Primary Hypertrophic Osteoarthropathy in a Patient With Rheumatoid Arthritis

To the Editor:

Hypertrophic osteoarthropathy (HOA) is a syndrome characterized by digital clubbing and periostosis of the tubular bones. The secondary form is considered an ominous sign of severe underlying illness, most commonly lung cancer.[1] The primary form of HOA, pachydermoperiostosis, is a rare genetic disorder whose major diagnostic criteria include digital clubbing, periostosis, and pachydermia.[2,3] It is a hereditary sex-linked disease and is not known to be associated with underlying systemic illness.[4]

A recent report[5] reported the case of a patient with primary HOA and ankylosing spondylitis. We describe the case of a 54-year-old man with a familial incomplete form of primary HOA and seropositive rheumatoid arthritis diagnosed at age 25. He presented to our rheumatology outpatient clinic complaining of diffuse join pain and swelling as well as 2 hours of morning stiffness. His review of systems was negative for nightsweats, weight loss, or pulmonary symptoms. On examination, he had swelling of his shoulders, elbows, wrists, metacarpophalyngeal joints, knees, and ankles. He had no thickening of his skin. He was also noted to have clubbing of all his upper and lower extremity digits, which he stated had been present since age 20. He noted that his brother and father had similar-looking digits. The patient's brother came to a follow-up visit and was also found to have bilateral upper and lower extremity digital clubbing (Fig. 1A).

Our patient's evaluation for rheumatoid arthritis included the following: a positive rheumatoid factor of 105 IU/mL and CCP of 66 U, erythrocyte sedimentation rate 53 mm/h, C-reactive protein 7 mg/L, antinuclear antibody negative, thyroid stimulation hormone normal, negative hepatitis B and C serology, and negative tuburculin skin testing. Magnetic resonance imaging of the wrist showed diffuse marked joint space narrowing of the bones of the wrist, multiple erosions at the radiocarpal joint, synovial hypertrophy, and joint effusion all consistent with rheumatoid arthritis (Fig. 1B). Plain radiographs were likewise consistent with RA but also showed diffuse symmetric periostosis (Fig. 1C).

Because of the periostosis and clubbing, though chronic and familial, an extensive malignancy workup was performed, which included negative serum and urine protein electropheresis, negative chest, abdomen and pelvis computed tomography scans, and negative whole body positron emission tomography/computed tomography scan. Since no underlying etiology could be found and the clubbing was familial, a presumptive diagnosis of incomplete (without pachydermia) primary hypertrophic osteoarthropathy was made.

RA is not a cause of secondary HOA and has not been reported in the literature to coexist with primary HOA. Patients with primary HOA tend to have arthropathy but all inflammatory serologies are negative.[3] Similar to the previously reported case of AS and HOA, the coexistence of RA and HOA in our patient is likely coincidental. However, the rare coincidence of 2 cases of the same unusual disease presenting in conjunction with an inflammatory arthritis is serendipitous and piques an interest in searching for a possible pathologic association.

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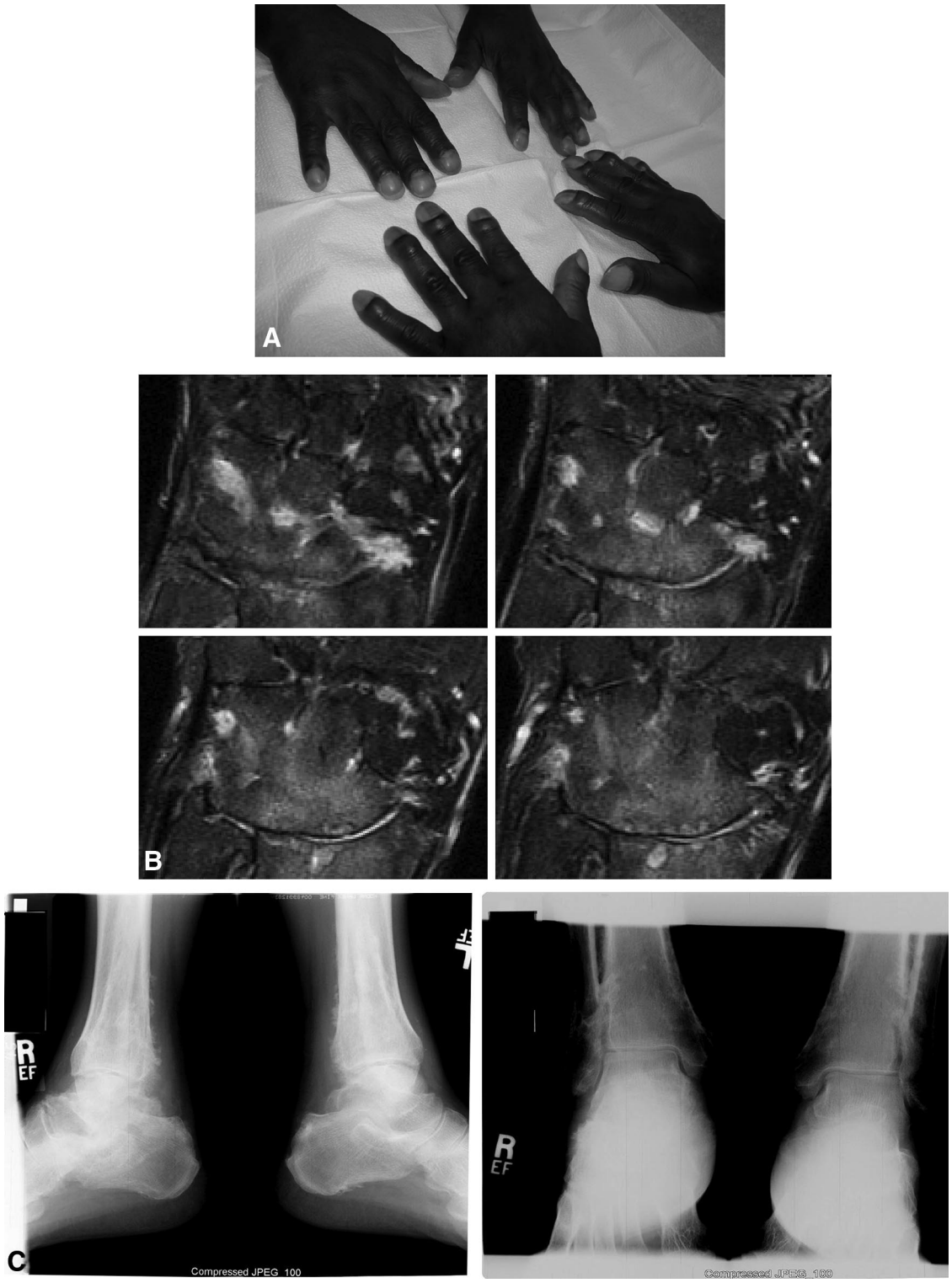

FIGURE 1A. Digital clubbing of patient and brother; 1B. MRI of wrist showing joint space narrowing, erosions, and effusion; 1C. Ankle radiographs showing bilateral symmetric periostosis.

Comment on Recurrent Henoch-Schonlein Purpura in Children

To the Editor:

We read with great interest the recent contribution by Prais et al. in *J Clin Rheumatol*.[1] They reported that 7 (2.7%) of the 260 patients with Henoch-Schönlein purpura (HSP) had experienced recurrences and there were no predictive factors for recurrence.

Although the incidence of recurrence (or relapse) in HSP is variable according to the different studies, it was relatively higher (25%, 52 of 206) in our previous study[2] when compared with the Prais et al.'s report. Also, the recurrence of HSP was related to an older age (>10 years), persistent purpura (>1 month), severe bowel angina at onset, and recurrence itself was an independent risk factor for nephritis thereafter.[2] Similarly, Rigante et al. had demonstrated a possible relationship between persistent purpura and relapsing course in HSP,[3] and Alfredo et al. recently showed that late onset was the only variable related to recurrence of HSP.[4] Although statistical analysis was not possible because of the small number of recurrence in Prais et al.'s study, they showed that 2 of 7 HSP patients (no nephritis at onset) with recurrences had developed nephritis at the second episodes in Table 1.[1]

Prais et al. also described that no further recurrent episodes were reported on long-term follow-up (at least 2 years) and no persistent symptoms were found.[1] However, we should consider a distinct group of multiple recurrent or chronic nonrenal HSP. Alfredo et al. recently pointed out that patients with HSP (especially late onset) should be followed-up over the long-term, because recurrent and chronic cases account for more than 20% of the total, although monocyclic cases were more common among children with early onset disease.[4] Rettig and Cron reported that methotrexate might be needed as a steroid-sparing agent in nonrenal chronic HSP with multiple recurrences.[5]

Therefore, further epidemiologic studies should be performed to evaluate the incidence, predictive factors, clinical course of recurrent HSP, and to elucidate the relationship between recurrence and nephritis.

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The Author Responds

To the Editor:

We thank Shin et al. for their valuable comments on our recently published article.[1]

We reported a relatively low rate of Henoch-Schonlein purpura (HSP) (2.7%) recurrences compared with previous reports. This fact was explicitly discussed in the discussion section, and we believe that the setup of our tertiary pediatric medical center and the specific population (hospitalized children) explains these discrepancies.

Regarding the long-term follow-up, we agree that longer and prospective studies should be performed to assess delayed consequences of HSP. Our group had reported the development of arterial hypertension several years after an episode of HSP even in patients without renal involvement at presentation.[2]

We support Shin et al. conclusions that further epidemiological studies are needed to elucidate the long-term sequelae of HSP.

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