

DEFINITION

1. **Paget's disease of bone**, which is more accurately described as **osteitis deformans**, is a focal or multi-focal chronic bone disease in which there is resorption and regeneration of bone, which causes softening, thickening and deformity together with an increase of abnormal bone mass at each focus. The condition is not a diffuse one of bone.

CLINICAL MANIFESTATIONS

2. The lumbar vertebrae, sacrum, skull and pelvis are most often affected although limb bones may be involved. The disease may be confined to one bone or may be widespread.
3. The disease typically affects the elderly being rarely noted before the age of 50 years. It is frequently asymptomatic being discovered as an incidental finding on X-ray.
4. Deep bone pain, usually worse at night, may be a symptom. The development of deformity, spinal curvature, enlargement of the skull or bowing of the limbs may be observed, pathological fractures may occur and joints adjacent to the effected bone are liable to develop osteoarthritis.

AETIOLOGY

5. The disease is relatively rare in Asia and Africa but quite common in the USA, UK, Australia, France and Germany.
6. Men are affected more than women and, although a number of families with more than one member affected has been reported, the condition is rare in sons of fathers with the disease.
7. Claims have been made for an inborn error of connective tissue metabolism, a disorder of hormone secretion, and acquired vascular disorder or an auto-immune state but supporting evidence for these claims has been lacking.
8. A discovery of nuclear inclusion bodies (resembling those of measles) in the osteoclasts in Paget's disease and in no other metabolic bone disease has suggested a virus infection, possibly a "slow" virus. As yet, definite proof of this concept has not been obtained nor has it been refuted.
9. There is no evidence that the course of the condition is affected by any environmental factor.

CONCLUSION

10. Paget's disease of bone (or Osteitis Deformans) is a focal or multifocal disorder of bone for which no cause has been identified. It is thought by some experts that the condition may be viral in origin. Its course is unaffected by environmental factors.

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