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Acro-osteolysis and calcinosis in patient with scleroderma: A case report

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ABSTRACT

Acro-osteolysis is a rare disease characterized by bone resorption involving the distal phalanges of the hand. We present a unique case of progressive acro-osteolysis of the distal phalanges and articular calcifications in a patient with scleroderma. The calcified deposit in a proximal interphalangeal joint was excised under local anesthesia. The medical treatment was arranged under the supervision of a rheumatologist.

Scleroderma is an autoimmune disease characterized by excessive collagen deposits in connective tissue. It is classified into two subtypes, limited and diffuse. Limited scleroderma, previously referred to as CREST syndrome (calcinosis, Raynaud phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia), usually manifests as cutaneous involvement, mainly affecting the upper extremities. Symptoms include Raynaud phenomenon, thickened skin on the fingers, and musculoskeletal involvement. The fingertips, especially, are involved, with pain and functional impairment of the hands. Acro-osteolysis, which affects the fingertips, is a result of vascular insufficiency or tight skin due to connective tissue diseases such as psoriatic arthritis, scleroderma, and rheumatoid arthritis.[1,2] Calcinosis of the hand, another manifestation of limited scleroderma,[3] can cause painful swelling and tenderness of the involved area. An association between calcinosis and acro-osteolysis has been reported in previous studies. Johnstone et al[4] reported that patients with acro-osteolysis were more likely to have severe calcinosis. Avouac et al[5] also showed that calcinosis and acro-osteolysis were associated, with digital ulceration or vasculopathy in common. We report on a patient who presented with concurrent acro-osteolysis and calcinosis in both hands, and was subsequently diagnosed with limited scleroderma. We also present a review of relevant literature.

Case report

A 63-year-old woman, with a medical history of thyroidectomy for thyroid cancer, presented with sharp pain in the fingertips of both hands. An initial radiograph of the right hand showed mild lytic changes of the second distal phalanx and narrowing of the joint space in several interphalangeal joints (Fig. 1). A few days after initial treatment with nonsteroidal anti-inflammatory drugs for osteoarthritis of the hands, she was lost to follow-up. She presented to our hospital 2 years later with exacerbation of symptoms, including Raynaud phenomenon, caused by exposure of the extremities to cold, which resulted in numbness, coolness, and pallor of the skin. Thickening of the skin on her hand, clubbing, fingertip tenderness, and painful swelling and redness of the fourth proximal interphalangeal joint were noted. Right hand serial radiographs revealed bone resorption of the distal phalanges, or acro-osteolysis, with calcified deposits in several hand and wrist joints (Fig. 1). Laboratory findings revealed a white blood cell count of 8100/mm^3, erythrocyte sedimentation rate of 64 mm/h, C-reactive protein level of 0.06 mg/dL, and a positive antinuclear antibody test. The

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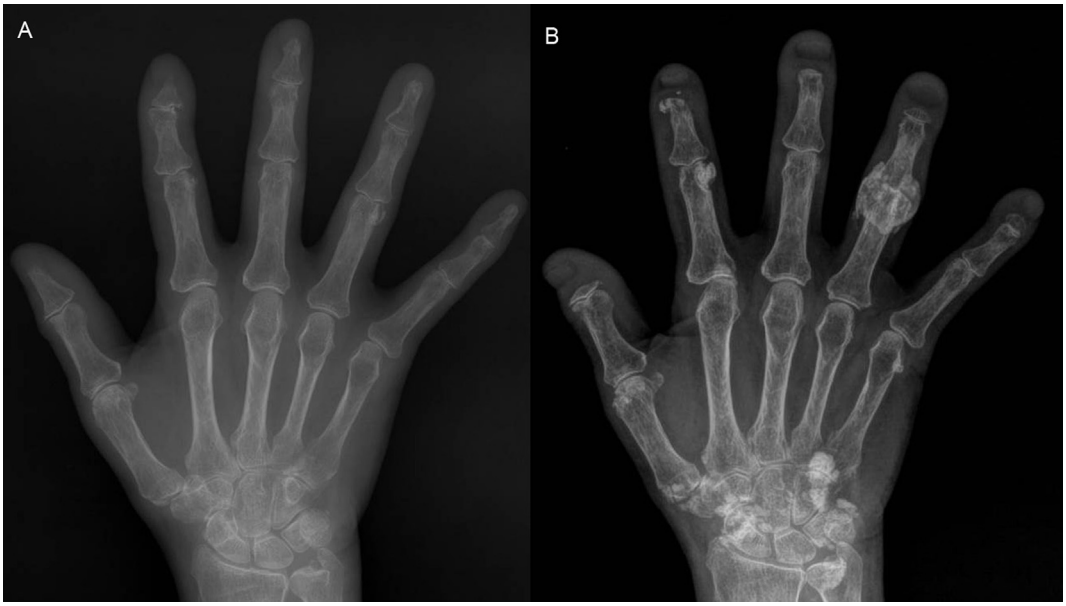

Fig. 1. (A) Initial radiography of the patient's right hand showing mild osteolytic lesions of the 1st and 2nd distal phalanges with joint space narrowing in several interphalangeal joints. (B) Only two years later, aggravating acro-osteolysis is evident in all distal phalanges with multiple calcifications observed.

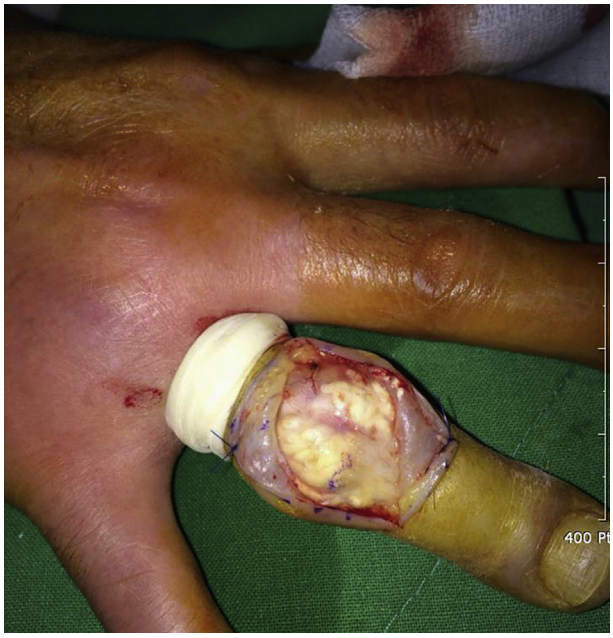

Fig. 2. Calcification embedded in the 4th proximal interphalangeal joint causing inflammation and pain.

calcified deposit of the right fourth proximal interphalangeal joint was excised under local anesthesia (Fig. 2). Periarterial sympathectomy via the palm and volar wrist was planned if acro-osteolysis worsened despite medications. A biopsy and bacterial culture of the joint were performed. The culture revealed no bacterial growth in the joint, but biopsy showed chronic inflammation, necrosis, and fibrosis. She was diagnosed with limited scleroderma based on her clinical and laboratory findings. She is currently being treated by a rheumatologist with methotrexate and a steroid that has significantly improved fingertip pain. Acro-osteolysis did not worsen with appropriate medication for scleroderma. Further surgery, including periarterial sympathectomy, was not needed.

Discussion

Acro-osteolysis is rare, and is characterized by bone resorption involving the distal phalanges of the hand. Bone resorption generally begins on the palmar surface of the tuft. Progressive resorption leads to “penciling” or sharpening of the phalanx. In severe cases, all of the distal phalange may be destroyed. In rare instances, the middle phalanges may also be involved.[2]

Primary acro-osteolysis etiologies include Hajdu–Cheney syndrome, Rothmund–Thomson syndrome, Werner syndrome, Giaccai syndrome, and Haim–Munk syndrome. Primary acro-osteolysis is usually asymptomatic, but is associated with characteristic radiologic findings.[6] However, almost all cases of acro-osteolysis result from secondary causes, including burns, Raynaud phenomenon, scleroderma, psoriatic arthritis, rheumatic disease, and leprosy. Acro-osteolysis in scleroderma correlated with the duration of the disease. Koutaissoff et al[7] reported that scleroderma patients with tuft acro-osteolysis had a statistically significant longer disease duration, averaging 16 years. In their study, acro-osteolysis was also associated with disease activity. We experienced a unique case that acro-osteolysis progressed rapidly to whole aspect of distal phalanges and calcifications were also scattered in several joints due to limited scleroderma in only two years. Our case of acro-osteolysis developed in a relatively short time, with disease activity being the primary contributor.

Secondary etiologies are associated with vascular insufficiency,[8] and aggressive treatment of insufficiency or control of the underlying disease may help prevent acro-osteolysis. There is no radical cure for acro-osteolysis due to scleroderma, but calcium channel blockers, endothelin receptor antagonists, and prostaglandin I2 help prevent Raynaud phenomenon, a prodromal symptom of acro-osteolysis and ulceration of fingertips. Nonsteroidal anti-inflammatory drugs, steroids, or immunosuppressants reduce local symptoms of scleroderma. Studies showed that antiresorptive agents may slow progression of primary acro-osteolysis by reducing bone turnover.[9] Botulinum toxin A injected in the hand was effective for the treatment of vasospastic disorders secondary to collagen vascular diseases, and is an alternative therapy for scleroderma.[10,11]

Surgical interventions include treatment of infections, excision of calcinosis, arthrodesis, and sympathectomy, and can benefit scleroderma-involved hands.[12] Sympathectomy strips the adventitia of common digital arteries and results in sympathetic denervation and improvement in digital blood flow.[13] A long-term follow-up study reported that periarterial sympathectomy was effective in treating digital ischemia due to vasospasm.[14] Most surgical interventions are indicated for vasospastic disorders when conservative treatments, including pharmacologic therapy, fail. We did not perform periarterial sympathectomy due to good response to pharmacologic therapy.

There is no standard treatment for scleroderma-involved hands, and optimal treatment requires collaboration of rheumatologists and hand surgeons. As in this case, excision of calcifications can treat drug-resistant inflammation. Sympathectomy combined with drug therapy may benefit intractable vascular insufficiency of the hands. Because of the potential for rapid progression of acro-osteolysis, as in this case, aggressively ruling out secondary causes and administering appropriate treatment are recommended for patients presenting with fingertip pain and corresponding osteolytic lesions of distal phalanges on radiography.

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