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TEST YOURSELF: ANSWER

Multicentric reticulohistiocytosis

Sayed Ali · Stephen Huebner

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40-year-old female with bilateral shoulder pain and a skin rash: diagnosis and discussion

Diagnosis: Multicentric reticulohistiocytosis

Discussion

The chest radiograph shows mild cardiomegaly with no evidence of adenopathy or lung disease. A bilateral destructive shoulder arthropathy is evident on the chest film and is better appreciated on the single anteroposterior (AP) views of both shoulders. The joint spaces are mildly widened, and erosions and cystic changes are seen in both glenoids and humeral heads. The distal left clavicle is tapered. An earlier hand radiograph shows marked joint space widening involving the distal interphalangeal (DIP) joints, and to a lesser extent the proximal interphalangeal and carpometacarpal joints of the third through fifth digits. Soft tissue swelling is noted. Of note, periarticular osteoporosis is not present. Extensive skin lesions with a nodular "cobblestone" configuration are present on the face, as well as on the hand where they have a diffuse infiltrative appearance.

The initial impression, given the eroded distal left clavicle and bilateral destructive arthropathy, would be either rheumatoid arthritis or amyloid arthropathy in a patient with chronic renal failure. The absence of osteoporosis and the presence of joint space widening rather than narrowing make rheumatoid arthritis unlikely. The mild cardiomegaly and destructive arthropathy with joint space widening could be seen in renal osteodystrophy with amyloid arthropathy [1], but there was no history of renal disease.

Both systemic lupus erythematosus (SLE) and scleroderma might be considered in the differential diagnosis. Both entities may be associated with soft tissue calcification, which is not evident in this patient. Lupus may demonstrate an erosive arthropathy variably affecting the DIP joints, but not with joint space widening as shown here. Scleroderma can demonstrate distal clavicular erosion as well as a destructive arthropathy and may have soft tissue calcification. However, there is no soft tissue loss or acroosteolysis in the hands to support the diagnosis of scleroderma.

Gout could have this radiologic appearance, but would typically demonstrate joint space preservation initially, followed by eventual joint space narrowing later in the course of disease [2]. Joint space widening as seen in the shoulders and hand in this case, would not be expected. Furthermore, gout would be unlikely in a premenopausal 40-year-old female as it most commonly affects men, and when it occurs in women it generally affects women in their 60s or older.

Another differential would be a neuropathic shoulder in syringomyelia. This is usually an atrophic neuroarthropathy in the shoulder, and it would not demonstrate tapering of the clavicle as seen in this patient. Radiographs of a neuropathic shoulder frequently demonstrate evidence of subluxation or dislocation, which is not demonstrated in this patient.

Bilateral avascular necrosis could account for the irregularity of both humeral heads, but there is no history of steroid use or other risk factors for avascular necrosis.

None of the above conditions would however be expected to have the extensive dermatologic manifestations shown here. Other arthritides that have dermatologic manifestations include psoriatic and reactive (Reiter's) arthritis. Both typically demonstrate periosteal and enthesal new bone formation and eventual joint space narrowing [3], which are not demonstrated here. Reactive arthritis also occurs overwhelmingly in males, and typically has skeletal manifestations at multiple sites including the sacroiliac joints. Osteopenia is usually absent in reactive arthritis, as in this case. Unlike this case, however, the skin lesions in both psoriatic and reactive arthritis are plaque-like without a raised nodular pattern.

The case presentation can be found at doi:0.1007/s00256-012-1507-1.

S. Ali (✉) · S. Huebner
Department of Radiology, Temple University Hospital,
3401 North Broad Street,
Philadelphia, PA 19140, USA
e-mail: saychink@gmail.com

A final important differential is chronic sarcoid arthritis. Sarcoid may show raised nodular skin lesions as well as cysts and bone destruction, but a destructive arthritis is much less common. Characteristic bone manifestations of sarcoid include abnormal trabeculation in one or more phalanges and "punched out" lytic lesions, typically also in the phalanges. Late findings include joint space narrowing and periarticular osteoporosis [2]. In addition, cases of destructive arthritis in sarcoid are typically accompanied by systemic disease. Although the lungs, mediastinum, and hila were normal in this patient, it should be noted that the pulmonary manifestations and adenopathy of sarcoid may be transient or improve with steroid therapy. There was, however, no history of steroid use in our patient.

A myocardial biopsy for clinical myocarditis showed histiocytic infiltration.

Given the lack of osteoporosis, marked bilateral joint destruction with joint space widening in the shoulders and hands, and extensive nodular skin deposits, the diagnosis is consistent with Multicentric Reticulohistiocystosis (MRH).

Multicentric reticulohistiocytosis is a rare condition, with fewer than 200 cases reported in the literature [4]. It most commonly affects middle-aged women, and is characterized by the deposition of lipid laden histiocytes, particularly in the skin and synovium. It has previously been called lipoid dermatoarthritis, since biopsy of the skin lesions may yield several lipid-like components rather than a specific lipid. Other names given to it include giant cell histiocytosis and giant cell reticulohistiocytosis. The intra-articular deposition results in relatively symmetric and painless joint destruction, typically affecting but not confined to the DIP joints [3]. DIP joint involvement is present in 75 % of patients. Less commonly involved joints include the proximal interphalangeal and metacarpophalangeal joints, shoulders, elbows, wrists, vertebrae, hips, knees, ankles, and joints of the feet [5]. Radiologically, there is joint space widening and erosive and cystic change, but no periarticular osteoporosis. The arthritis is characterized by a lack of productive change differentiating it from erosive osteoarthritis. In this case, the bilateral shoulders, left acromioclavicular joint and hands are involved.

Skin involvement precedes the arthritis in 18 % of MRH patients, and occurs simultaneously with the arthritis in 21 % of patients. It follows the arthritis in the remaining cases, beginning an average of 3 years after the onset of arthritis [6]. The skin lesions are characteristic, with a flesh-colored to reddish-brown papular or nodular "cobblestone" configuration and a "coral bead" appearance to the nailfolds [4]. In addition to the skin and articular manifestations, involvement of other organs including cardiac muscle, skeletal muscle, the pleura, and the gastrointestinal tract have been reported [7].

Recognition of the characteristic features on hand radiographs can be helpful in diagnosing MRH. However, in the present case, myocardial biopsy was felt to be warranted because of clinical myocarditis, which can be seen in a variety of entities including, but not confined to, MRH.

Multicentric reticulohistiocytosis is associated with malignancies in up to 30 % of patients [6], and some consider it a paraneoplastic syndrome [8]. Likewise, several autoimmune diseases have been associated with malignancy, including rheumatoid arthritis and systemic lupus erythematosis [9, 10]. MRH also occurs in association with autoimmune diseases in 5–20 % of patients including lupus, scleroderma, hypothyroidism, diabetes, primary biliary cirrhosis, idiopathic inflammatory myopathy, and celiac disease, and has also been reported in association with rheumatoid arthritis and Sjogren's syndrome [11]. Treatment is with steroids or chemotherapeutic agents [12]. Anti-tumor necrosis factor α agents and alendronate may also be efficacious [7].

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