Sarcoid dactylitis

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SUMMARY Dactylitis is a rare rheumatological complication of sarcoidosis. It may be accompanied by underlying bone changes, and management is often difficult. We report these 4 cases of dactylitis in which there have been significant bone changes and associated management problems. One case is further complicated by biopsy-proved sarcoid synovitis, uncommon in a British resident, and 2 cases show destructive bone changes, which have rarely been reported in sarcoidosis.

Case reports

Case 1. A nursing sister of West Indian origin presented in 1974 aged 43 with a 9-month history of arthropathy affecting the proximal interphalangeal joints of her right middle and her left index finger and the proximal interphalangeal and metacarpophalangeal joints of the left thumb. Rheumatoid factor was negative. She was treated with indomethacin followed by gold, both to little effect. A synovial biopsy (Fig. 1) of a proximal interphalangeal joint showed noncaseating granulomata. She developed biopsy-proved lupus pernio.

In February 1979 her lupus pernio and arthropathy deteriorated, and she had developed pain and swelling in several of her digits. She was given a 7-week course of ACTH and in January 1980 was given 10 mg of prednisolone on alternate days. This was gradually reduced, but neither her dactylitis nor her lupus pernio improved. It was difficult for her to continue working as sister of a geriatric ward.

X-ray of her hand July 1981 (Fig. 2) showed altered trabecular pattern of many phalanges, a large cystic lesion, marked narrowing of the left thumb proximal phalanx, and periosteal reaction in several
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Fig. 3 Case 1. X-ray left foot, 1981.

Fig. 2 Case 1. X-ray left hand, 1981.

Phalanges. X-ray of her left foot (Fig. 3) showed a similarly altered trabecular pattern and a large cystic lesion in the second toe proximal phalanx. The third and fourth proximal phalanges were abnormally thin. Pictures of her hands (Fig. 4) and feet (Fig. 5) showed marked soft tissue changes generally corresponding to the underlying bony changes on x-ray.

In October 1981 prednisolone was reduced; the dactylitis and lupus pernio have remained unchanged. She continues as a nursing sister and despite some discomfort is now managing well.

Case 2. In April 1977 a male Indian doctor aged 40 presented with bilateral conjunctivitis, right uveitis, and left facial palsy. Chest x-ray showed bilateral hilar adenopathy and a conjunctival biopsy non-caseating granulomata. He was treated with 40 mg of prednisolone daily and steroid eye drops. As steroid therapy was reduced he developed painful swelling of several fingers lasting some months. Steroid therapy was discontinued in May 1980, and in June 1980 he presented with a history of 3 months' pain and swelling of several fingers. Aspirin had not relieved his symptoms. On examination he had pain and swelling in the proximal phalanges of his left index and ring fingers and in the middle phalanx of his right little finger. A year later there was increase in pain and swelling in many digits, and he was having considerable difficulty in carrying out his work as a family doctor.

Selected x-rays of his left ring finger proximal phalanx and right little finger middle phalanx taken in June 1980 showed marked alteration of the trabecular pattern and destructive changes (Fig. 6). An x-ray of the right forefoot taken in June 1981 (Fig. 7) showed generally abnormal bony architecture, and a terminal tuft of the middle toe distal phalanx had been lost. By October without specific treatment for his dactylitis his pain and swelling had improved. He was started on systemic steroids 40 mg a day for recurrent uveitis, and his dactylitis remained quiescent.

Case 3. A West Indian man working in a leather factory developed lupus pernio of his nose and upper airways in 1971 at aged 55. In 1973 he complained of swelling of his left index finger. By 1975 he required treatment with systemic steroids for worsening nasal obstruction. Symptomatically his nose and dactylitis improved. The bony changes, however, have progressed as shown (Fig. 8).

Case 4. A West Indian car mechanic presented in 1979 aged 24 with a 2-month history of pain and
swelling in several fingers and toes. On examination he had cervical and axillary adenopathy, sausage shaped swelling of several fingers, and recent dystrophic changes in his toenails. The dactylitis progressed and the nail changes affected his hands as well. X-rays in April 1981 (Fig. 9) showed loss of the normal cortex in many of the phalanges, with multiple small lytic lesions.

In July 1981 a lymph node biopsy showed non-caseating granulomata. His dactylitis had so worsened that he was unable to work (Fig. 10). He was treated with a course of local radiotherapy to his hands and feet. A total of 1500 rads in 3 fractions was given to his right hand and 1000 rads in 4 fractions to his left. Despite this therapy the swelling and ulceration progressed.

In September 1981 he was started on prednisolone 20 mg daily, chloroquine 250 mg on alternate days, and methotrexate 5 mg weekly. By November 1981 the soft tissue changes were much improved and the mobility of his hands increased. He could return to work.
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Fig. 6 Case 2. X-ray left ring finger and right little finger.

Fig. 7 Case 2. X-ray right forefoot.

Fig. 8 Case 3. X-rays left index finger, 1973, 1974, and 1978.

Fig. 9 Case 4. X-ray of hand, April 1981.
By February 1982 he was taking only 7·5 mg of prednisolone daily. By March his symptoms had recurred and he was having considerable difficulty in working.

Discussion

Sarcoidosis has an incidence of 0·02–0·2% in the general population.1 Rheumatological complications are common, and arthritis occurs in up to 40% of these patients.2 An acute arthritis associated with erythema nodosum and hilar adenopathy is most common and tends to disappear without residua. A chronic arthritis also occurs, and patients usually have evidence of active sarcoidosis in other organs.4 Generally the x-rays of sarcoïd arthritis appear normal unless they are associated with bony lesions of the phalanges.5 Tenosynovitis is also reported.5 6

Bone disease occurs frequently. Depending on radiological investigation the incidence varies from 1·2 to 13%, but it is asymptomatic in 42% of these patients. The x-rays show altered trabecular pattern, minute cortical defects, and cystic lesions. Sarcoid dactylitis occurs in 0·2% of patients and may or may not be associated with underlying bone changes due to granulomas involving the bone marrow.7

These 4 patients have both soft tissue changes and marked bony involvement. In 2 patients there are destructive bony changes. Such changes are rare and Neville et al. reported 3 cases in their series of 508 and Mandi et al. one in 725.8 Case 1 is further complicated by biopsy-proved sarcoïd synovitis, rare in a British resident, even of West Indian origin.

In all 4 cases management was a problem and their working lives were interfered with. Steroids are known to produce symptomatic relief in some patients, as in case 3. However, it was necessary to treat case 4, of universal dactylitis, with chloroquine and methotrexate after failure to respond to radiotherapy. He rapidly relapsed when treatment was reduced to 7·5 mg of prednisolone only. This case compares well with a report of a Caucasian also presenting with subacute universal dactylitis who had bony involvement and some destructive changes.9 The authors do not comment on the management of their case but point out the rarity of such a presentation of sarcoidosis.

Despite soft tissue improvement bone disease can progress, as in cases 2 and 3, where dramatic destructive changes continued. Of Neville et al.’s 3 patients with destructive bony changes 2 required amputation for pain relief.7

In these cases sarcoïd dactylitis is associated with evidence of granulomatous sarcoidosis in other organs (Table 1). Besnier in 1889 originally described the association of sarcoïd dactylitis with lupus pernio.10 An increased incidence of bony

Table 1 The associated features present in the 4 patients

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involvement occurs with eye disease. None of our patients is Caucasian, and it is known that the incidence of skin granulomas in sarcoidosis varies both geographically and racially, which may be significant. The difficulties in treating the patients reported here may be a reflection of the severe underlying bony changes present. Steroids can give symptomatic relief, but bony involvement may progress, and the problem of treating dactylitis with severe underlying bony changes remains.

We are most grateful to Dr D. G. James for his helpful advice in the management of case 4.

References