

PAGET'S DISEASE (OSTEITIS DEFORMANS) *

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(Pages 130 to 133)*

Fifteen cases of Paget's disease seen during a period of 14 years are presented. Maximum number of cases were in the sixth decade. The incidence of sarcomatous complication was 26.6 per cent, however, as the series is small it has no statistical significance. It is probable that Paget's disease is prevalent in greater number among Indians than has been thought of earlier.

Paget's disease is a slowly progressive disorder of the skeleton of unknown etiology characterised by abnormal destruction and production of bone. It was first described by Sir James Paget in 1876; but Czerney of Freiberg had already described the condition in 1873 and given the name 'Osteitis deformans'. After the publication of Paget's classical papers an increasing number of cases were reported in the literature especially from the west. There had been only occasional reports of the disease in India (Chakrabarty and Bharadwaj 1963, Shanmugasundaram 1970, Rangachari et al. 1975). This disease is not so rare in our midst though there is paucity of published reports.

MATERIAL

The present article is based on the study of 15 cases of Paget's disease seen at the Department of Orthopaedics, Govt. General Hospital, Madras-3, during 1968-81. The senior author (T.K.S.) has seen nearly 50 patients during the years 1959-81. At times, the diagnosis is incidental. Progressive deformity of the limbs, pathological fracture, malignancy or low back ache are the predominant features which bring them to the hospital.

OBSERVATIONS

The maximum incidence of Paget's disease in this series was between 50 to 59 years, i.e. 7 cases out of 15. The mean age was 56.6 years—the youngest patient being 45 and the oldest 80 years. There were 11 male patients and 4 females.

Of these 15 cases one was mono-ostotic in the left humerus; all others were polyostotic, 4 cases presented with malignant changes (of the 50 cases seen by the senior author only one presented with malignant transformation to fibrosarcoma). Five patients presented with painful swelling in the extremities, 4 presented as low back ache, and another 4 with progressive deformity of the limbs also difficulty in hearing. Three patients presented as fracture following trivial injury. One of the patients presented with pain and progressive limitation of movements of left hip. Radiologically, the skull showed typical changes in 14 and the pelvis in 12 cases. Involvement of the lumbar spine and femur was seen in 5 cases each. Deformity of the legs and X-ray changes in tibia were seen in 6 cases. The histological picture of the malignant lesions were fibrosarcoma in one case and osteosarcoma in the other 3. The case of

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fibrosarcoma was living, at one year after amputation, with secondaries in the lungs. All the cases of osteosarcoma died within one year of presentation.

Increased pulse pressure, high cardiac output and cardiac enlargement were observed in one of the cases where the disease was active. Calcification in the main arteries was observed in 5 patients.

Biochemical findings : The serum calcium, phosphorus and alkaline phosphatase estimations were done in 12 out of the 15 cases. The serum calcium and phosphorus were within normal limits in all the cases. The alkaline phosphatase level was raised in 10 cases and normal in 2 cases. The level of alkaline phosphatase was more than 100 K.A. units in 5 cases, between 50 to 100 units in 4, and between 15 to 50 units in one case.

ILLUSTRATIVE CASES

Case 1 : Narayanasamy, 61 years male, presented with pain and progressive limitation of movements of left hip of 2 years duration. He had a large skull (circumference 62 cm) and was using a hearing aid (Fig. 1). The range of movement in the left hip was flexion only from 70 to 90 degrees. Serum alkaline phosphatase level was 150 K.A. units. Radiograph of the pelvis showed increased density and coarse trabeculae (Fig. 2). There was remarkable increase in the thickness of the skull with invagination of the base of the skull (Fig. 3).

Case 2 : Manickam, 50 years male, presented with painful swelling of right leg of 2 weeks duration (Fig. 4). There was a warm, tender swelling (10 cm × 10 cm) of variable consistency arising from the right tibia. The alkaline phosphatase was 60 K.A. units. Radiographic survey showed sclerosis of upper end of tibia with areas of destruction (Fig. 5) and involvement of pelvis, skull and humerus.

Histopathological examination of the tibial



Fig. 1.
Clinical photograph of case 1, showing the large size of the head, and the hearing aid.



Fig. 2.
Radiograph of the pelvis of case 1, showing increased density, coarse trabeculae and obliteration of joint space of the left hip.

swelling revealed that it was fibrosarcoma complicating Paget's disease of tibia. An above knee

amputation was done followed by chemotherapy with cyclophosphamide and methotrexate. This patient was reviewed recently one year after the amputation and he had pulmonary secondaries. He is the only patient alive today after the malignant change in Paget's disease.

DISCUSSION

Paget's disease is commonly seen in Anglo-saxons, the highest incidence being in England and

Western Europe. The present series differs from the Western literature as regards the incidence of malignancy which was 26.6 per cent (4 out of 15). Lichtenstein (1977) stated that malignancy in Paget's disease was less than 10 per cent while Jaffe (1958) thought it was less than 2 per cent. The incidence of sarcomatous transformation in 4 out of 15 cases seen in a single department in India indicates that large number of undetected cases of Paget's disease must be prevalent in the South Indian population. Many patients remain asymptomatic and hence go undetected. The impression of rarity of Paget's disease among Indians is probably incorrect. Sarcomatous change in Paget's disease is a serious and fatal complication. The mean survival in Paget's sarcoma was 11.1 months in Price's series (1969). In the present series also only one patient survived more than a year from the time malignancy was diagnosed.



Fig. 3.

Radiograph of the skull of case 1, showing remarkable increase in thickness of the skull with invagination of the base of the skull.



Fig. 4.

Clinical photograph of case 2, showing the ulcerated tumour of right tibia.



Fig. 5.

Radiograph of the leg of case 2, showing sclerosis of the upper end of tibia with osteolytic areas.

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