Treatment of Pain in SAPHO (Synovitis, Acne, Pustulosis, Hyperostosis, and Osteitis) Syndrome

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SAPHO (synovitis, acne, pustulosis, hyperostosis, and osteitis) syndrome includes a variety of musculoskeletal disorders and associated skin conditions. Diagnosis can be difficult but should be considered in the differential in patients with lytic, sclerotic, or hyperostotic bone lesions and pain. Appropriate and prompt treatment, including use of nonsteroidal medications such as ibuprofen or naproxen, colchicine, corticosteroids, and bisphosphonates, and use of disease-modifying agents such as methotrexate, sulfasalazine and infliximab, can produce improvement of symptoms. Multidisciplinary care, including the disciplines of rheumatology and dermatology, should be considered.

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INTRODUCTION

This report discusses the clinical presentation, diagnosis, and treatment options for patients with SAPHO (synovitis, acne, pustulosis, hyperostosis, and osteitis) syndrome. We present the case of a patient who was misdiagnosed, leading to delayed and inappropriate treatment that resulted in continued and possibly increased pain.

CASE PRESENTATION

A 50-year-old woman was referred by her primary care provider to the Pain Management Center for evaluation and recommendations for chronic pain control. The patient had a history of pain in her shoulder and chest since she was in her 30s. She described an insidious onset. During the past several years, she noted a constant ache with intermittent episodes of severe deep pain in the bones of her shoulder and chest area. The pain had no relation to position or activity. After experiencing several years of increasing pain, she was eventually evaluated by an orthopedic surgeon at an outside hospital (not part of the system) and underwent an extensive workup. The results of a computed tomography (CT) scan showed bony abnormalities suggestive of chronic osteomyelitis at the sternoclavicular joints and sternum (Figure 1). A nuclear 3 phase bone scan showed increased radiotracer uptake in the right sternoclavicular joint and the manubrium. Delayed images on the bone scan were compatible with chronic osteomyelitis. Magnetic resonance imaging (MRI) of the cervical and thoracic spine showed abnormally decreased signal of the C4 and C5 and T5 through T10 vertebral bodies on T1-weighted sequences (Figure 2), with postcontrast images revealing enhancement of the same vertebral bodies (Figure 3). The patient was diagnosed with sternoclavicular joint hyperostosis and sternal osteomyelosclerosis. She subsequently underwent sternum and clavicle resection (Figure 4). Surgical pathology results of the core bone biopsies were negative for infection or malignancy.

After initial pain relief that lasted for several weeks, she experienced recurrent and continued pain in her right shoulder and chest region. After her surgery she also noted episodic pain in multiple joints, primarily her right ankle, along with self-limiting skin lesions that accompanied the ankle pain. To manage her pain, she tried over-the-counter acetaminophen, which provided minimal benefit. She experienced increasing pain and flare-ups, and thus trials of the short-acting opioid medications hydrocodone/acetaminophen and oxycodone were initiated, with the addition of a long-acting medication,

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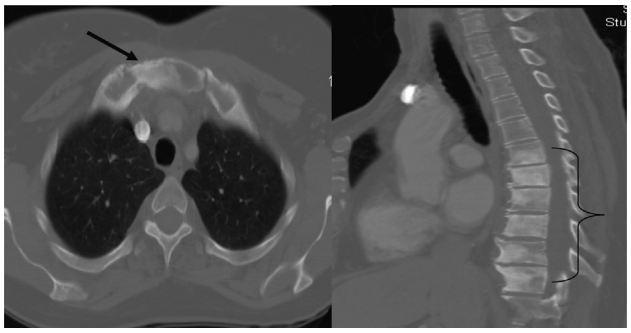

Figure 1. A computed tomography scan (bone window) showing bony abnormalities. The left image shows the sternal-clavicular joint and sternum (black arrow), and the right image shows the thoracic spine (brace).

extended-release morphine, after several months. Despite these escalating doses, the patient noted only modest pain control after 3 years. At the time of presentation, her pain was managed with extended-release morphine, 90 mg 3 times daily, with as-needed oxycodone, 15 mg 4 times a day.

At her initial evaluation at the Pain Management Center, examination findings were unremarkable other than well-healed scars and deformities from her past resection and some enlargement of her right ankle region. Given her history and current medication regimen, a multimodal regimen was considered, including nonsteroidal anti-inflammatory drugs (NSAIDs) such as ibuprofen or naproxen and neuromodulators such as gabapentin or pregabalin. Initially, ibuprofen at a dose of 200 mg 3 times a day, along with a proton pump inhibitor, was added to her opioid regimen. She was also referred to the chronic pain psychology service in the department of behavioral heath and psychiatry for comprehensive pain management.

The patient noted significant pain relief with low-dose ibuprofen without the need for oxycodone. The morphine was tapered and discontinued over a 6-month period, and

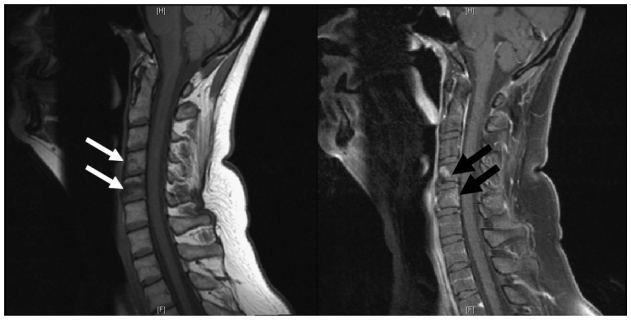

Figure 2. A sagittal T1-weighted magnetic resonance image (MRI) of the cervical spine showing decreased signal (left, white arrows) and a postcontrast MRI demonstrating enhancement (right, black arrows) within the C4 and 5 vertebral bodies.

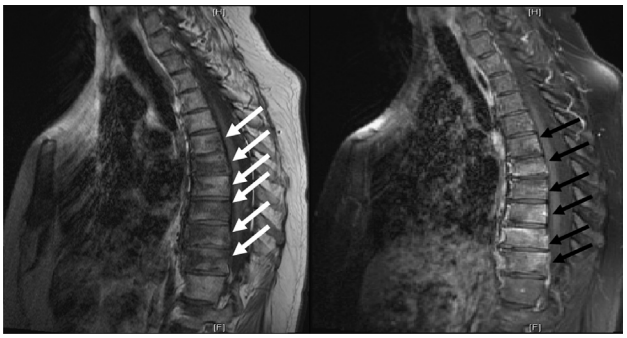

Figure 3. A sagittal T1-weighted magnetic resonance image (MRI) of the thoracic spine showing decreased signal (left, white arrows) and a postcontrast MRI demonstrating enhancement (right, black arrows) within the T5 through T10 vertebral bodies.

the ibuprofen dose was gradually increased to 2400 mg per day in divided doses. However, the right ankle pain recurred with swelling and skin pustules. She was treated with a short course of methylprednisolone with improvement in pain but in not the skin lesions (Figure 5). As a result, a dermatology consultation was obtained.

A dermatology consultant classified the lesions as erythematous, with multiple pustular scaling. Skin biopsy results showed marked superficial to mid-dermal acute and chronic inflammation with microabscesses. The patient had marked benign epidermal hyperplasia with patchy moderate acute inflammation and spongiosis. A Gomori-Grocott

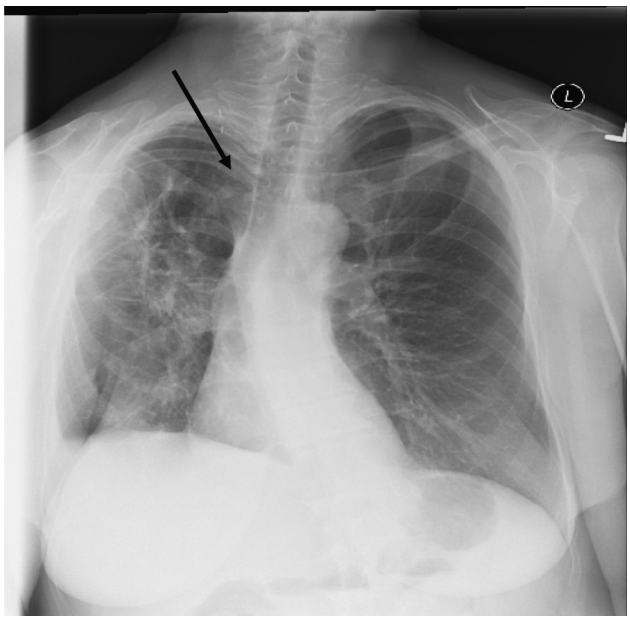

Figure 4. Radiograph of the right clavicle and sternum removal (arrow).