

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