Sarcoidosis in bone

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Summary

Radiologically demonstrable bone change is found in about one in every six patients with sarcoidosis. The hands and feet are affected and rarely the trunk, an unusual distribution for a disseminate disease.

Osteoporosis, cortical thinning and mottled rarefaction are the earliest signs, followed by cystic changes due to the coalescence of adjacent lesions and perhaps pathological fractures.

The nasal bones may be directly involved in patients with lupus pernio. Changes in the rest of the skull, in the spine and pelvis are rare and may be bizarre.

Whatever the aetiology, sarcoidosis is a general disseminate disease. It would be remarkable if skeletal involvement did not occur but its radiological demonstration is relatively infrequent. The commonly quoted incidence of radiological change (Holt & Owens, 1949; Stein, Israel & Sones, 1956) is about 17%.

Considered as a general disease the distribution of the radiological changes in bone is unusual. The periphery of the body, the hands and feet are affected rather than the central parts of the skeleton. The spine, pelvis and skull are rarely shown to be abnormal. The distribution pattern contrasts with that of other diseases capable of wide dissemination, as for example, carcinoma and tuberculosis.

Local involvement of bone as part of a local sarcoid lesion occurs and no doubt this is the cause of the changes in the nasal bones in the sarcoid lesion traditionally referred to as lupus pernio (Curtis, 1964). It is a curse of diseases of uncertain aetiology to be bedevilled by innumerable descriptive titles.

The peculiar nature of the bone changes in the hands and feet was first recognized by Kriebich (1904) over 60 years ago. Biopsy was naturally practised in the early years, though this is indefensible on peripheral bone now. Perivascular infiltration of the Haversian systems by sarcoid deposits gives rise to general osteoporosis and cortical thinning. Radiologically these are the earliest detectable signs. The later cystic changes are due to the gradual coalescence

of adjacent lesions which, when they heal, are replaced by fibrous tissue and so persist as radiological cysts. Pathological fractures can occur in very active cases.

In the bones of the hands and feet the destructive process in the phalanges results in a gradual enlargement of the bone lacunae so that a mottled rarefaction is produced with tiny dots of diminished density in the medulla. The process may be uneven and the distal ends of the proximal and middle phalanges and the proximal ends of the distal phalanges often show this change best. As the process increases the cortical bone is thinned and expanded (spina ventosa) and pathological fractures may occur. Fleischner (1924) illustrated examples of the mutilation that can occur. The absence of periostitis is a feature and probably is in keeping with the lack of bone reaction as is also the absence of sequestra and, clinically, the painless nature of the condition.

Schaumann (1926) pointed out that the medullary cavity of the bone is the primary seat of the disease and medullary bone may be infiltrated throughout the body skeleton without any abnormality being radiographically detectable provided that cortical bone destruction has not occurred. This is a cause of diagnostic difficulty not peculiar to sarcoidosis.

Bone changes in the trunk are rare. Bloch, Movson & Seedat (1968) illustrated a classic case of extensive sarcoid disease in a Bantu boy of 15 who had iridocyclitis, bilateral parotid swellings, large liver and spleen and maculo-papular skin rash. The patient complained of back pain which radiography showed was due to the presence of a cystic lesion in the body of the eleventh dorsal vertebra round which there was some marginal sclerosis. A similar lesion was shown in one ilium and there were changes in the hands and enlarged hilar glands. Biopsy of the lesion in the ilium showed granulomatous sarcoid lesions.

Olsen (1963) presented the fourth recorded case of skull involvement. His patient was a woman of 51 years who had glands in the neck and an ulcerated vulval lesion. Radiography of the skull showed

FIG. 1. Hands of a young male, aged 37, with sarcoidosis. 'Cystic' areas in distal ends of middle phalanx of both ring fingers and a similar area in proximal phalanx of right middle finger. Trabecular absorption in some areas, e.g. base of middle phalanx right middle finger. Pathological fractures of bases of distal phalanx of both ring fingers.

several sharply discrete areas of bone destruction in the vault with no surrounding reaction and these areas returned to normal spontaneously in 22 months. All the areas affected were biopsied with positive result for sarcoid.

To conclude, the bone changes encountered in patients with sarcoidosis are such as might be expected to occur in a relatively benign but chronic disseminate condition where reparative tissue reaction is poor. It is clearly beyond the scope of radiology to suggest any aetiological factor but it would seem possible that an infective cause of such lesions could certainly account for the diversity and sometimes strange distribution of the bone lesions encountered.

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Plate 1

Explanation to Plate 1

1a and 1c. A male aged 32 years with lupus pernio affecting the left ear, right nostril and both great toes; associated cystic changes in the proximal phalanges of both great toes and slight glandular enlargement in the superior mediastinum.

1b. A female aged 47 years with widespread granulomatous infiltration of the scalp and marked loss of hair. Chest X-ray showed general increase in lung marking consistent with sarcoidosis.

1d. Annular lesion of sarcoidosis involving the scalp margin.