International Orthopaedics (SICOT) (1987) 11: 367–369

International
Orthopaedics

Lymphangiomatosis of the arm with massive osteolysis

A case report

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Summary. *A case of massive osteolysis of the arm is described where the typical bony lesions were associated with soft tissue lymphangiomatosis.*

Résumé. *Description d'un cas d'ostéolyse massive du membre supérieur dans lequel des lésions osseuses typiques étaient associées à une lymphangiomatose des parties molles.*

Key words: *Osteolysis, Lymphangiomatosis, Arm*

Case report

A 54-year-old man, was admitted to the Orthopaedic Clinic of the University of Pavia on June 10th, 1975 because of progressive weakness and stiffness of the lower limbs.

There was no relevant family history. The only injury reported was an acromio-clavicular dislocation at the age of 17 years. When he was 20 years old, a soft swelling around the left elbow was first noticed. It grew slowly and eight years later a biopsy was carried out, on which the diagnosis of lymphangioma was made. The lesion continued to grow slowly and later new swellings appeared on the shoulder and wrist. Massive bone resorption of the shoulder bones was first observed at the age of 35.

Attempts were made to remove the swellings when the patient was 38 and 48 years old, but on both occasions the operation had to be abandoned because of severe bleeding. Three years later, when he was 51 years old, the left arm was irradiated.

On admission, examination showed an increase in the length and volume of the left arm due to multiple soft and flabby swellings, with deep folds of the skin in the shoulder and elbow regions. Smaller swellings were present on the wrist, middle, ring and little fingers (Fig. 1). The skin was white and there was no increase in local temperature. The clavicle, scapula and the upper end of the humerus could not be felt. The function of the shoulder and elbow was entirely lost, but there was an abnormally wide range of passive movement of the shoulder. Active movements of the wrist and fingers were preserved. Skin sensation was normal. A marked cervical kyphosis, with a limited range of movement of the neck, was present.

Radiographs revealed that the whole of the scapula and the outer part of the clavicle had dis-

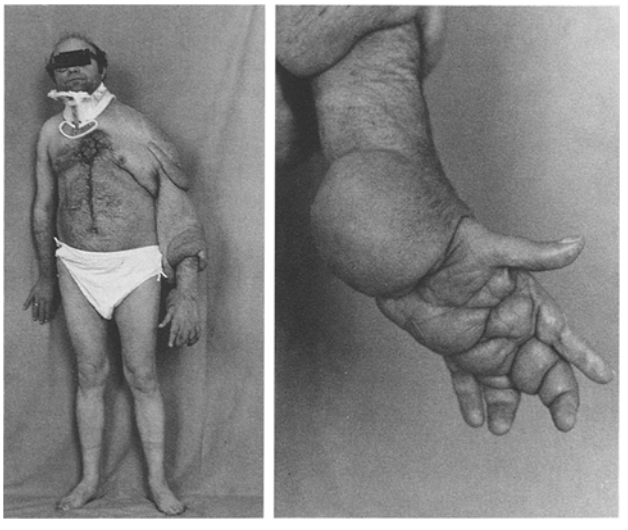

Fig. 1. Soft tissue lymphangiomatosis of the left upper limb

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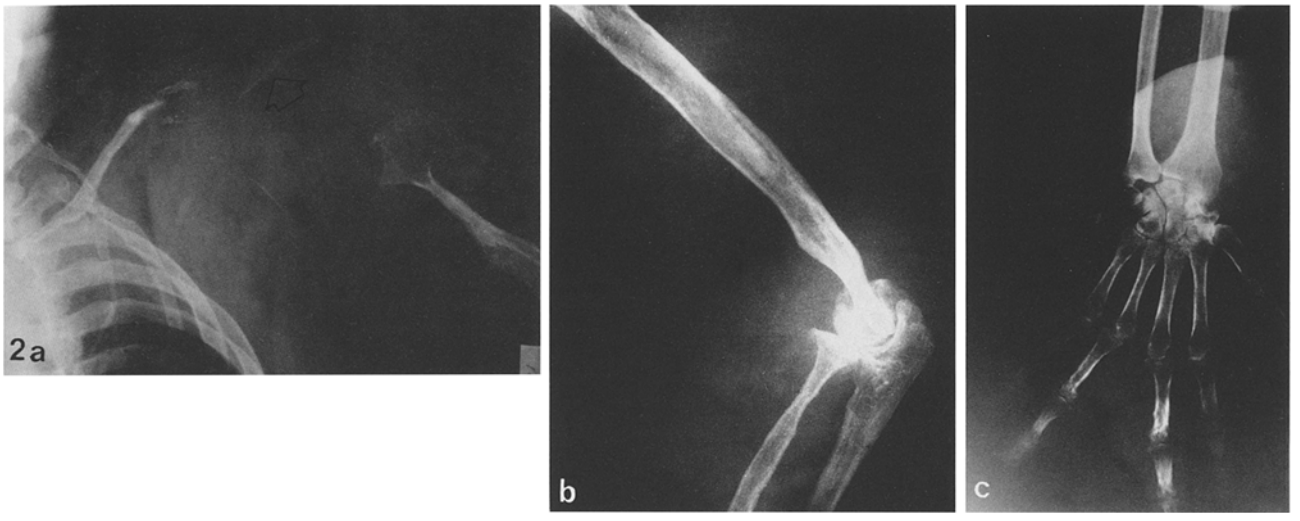

Fig. 2. a The outer part of the clavicle and the entire scapula has been resorbed (a remnant of acromion is shown by an *arrow*). The remainder of the clavicle and the first rib are reduced to a thin band. The humeral head is irregular and osteoporotic. There is tapering and concentric narrowing of the diaphysis; **b** Similar lesions are seen in the distal humerus, and in the proximal radius and ulna; **c** Tapering and concentric narrowing of the metacarpals and phalanges of the middle, ring and little fingers

appeared, while the medial part of the clavicle and the first rib were reduced to a thin band. The upper end of the humerus was severely deformed. The head was irregular and osteoporotic. The proximal shaft was tapered and concentrically narrowed. Similar lesions were present in the distal part of the humerus, and in the proximal radius and ulna. In the hand, half of the carpal scaphoid was resorbed, while the metacarpals and phalanges of the middle, ring and little fingers had undergone varying degrees of tapering and concentric narrowing (Fig. 2). Osteolysis of the 5th and 6th cervical vertebrae had caused the acute kyphosis of the neck (Fig. 3). Arteriography

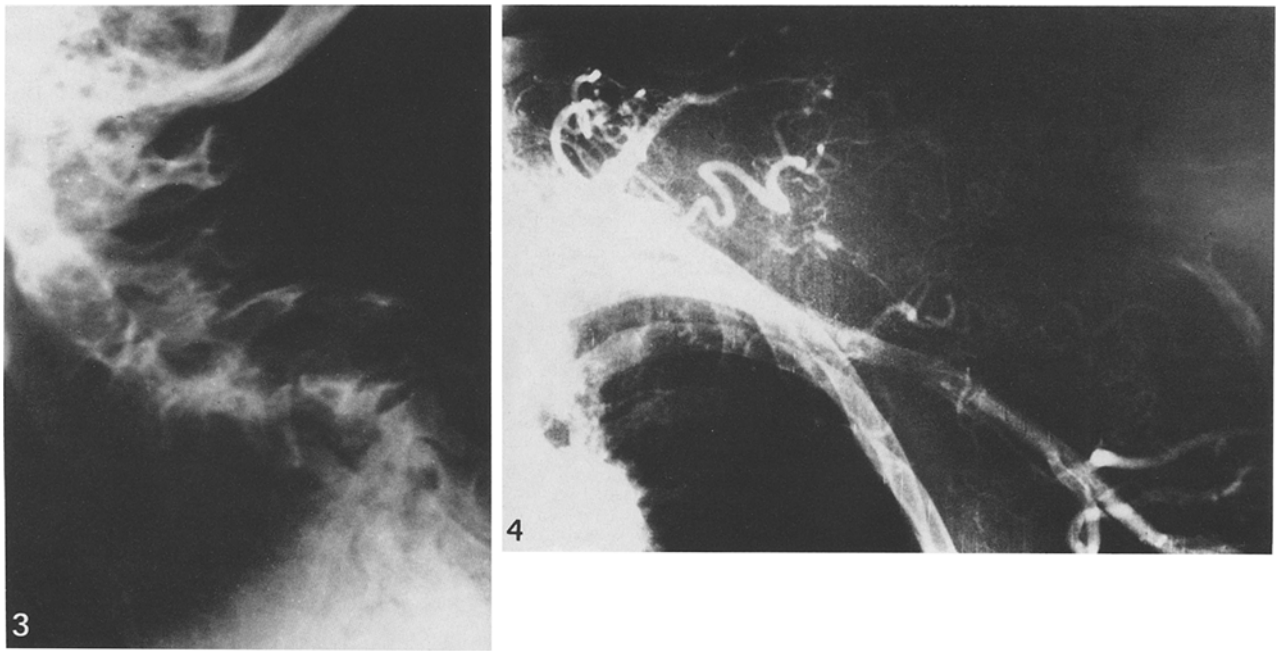

Fig. 3. Acute kyphosis of the cervical spine due to collapse of the 5th and 6th vertebrae

Fig. 4. Arteriography of the shoulder region shows convoluted, collaterals of the subclavian artery in the area of the absent bones

showed a network of convoluted collateral vessels from the subclavian artery extending into the region of the absent bones (Fig. 4), and the emptying time was longer than normal.

Laboratory tests were normal, except for an increase in eosinophils. The only treatment used was a cervical collar, and after 6 years, the disease had not progressed further.

Discussion

The association between massive osteolysis and haemangiomatosis or lymphangiomatosis has been firmly established by Gorham [5] and by Gorham and Stout [6] in 1955 and 1956. This has been confirmed in most of the subsequent reports of the disease [1, 2, 3, 9, 10, 11, 12]. In only three cases were skin haemangiomata present in the region of the osteolysis [4, 7, 8]. The peculiarity of our case is the striking lymphangiomatosis of the soft tissues and the extent of the bony lesions from the cervical spine and first rib to the phalanges of the fingers. To our knowledge, such wide involvement of bones has not been reported before.

Soft tissue lymphangiomatosis was observed first at the elbow in this case, and thereafter spread proximally and distally. We do not know whether the soft tissue lesions preceded the osteolysis or how the bone resorption progressed. When the patient came to us, there was a remarkable topographical correlation between the soft tissue swellings and the bone involvement.

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