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Skin manifestations associated with chronic recurrent multifocal osteomyelitis in a 9-year-old girl

To the Editor: A 9-year-old girl presented with a 15-month history of severe joint pain limited to the right ankle. She had been treated for fatigue fracture and epiphysitis, but continued to require the use of crutches. The patient had pronounced muscular atrophy of the right leg, swelling and hyperthermia at the heel, and plantar pustulosis. She had mild paronychia on most fingers of the right hand and progressive changes in the fingernails, which were characteristic of nail psoriasis (Fig 1). Thus, psoriatic arthritis or osteitis was suspected, and oral naproxen (200 mg twice daily) therapy was prescribed. Whole-body magnetic resonance imaging (MRI) was performed and demonstrated inflammatory bone lesions, osteolysis, and sclerotic lesions (Fig 2). Laboratory parameters were within normal ranges. Her family history was unremarkable for similar cutaneous or musculoskeletal pathology. Chronic recurrent multifocal osteomyelitis (CRMO) with multifocal bone lesions, plantar pustulosis, and nail involvement was diagnosed. Oral methotrexate therapy (15 mg/week) was initiated and naproxen was continued. After 6 months, the joint pain resolved, and muscular atrophy, palmar pustulosis, and nail lesions improved.

CRMO is an acquired aseptic autoinflammatory bone disease that presents predominantly in girls and is characterized by pain that is worse at night, with or without fever. Typically there is a discrepancy between the mild symptoms and extensive bone inflammation. Sedimentation rate and C-reactive protein (CRP) values may be elevated,



Fig 1. Chronic recurrent multifocal osteomyelitis. Onycholysis, nail pits, oil spots, and discoloration of the nails as well as erythema, hyperkeratosis, pustules on the sole of our 9-year-old female patient.

while the white blood cell count and other laboratory parameters are usually normal. The diagnosis of CRMO is mainly reliant on imaging studies. Conventional radiography initially shows osteolytic bone lesions with development of peripheral sclerosis in the course of the disease. MRI may show early lesions such as edema of bone marrow and inflammation of soft tissue. In order to diagnose CRMO, two major or one major and three minor criteria must be fulfilled.¹⁻³ Major criteria are osteolytic or sclerotic bone lesions, multifocal bone lesions, palmoplantar pustulosis or psoriasis, and sterile bone biopsy with signs of inflammation, fibrosis, or both. Minor criteria are normal blood cell count, good general health, slightly to moderately elevated CRP and erythrocyte sedimentation rate, clinical course of at least 6 months, hyperostosis, association with autoinflammatory diseases other than palmoplantar pustulosis or psoriasis, and a first- or second-degree relative with nonbacterial osteitis, or autoimmune or autoinflammatory disorders.

Some authors believe CRMO to be a juvenile variant of the SAPHO syndrome (synovitis, acne, pustulosis, hyperostosis, osteitis). However, to what extent CRMO and SAPHO present a spectrum of one disease or separate entities remains controversial.²

There is no standard therapy of CRMO; however, nonsteroidal antiinflammatory drugs (NSAIDs) are considered to be first-line treatment with a favorable response rate in up to 80% of patients. Patients may require therapy to control skin and bone lesions, and NSAIDs can be used during attacks or to prevent attacks.⁵ NSAID therapy is usually continued until patients are symptom-free for at least 3 months. When NSAID therapy is inadequate, primary treatment options are bisphosphonates and tumor necrosis factor antagonists,⁵ and strong data

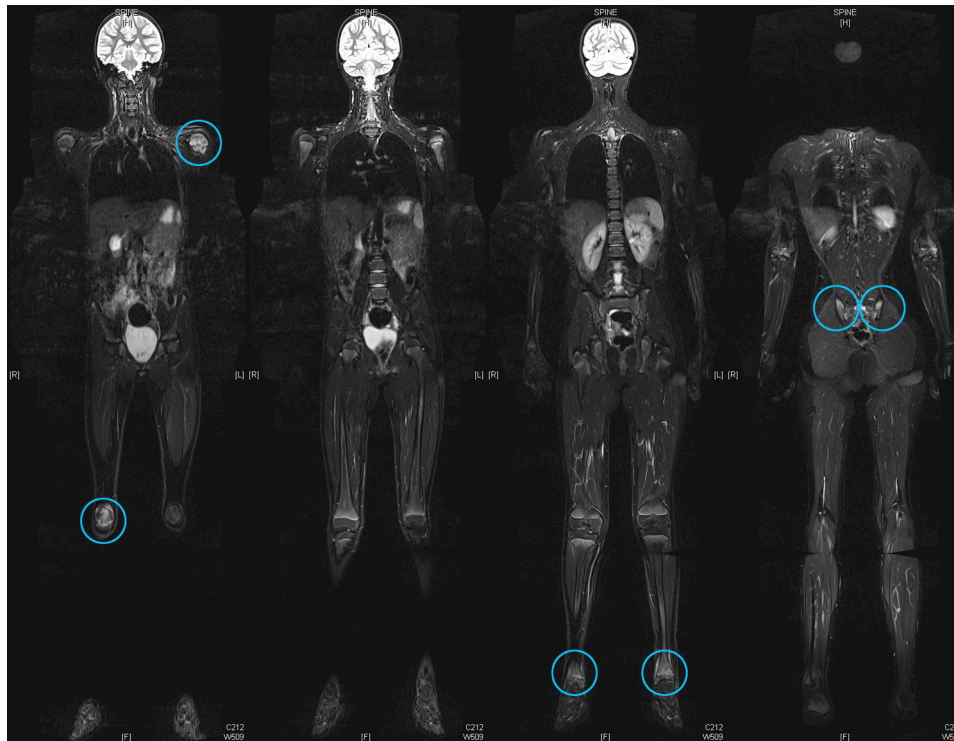


Fig 2. Chronic recurrent multifocal osteomyelitis. Whole-body magnetic resonance imaging detecting inflammatory bone lesions, osteolyses, and scleroses most prominent in the left shoulder, the right foot, and both elbows and ankles.

exist for pamidronate.⁴ Alternative treatments include corticosteroids, methotrexate, sulfasalazine, azathioprine, and colchicine.³⁻⁵ Spontaneous remission is possible; however, CRMO is a disorder that resolves after many years with episodes of remission and relapse, most often without permanent sequelae.

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Are arteriovenous malformations a causative factor for hypertrophic and nodular port-wine stains?

To the Editor: Arteriovenous malformation (AVM), a vascular hamartomatous malformation, is rarely found on biopsy of hypertrophic and nodular port-wine stains (PWS).^{1,2} Digital subtraction angiography (DSA) is the gold standard method for