Intra-osseous synovial sarcoma

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Summary

Intra-osseous synovial sarcoma is very rare and its mode of origin is conjectural. Our patient's tumour was situated within the lower femur. The sites of origin of the few intra-osseous synovial sarcomas so far documented may support the concept of intra-osseous synovial rests.

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Tenosynovial sarcomas may take a variety of morphological forms and may be found at a variety of sites, the commonest being the thigh, foot, knee, shoulder and forearm. In all these common locations and even in other rare sites, e.g. neck, abdomen and back, the tumour arises within soft tissues.

We have recently encountered a primary synovial sarcoma which arose *de novo* within the femur. Because of the extreme rarity of such an occurrence and the possible support which it provides for the concept of 'synovial rests' within long bones, the following case is reported.

Case report

A 16-year-old girl was admitted to hospital with a fracture of the lower end of the right femur resulting from a fall while walking. Radiography (Fig. 1) showed that the fracture was due to a malignant tumour which appeared to be arising from within the bone. Biopsy of the lesion confirmed the presence of a malignant tumour (Fig. 2), biphasic in character and composed of plump, spindle-shaped cells showing abundant mitotic activity, while other areas showed narrow spaces lined by pseudo-epithelial cells. On histological examination the appearance was typical of the biphasic synovial sarcoma. Scattered osteoclast-like giant cells were present in some areas. Tomography of the chest revealed no evidence of metastatic spread. The patient subsequently underwent subtrochanteric amputation. Examination of the amputated leg confirmed the intra-osseous origin of the tumour. Postoperative chemotherapy was refused.

Discussion

Synovial sarcoma may involve bone, either by direct spread or by haematogenous metastasis. The origin of such a tumour from within bone appears to be an exceptional event, which is not mentioned in modern texts; in a survey of the recent literature we have been able to find only a few reports. The tumour reported by Srivastava and Gupta[1] in 1974 also involved the lower femur; synovial sarcomas arising within the tibia have been reported by

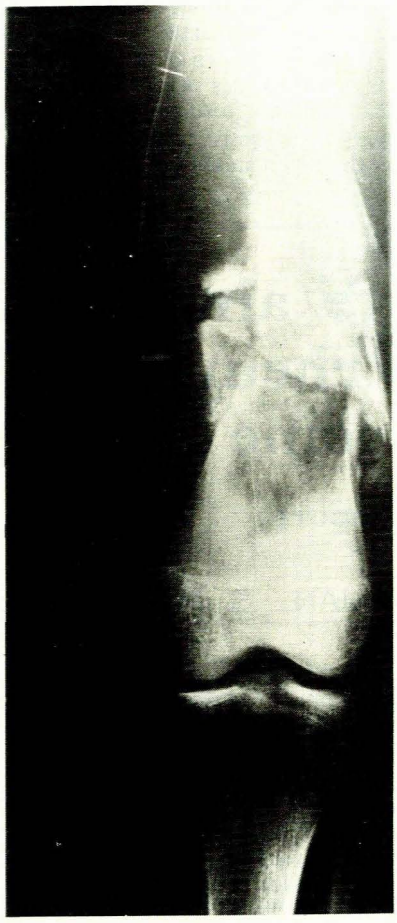

Fig. 1. Radiograph of right leg showing pathological fracture through an area of radiolucency caused by an intra-osseous osteolytic malignant tumour. Subperiosteal formation of new bone is observed proximal to the fracture.

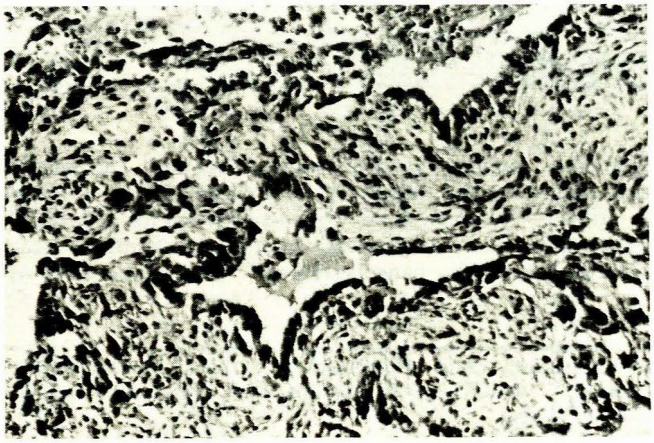

Fig. 2. Microscopic appearance of the tumour showing the formation within the sarcoma of spaces lined by pseudo-epithelial cells (H and E x 150).

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others[2,3] and Evans *et al.*[4] reported a patient with a congenital synovial sarcoma of the humerus. A synovial origin has also been mooted for adamantinoma of the tibia.

The mode of origin of synovial sarcoma from within bone is conjectural. Srivastava and Gupta[1] proposed that fibroblasts and other mesenchymal cells within bone may be stimulated to differentiate into synovial tissue which may undergo malignant change. In a discussion regarding the pathogenesis of simple bone cysts, Mirra[5] suggested that they represent a congenital rest of synovial tissue which has been displaced into the thin cortical metaphyseal region of bones at the synovial-capsular bone reflection. The marked predominance of cysts at the humerofemoral location can be explained by the fact that these two bones have the largest area of capsular-to-metaphyseal bone reflection. It is not inconceivable that synovial sarcoma may also arise from such intra-osseous synovial rests.

The presence of giant cells within the stroma of the tumour in our patient is probably related to the intra-osseous situation of the tumour; similar giant cells were also present in the synovial sarcoma of the tibia reported by Lederer and Sinclair.[3]

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Severe airway obstruction associated with rheumatoid arthritis and Sjögren's syndrome

A case report

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Summary

Numerous pulmonary complications associated with rheumatoid arthritis (RA) and Sjögren's syndrome (SS) have been described. Mild airway obstruction has recently been recognized in these disorders, severe obstruction occurring in only a few patients with RA. We report a patient with RA and SS who developed severe irreversible airway obstruction, an association not hitherto described. The airway disease failed to respond to bronchodilators, steroids or immunosuppressive agents.

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Numerous pulmonary manifestations of both rheumatoid arthritis (RA) and Sjögren's syndrome (SS) have been described.[1-5] Although airway obstruction in RA has been noted for some time, this complication was originally attributed to the smoking habits of the patients studied.[6-8] Later studies revealed mild airway obstruction in a significant proportion of patients with RA who were non-smokers.[4,9] Recently a severe form of airway obstruction, characterized by obliterative bronchiolitis, has been described in a few patients with RA.[2] In contrast, SS is usually associated with restrictive ventilatory defects,[3] and obstructive airway disease has been reported only in 1 study.[10] We report a patient with seropositive RA and SS associated with severe irreversible airway obstruction. The lung disease has remained refractory to treatment with bronchodilators, steroids and immunosuppressive agents.

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Case report

A 54-year-old White woman presented in October 1979 with symmetrical polyarthritis involving the feet, wrists, proximal interphalangeal joints, shoulders, elbows and knees. A rheumatoid factor test was positive, and she was treated with several non-steroidal anti-inflammatory drugs with poor symptomatic relief. She was referred to the Rheumatology Clinic at the Johannesburg Hospital 1 year later for further assessment. In addition to joint pain she complained of severe grittiness of the eyes and a dry mouth for the previous few months. Although her physical activity had been severely curtailed by the arthritis, a progressive decrease in effort tolerance and a non-productive cough had been present for the past few months. There was no past history of cough, asthma, or exposure to industrial or chemical agents. She was a non-smoker, had never been treated with steroids, penicillamine or gold, and denied any antecedent chest infections.

Examination revealed typical rheumatoid changes in the hands and feet with acute synovitis of the wrists, proximal interphalangeal joints, metacarpophalangeal joints, shoulders, knees, ankles and toes. She was tachypnoeic at rest and generalized fine inspiratory crepitations were audible, most marked at the midzones and bases of both lungs. The rest of the examination was negative. Investigation showed a mild hypochromic microcytic anaemia (haemoglobin concentration