The patient is a 20-year-old woman who presented with sausage-shaped swelling of the hand’s fingers and toes since two years ago. She did not have pain or arthralgia but she complained of swelling of the fingers and toes and morning stiffness of 30 minutes duration. No other symptom was reported.

On physical examination, marked spindling and swelling of the hand’s fingers and toes were observed (Figure 1). No arthritis was noted. Other physical examinations were all normal.

Complete blood count showed the following results: white blood cell count: 6.2 (×10⁹/L) with 69.7% neutrophil and 16.6% lymphocyte, red blood cell count: 3.86 (×10¹²/L), hemoglobin: 11.1g/dL, MCV: 86.8 fL, MCH: 28.8 pg, MCHC: 33.1g/dL, and platelet count: 228 (×10⁹/L).

Serum calcium was 9.4 mg/dL, serum phosphorus was 3.8 mg/dL, and serum alkaline phosphates was 120 U/L.

Radiography of the chest, hands, and the feet were also performed (Figure 2).

What is Your Diagnosis?

See the next page for the diagnosis.
Sarcoidosis has an incidence of 0.02 – 0.2% in the general population.1 Sarcoal bone lesions were first described by Perthes (1920) and Jungling (1928) under the designation "tuberculoid osteitis".2 The lesions occur in only 1% to 15% of all patients with sarcoidosis and are usually asymptomatic. The small tubular bones of the hands and feet are selectively involved (90% of cases). It is important to distinguish these bone lesions from benign acute sarcoid arthropathy. Dactylitis has been reported to occur in 0.2% of all patients with sarcoidosis. Most patients with dactylitis have active systemic sarcoidosis with involvement of several organs.2 Dactylitis is most common in younger individuals. In most cases, the soft tissues surrounding the phalanges are invaded by a granuloma, and bone resorption occurs secondarily.

Our patient presented with swelling of the fingers and toes. In the figure presented here, spindling and swelling of the 4th and 5th fingers of the left hand and the 1st, 3rd, 4th, and 5th finger of the right hand is evident.

The plain radiography of the hands showed soft tissue swelling, lace-like trabecular pattern, cystic changes in the proximal phalanx of the 1st, 4th, and 5th fingers of the right hand, and same changes in the proximal phalanx of the 1st and 5th fingers and middle phalanx of the 4th finger of the left hand.

The findings of lace-like trabecular pattern and cystic changes of the bones led us to perform a chest radiography to rule out sarcoidosis. It showed bilateral symmetrical hilar and right paratracheal lymphadenopathy confirmed by chest computed tomography (Figure 3).

Serum angiotensin convertase enzyme level was measured 68 u/L (reference range: 0.8 – 52). So the diagnosis of sarcoidosis with dactylitis was confirmed. Our patient did not have active involvement of other organs.

Compared with the wider spectrum in children, sausage-shaped digits have only a few known causes in adults including Reiter's syndrome, psoriatic arthritis, sarcoidosis, flexor tendon sheath infections, and gout.3 So, in every patient with sausage-shaped swelling of the digits, careful history taking, physical examination, and appropriate paraclinical tests are needed to find the cause.

The treatment of sarcoid bone lesions is challenging. Prolonged high-dose glucocorticoid therapy is often needed. Bolus methylprednisolone has been advocated to block the activity of the disease, thereby reducing the cumulative glucocorticoid dosage needed.2 Steroids can give symptomatic relief, but bony involvement may progress, and the problem of treating dactylitis with severe underlying bony changes remains.1

The patient received 17.5 mg/d prednisolone and 10 mg/week methotrexate. After one month of treatment, swelling of the digits was much better but longer follow-up of the patient is required to assess the efficacy of the treatment.

In summary, we reported a rare presentation of sarcoidosis accompanied by dactylitis. Sarcoal dactylitis should be suspected in every patient with sausage-shaped digits and lace-like pattern on radiography.

References