SAPHO: A Case of Skin and Bones

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Introduction

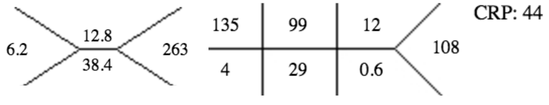
SAPHO is a relapsing inflammatory disease involving the skin, bones, and joints. It is a rare syndrome involving a constellation of common symptoms that requires high clinical suspicion to unify the diagnosis.

Case Summary

This is a case of a 23-year-old female soldier with chronic pain and pustular skin rash over a 10 year period. Her pain remained refractory to an arsenal of analgesics and physical therapy. She was referred to Dermatology for hand and foot dermatitis refractory to topical steroids. Initially, she received a diagnosis of severe Palmoplantar Pustular Psoriasis; however, courses of methotrexate, adalimumab, and etanercept were all ineffective.

Two weeks after stopping treatment with etanercept, she presented with severe neck pain without associated infectious or neurologic symptoms. Plain films showed a possible C5 fracture and subsequent CT and MRI revealed C5-C6 discitis with anterior bridging cervical syndesmophytes. This prompted admission for a CT-guided biopsy with concern for an infectious, inflammatory, or malignant etiology.

On exam, she had scaling palmoplantar plaques, tenderness at multiple costochondral junctions, and cervical midline tenderness with restricted movement in all directions. Labs and cultures were unremarkable other than a mildly elevated CRP.



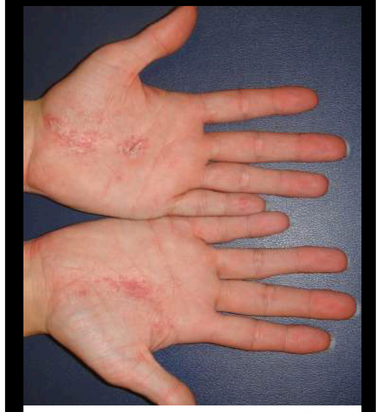


Figure 1. Photo: palmar plaques (2009)

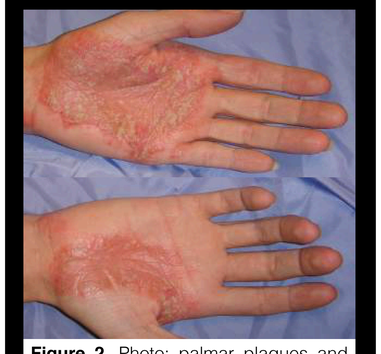


Figure 2. Photo: palmar plaques and pustulosis (2012)

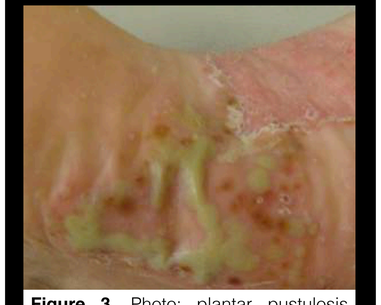


Figure 3. Photo: plantar pustulosis (2012)

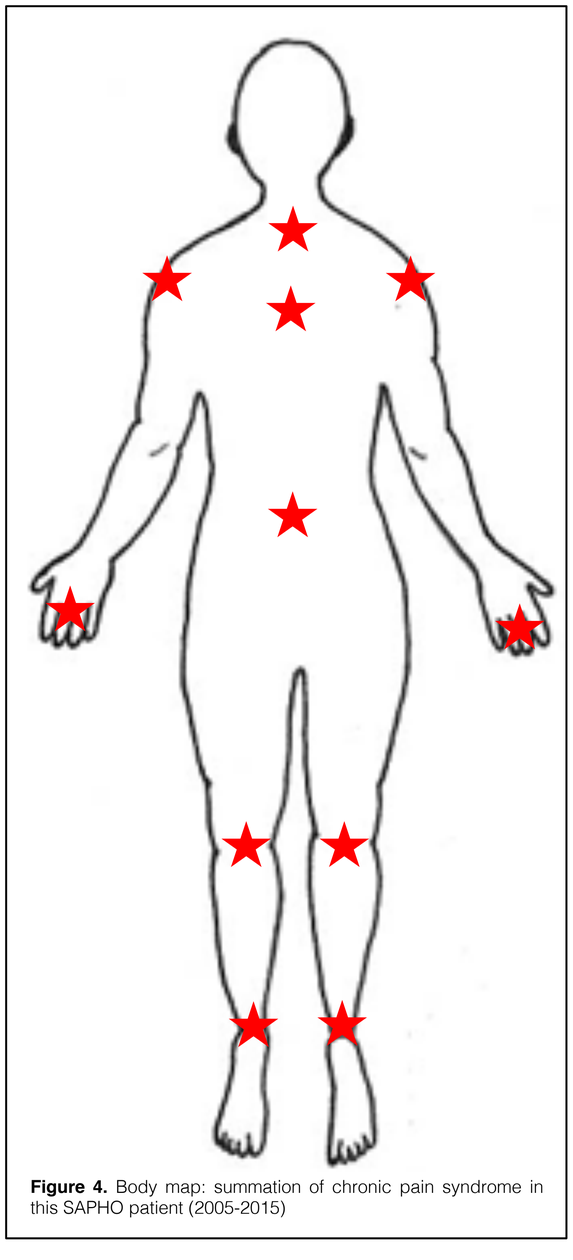


Figure 4. Body map: summation of chronic pain syndrome in this SAPHO patient (2005-2015)

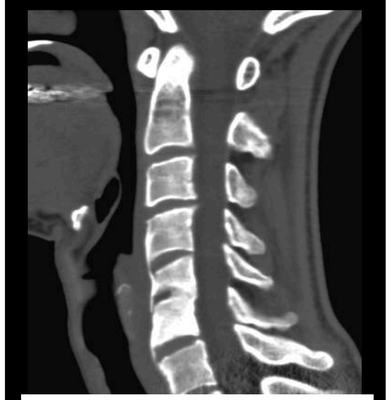


Figure 5. CT cervical: C5-C6 discitis (2013)

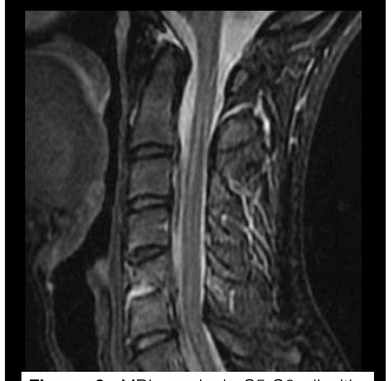


Figure 6. MRI cervical: C5-C6 discitis, anterior bridging syndesmophytes (2013)

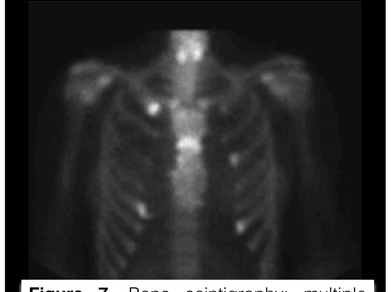


Figure 7. Bone scintigraphy: multiple signs of costochondritis (2013)

Bone scintigraphy showed multiple signs of costochondritis. With this constellation of pustolosis (hands and feet), synovitis (knees), hyperostosis (syndesmophytes), and osteitis (discitis), Rheumatology postulated a unifying diagnosis of SAPHO. General consensus among the consulting services was that the discitis was likely inflammatory, and thus, CT-guided biopsy was deferred. The patient was started on certolizumab with complete disease remission after 6 months of therapy and remains in remission after 12 months of follow up.

Conclusion

The prevalence of SAPHO in the general population is estimated to be 1 in 10,000. It is a disease primarily affecting young women and thought to be underdiagnosed due to lack of awareness in the primary care setting. Delay in treatment can lead to chronic pain and disfigurement of skin and joints. Given its chronicity and relapsing course, continuity of patient care and medical records can aid in recognizing the pattern of disease. Remember:

S ynovitis
A cne
P ustulosis
H yperostosis
O steitis

References

1. Zimmerman et al. Synovitis, acne, pustulosis, hyperostosis, and osteitis (SAPHO) syndrome: A challenging diagnosis not to be missed. Journal of Infection (2016) 72, S106-S114.

2. Kundu et al. Diagnosing the SAPHO syndrome: a report of three cases and review of literature. Clin Rheumatol (2013) 32:1237–1243.