Hypertrophic pulmonary osteoarthropathy in a patient with pulmonary alveolar microlithiasis

Salih Emri, Lütfi Cöplü, Z Toros Selçuk, A Altay Sahin, Y Izzettin Barış

Abstract
Hypertrophic pulmonary osteoarthropathy occurred in a patient with proved pulmonary alveolar microlithiasis, an association not previously reported.

Pulmonary alveolar microlithiasis is an uncommon disorder of unknown aetiology, characterised by microliths within the alveoli. Diagnosis is usually made from the radiographic appearance.1,2

Finger clubbing has been reported in pulmonary alveolar microlithiasis,3,4 but not hypertrophic pulmonary osteoarthropathy. We report such a patient.

Case report
Pulmonary alveolar microlithiasis was diagnosed in a 44 year old woman and confirmed by open lung biopsy. Three years later she reported intermittent bilateral pain in the knees and ankles of four months' duration. The pain was associated with swelling at rest and aggravated by walking. She also noted increasing dyspnoea on exertion, and a cough producing yellow sputum. There was no relevant family history. On examination she was obese and had finger clubbing. Her wrists, ankles, metacarpophalangeal joints, and metatarsophalangeal joints were swollen, erythematous, and tender in response to light palpation. Auscultation of the chest disclosed bilateral basal end inspiratory crackles. The erythrocyte sedimentation rate was 80 mm in one hour. Rheumatoid factor was present and C reactive protein was increased. Arterial blood gases while she was breathing air were: pH 7-42, arterial carbon dioxide tension (Paco2) 4.2 kPa, arterial oxygen tension (Pao2) 9.9 kPa. Pulmonary function tests showed mild restriction and a transfer factor (TLCO) of 39% predicted. A chest radiograph (fig 1) showed very fine, sand like micronodules of calcific density, diffusely affecting both lungs. The appearance was unchanged from a radiograph taken 10 years earlier.

Computed tomography of the chest showed diffuse bilateral calcified densities, predominately in the lower zones, with parahilar accentuation. The micronodules were calcific density and had a fine, well-defined calcific rim characteristic of this condition. The chest radiograph showed fine linear opacities, indicating bronchiolar involvement. The arterial and venous pressure was 12/8 kPa. The radiographic appearance of the chest was compatible with pulmonary alveolar microlithiasis,4,5 and the clinical features were consistent with the diagnosis of hypertrophic pulmonary osteoarthropathy.

Use of the Denver pleuroperitoneal shunt in the treatment of chylothorax secondary to filariasis

Pleuroperitoneal shunting has been used in the treatment of recalcitrant chylothorax for almost a decade and its successful use has been described in both congenital neonatal chylothorax6 and adult cases7 of traumatic and malignant aetiology. We believe, however, that this is the first case in which the use of the pleuroperitoneal shunt has been described in chylothorax secondary to filariasis.

Hypertrophic pulmonary osteoarthropathy in a patient with pulmonary alveolar microlithiasis

Figure 2  Bone scan showing increased uptake of radioisotope at the diaphysis and metaphysis of the metacarpal bones and both wrists.

Discussion

One hundred and sixty nine cases of alveolar microlithiasis have been reviewed.\(^2\) Clubbing of the fingers has been described in this condition but other features of hypertrophic pulmonary osteoarthropathy have not been reported.\(^7\) Joint symptoms in hypertrophic pulmonary osteoarthropathy range from mild arthralgia to a restricted range of movement and even ankylosis in advanced cases.\(^7\) Synovitis, frequently asymmetrical, affects metacarpophalangeal joints, wrists, elbows, knees, and ankles.\(^7\) Complement levels and results of rheumatoid factor and antinuclear antibody tests are usually normal but the erythrocyte sedimentation rate is increased.\(^7\) The presence of rheumatoid factor in our patient may be related to its occurrence in fibrosing alveolitis, in which 40% of patients were found to have it.\(^9\)

Radiographically, detectable periosteal thickening along the shafts of the long and short bones confirms the presence of hypertrophic pulmonary osteoarthropathy.\(^10\)\(^11\) Radionuclide bone scans are helpful where radiographic features are minimal or absent. Periarticular and periarticular uptake are consistent with synovitis. Isotopic imaging of the hands and feet is a sensitive method of determining the presence and extent of hypertrophic pulmonary osteoarthropathy.\(^12\) The diaphysis and metaphysis of the metacarpal and metatarsal bones may also be seen to be affected, with increased uptake at the distal phalanges.\(^10\) The skeletal survey in our case showed subperiosteal bone formation in the distal tibia and the bone scan showed slightly increased uptake in both wrists and metacarpophalangeal joints. This, associated with finger clubbing, confirmed the presence of hypertrophic pulmonary osteoarthropathy in the presence of definitely diagnosed pulmonary alveolar microlithiasis.