

PAGET'S DISEASE IN AN ANGLO-SAXON

by

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SUMMARY

A RECENTLY excavated skeleton from an Anglo-Saxon burial ground at Jarrow Monastery is described. Virtually all the bones are abnormal, having the morphological and radiological features of Paget's disease. It is one of the most convincing examples in the annals of palaeopathology and confirms the antiquity of this condition.

INTRODUCTION

Osteitis deformans (Paget's disease) was described as a clinical entity less than a century ago¹ but examination of skeletal remains has since revealed that the disease has existed from neolithic times. As early as 1889 Hutchinson² described an affected portion of a parietal bone removed from an ancient Egyptian tomb and, although the specimen cannot now be traced, his description of it leaves little doubt as to the diagnosis: "The bone is much thickened and composed throughout of fine porous tissue. Its surface is rough and marked by fine arborescent grooves and minute apertures. On its inner surface the channels for blood vessels are deepened." Later reports of the disease in ancient man have sometimes been based on inadequate evidence. In this report we describe a case of extensive Paget's disease, in an unusually well-preserved Late Saxon skeleton, which must rate as one of the most convincing examples yet recorded.

DESCRIPTION OF THE SKELETON

Several hundred Late Saxon burials have been excavated from Jarrow Monastery, Durham, during the past decade. One of these (No. 69 WC 16), probably datable to around A.D. 950, is that of a man, aged about sixty-five, and shows some interesting abnormalities. It lacks a little of the facial skeleton, the feet and the distal end of the leg bones but apart from that it is virtually complete and in very good condition, although the long bones and most ribs have suffered a few clean breaks from post-inhumation soil pressure. Of the surviving remains all except a few rib fragments and hand bones are extensively diseased. The following is a brief account of the major changes.

Almost the entire surface of every bone is rough and irregularly pitted or "grained", whilst most are thickened and distorted. The skull is abnormally large and thick (Fig. 1). Its maximum length is 205.4mm., its breadth 146.8mm., giving a Cranial

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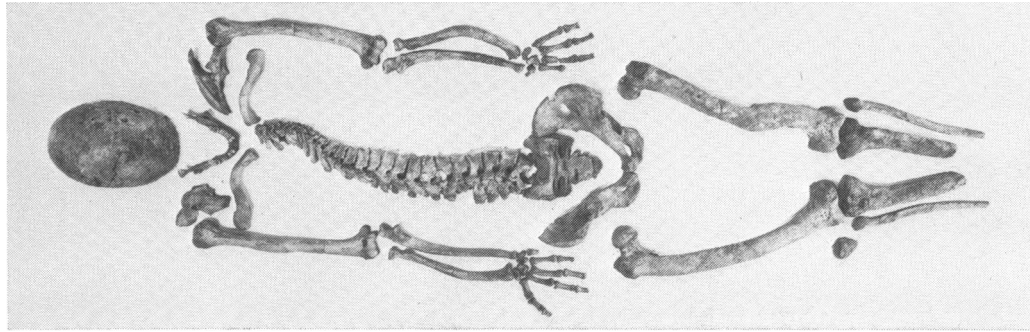


Figure 1.

Osteitis deformans of a Late Saxon (c. ninth to tenth century) skeleton from Jarrow. General view.

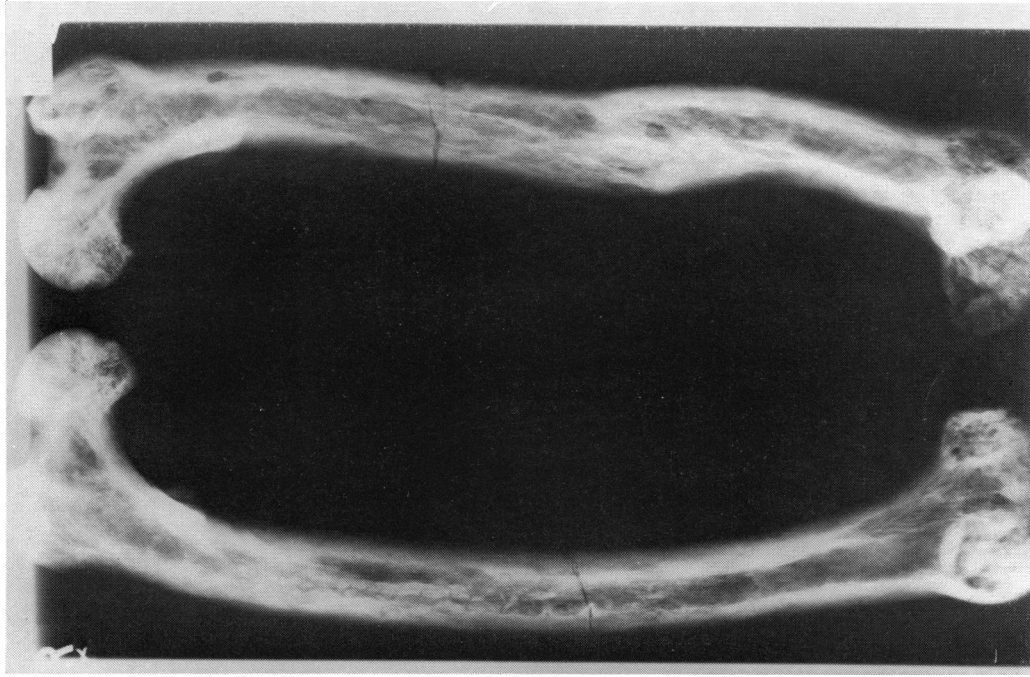


Figure 2.

Radiograph of femora with "bizarre" bone, thickened cortex and a L. mid-shaft fracture.

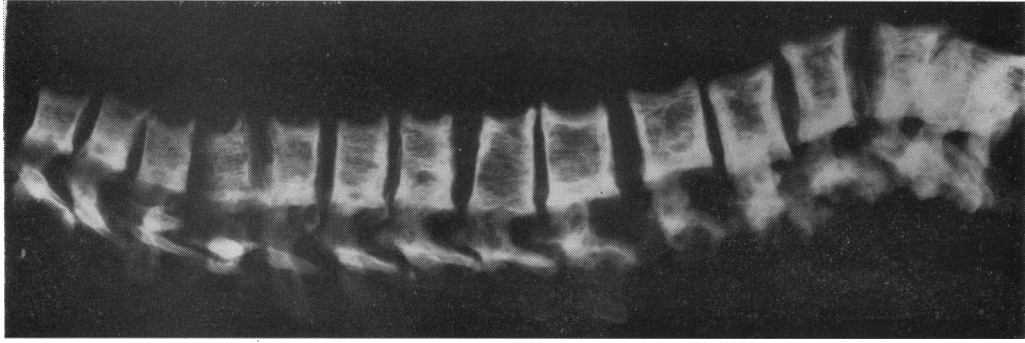


Figure 3.

Radiograph of vertebral segments T4-L5 to show "window frame" appearance and osteophytosis.

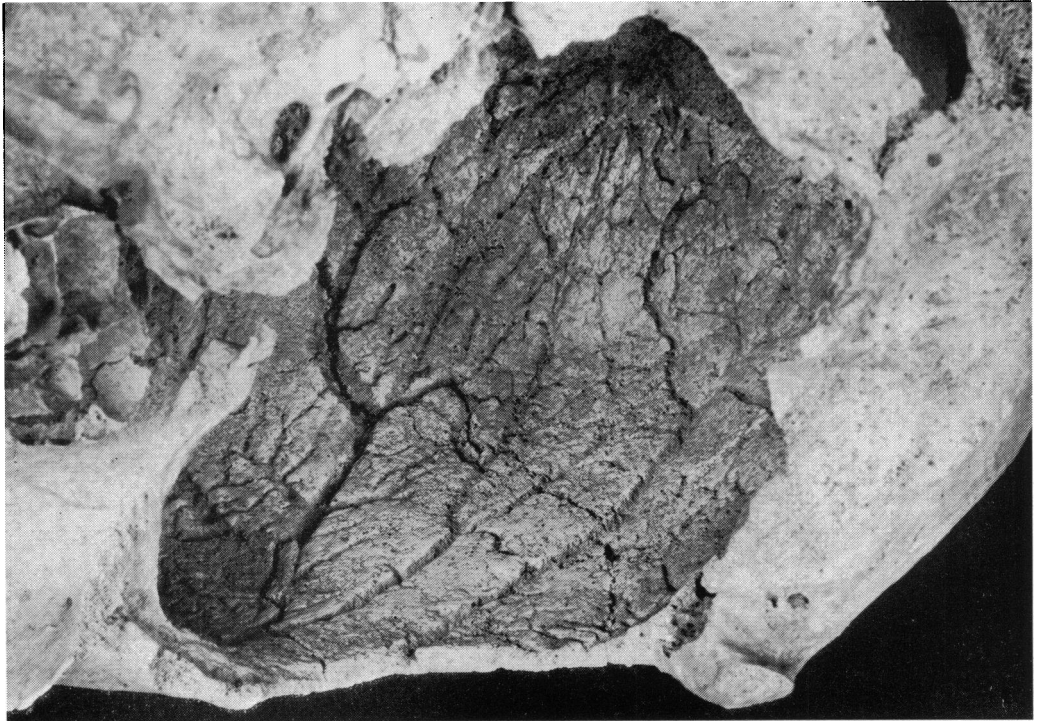


Figure 4.
Endocranial view showing huge channels for meningeal vessels.

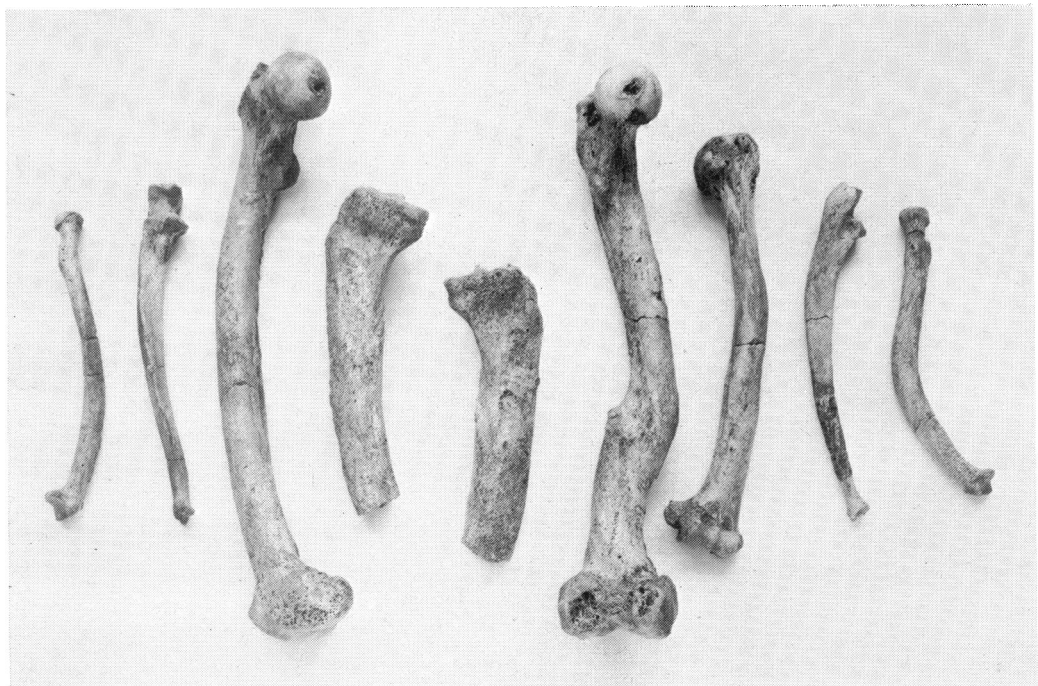


Figure 5.
The long bones showing curvature and torsion.

Index of 71.4 (dolichocranial). Its circumference through the optryon and opisthocranium is 573.8mm. The skull vault reaches 14mm. at the thickest part of the parietals and 22mm. at the internal occipital protuberance. Its surface shows light erosion but the underlying bone is finely granular or foam-like in texture. Endocranially the meningeal blood vessels have left deep channels in the bone (Fig. 4). The mandible is affected in the same way as the cranial vault whilst cementosis is extensive on some of the surviving teeth. The rest of the facial bones are damaged and incomplete but much of the maxillae survives and was also involved.

The vertebrae are similarly granular and irregular. In the thoracic region their bodies are slightly wedge shaped vertically and anteriorly so that, in life, this man must have had a well-marked kyphosis. Additional mid-thoracic asymmetry would also have left him scoliotic whilst, individually, several of the vertebrae have slightly "subsided" to give a splayed out effect, with the body broad in proportion to its height. Osteophytosis is present on most vertebrae and is severe on at least eight. The innominate show a "cupped" appearance of the bodies of the ilia which is due to a combination of lateral flaring in the anterior half of the crest and a pinching in of the bone immediately above the acetabulum. The inferior rami of the pubes and ischia also flare forwards and laterally so that the obturator foramina have come to resemble holes cut through the bottom of a shallow dish. When the left iliac crest is viewed from above its normal S-curve is greatly exaggerated. These innominate bones seem to have been nearly symmetrical but the posterior part of the right iliac crest is now slightly damaged and leaves a little uncertainty about this. When the pelvis is reconstructed by articulating its three components it somewhat resembles the triradiate pelvis of osteomalacia and this has undoubtedly arisen as the result of an abnormal softening of the bones. Both clavicles are also much thickened though asymmetrically, with an increased curvature. At least twenty of the ribs seem to be abnormal. This abnormality consists, as with the rest of the skeleton, of surface roughening, marked thickening and distortion or irregular curvature, with partial obliteration of the costal groove and of the marrow cavity. The scapulae are similarly affected.

Both humeri are bowed, the left more than the right (Figs. 1 and 5). When the left humerus is held so that the proximal part of its shaft is vertical, the transverse axis through the epicondyles deviates about 40° upwards and medially. It also has about 35° of antero-medial torsion. The humeral heads are surrounded by a low flange of osteoarthritic lipping, in addition to the rough and thickened state of the adjacent shaft.

The ulnae and, more especially, the radii are grossly distorted (Fig. 5). The right ulna has a shallow S-curve and both radii are strongly bowed radially, with nearly 70° of longitudinal torsion. In all these bones the medullary cavity is greatly diminished or obliterated.

The right femur is slightly bowed anteriorly, more so laterally (Figs. 1 and 5). Its shaft has about 25° of torsion on its long axis and its short, angled neck leaves the trochanteric-condylar length of the bone, at 419.3mm., 12.1mm. longer than its capito-condylar length. The left femur has a similar curvature, anteriorly and laterally, but this is partly distorted by a well-repaired mid-shaft fracture (Fig. 5).

Short Articles

This fracture was almost transverse but slight overlap of the two fragments has occurred and left the bone 10mm. shorter than its fellow. Wedges of callus bevel the overlap between the two fragments. There is also much torsion of this femur—at least 50°. In spite of the additional deformity to which it gave rise the fracture has healed well and the callus is nowhere excessive. It is regrettable that only the proximal two-thirds of the tibiae and three-quarters of the fibulae have survived, because these bones are grossly deformed. The tibial bowing is almost entirely anterior, that of the fibulae was more sinuous (Figs. 1 and 5).

DISCUSSION

This case is not one which calls for discussion or elaborate differential diagnosis: it is a classic or “textbook” example of Paget’s disease (osteitis deformans). The age of this man, together with the bowing and proliferation of the bones, are all highly characteristic of this condition, whilst numerous special features combine to establish the diagnosis beyond doubt. The great depth and size of the meningeal vascular grooves are almost pathognomonic and are a response to the enormously increased blood flow, through these vessels. The cranial thickening is also typical and so, too, is the hypercementosis of the teeth. In Paget’s disease the deformity of the clavicles and long bones is nearly always asymmetrical, as here, and is commonly associated with marked thickening of the cortex and extensive obliteration of the medullary cavity such as most of these bones reveal. This deformity is due to the extreme softness of Paget’s bone and its ready collapse as a result of weight-bearing in the case of the vertebrae, pelvis and lower limbs. The gross distortion of the non-weight-bearing upper limbs seems largely to be brought about by the spontaneous yielding of the bone to forces of muscular compression. Another outstanding feature of osteitis deformans, which this skeleton shows, is a femoral fracture. When this happens it is usually, as in the present specimen, a transverse break not an oblique one. If further proof is sought, it can be found in a radiograph of any of these bones, when the pathognomonic “bizarre” appearance is unmistakable (Fig. 2). Another special radiological feature here is the extreme thickness of the cortex which has largely obliterated the medullary cavities. This typically involves the metacarpals and phalanges (even when they are not greatly deformed) as well as the major long bones. The vertebral radiograph, alone, is almost sufficient to establish the diagnosis. It shows the “window frame” feature (Fig. 3) which is due to rarefaction of the trabeculae within the vertebral bodies and increased density of their surrounding bone.

Paget’s disease has often been diagnosed in prehistoric skeletons but perhaps the first reasonably convincing case was published by Pales.³ He reported a solitary neolithic R. femur from Lozère, which bore a close resemblance to the R. femur described above. Pales believed that his specimen was “*le seul cas reconnu et indiscutable de maladie de Paget préhistorique*”. In asserting this he ignored, with good reason, earlier contributions by Baudouin^{4,5} who had diagnosed the disease on somewhat slender grounds. Indeed, one of the most obtrusive features of the literature is the repeated diagnosis of osteitis deformans on the most threadbare evidence, sometimes from small scraps of bone which have been eroded or deformed

Short Articles

by post-inhumation processes. Thus, Fisher⁶ diagnosed it in an exceedingly fragmentary skeleton from Lynxville, Wisconsin, but Morse⁷ reports that a recent thorough examination of the specimen revealed no evidence to support this diagnosis. Denninger⁸ described five unevenly convincing cases from Illinois, all of which were either fragmentary, eroded or otherwise defective and cannot be accepted without much reservation.

Cautious workers have sometimes indicated the problematic nature of these cases by offering a range of alternative diagnoses, often constrained to do so by the inadequate amount of the skeletons to survive.^{9,10} Paget's disease becomes much easier to recognize if its typical microscopical "mosaic" pattern of bone can be detected. But as Putschar¹¹ has pointed out this may be difficult in archaic material since "the frail trabeculations of woven bone . . . are . . . often not sturdy enough to survive long burial" and Johnson¹² comments: "the pumice bone of Paget's disease will not remain, and much of the mosaic bone may have disappeared". Despite its macro-perfection the Jarrow specimen has proved to be just such a case: no good micro-section has been obtainable. Jaffe¹³ especially refers to the problem of distinguishing between syphilis and osteitis deformans when the diagnosis is based on nothing more than a solitary, incomplete bowed and thickened tibia.

Despite these difficulties a number of firm and probably accurate identifications of the disease have been made from widely separated localities. Møller-Christensen¹⁴ proposed one in a twenty-five-year-old man—an unusually young case—from Aebelholt. Heuertz¹⁵ recorded one from Ennery (Moselle). Rokhlin¹⁶ notes a somewhat less certain one from Russia. Milanese¹⁷ observed the condition in a neolithic humerus from Ponte S. Pietro. Many others have been suggested. But a perusal of the literature strongly suggests that many, perhaps most, reported cases of ancient osteitis deformans carry little conviction and that this is chiefly due to the fragmentary state of the specimens on which the diagnosis is made combined with uncritical enthusiasm in the authors. Very well-preserved and lacking only some fragments of face, feet and ankles, the Jarrow Monastery skeleton cannot be faulted as unduly defective. It takes its place, therefore, as one of the most convincing examples in the annals of palaeopathology and certainly the finest, perhaps the only, example of Paget's disease to emerge from an Anglo-Saxon burial ground.

ACKNOWLEDGEMENT

Our thanks are due to the excavator, Professor Rosemary Cramp, for inviting us to publish this case.

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Obituary

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OBITUARY

DOUGLAS JAMES GUTHRIE, M.D., D.LITT., F.R.C.S.E., F.R.C.P.E., F.R.S.E.
8 September 1885 – 8 June 1975

Douglas Guthrie was within three months of his ninetieth birthday when he died in Edinburgh on 8 June after a long illness. His name is well known to the present generation as the author of a successful and popular *History of medicine*, but when he retired in 1945 at the age of sixty he had already made significant contributions to his own specialty—the study and treatment of disorders of the ear, nose and throat—and the first edition of his *History* was already in the bookshops. His object in writing it had been to interest medical students and young doctors, but it achieved a far wider success with the general reader and so gave him the stimulus and the opportunity to develop his second career.

He was born at Dysart, Fife, where his father, the Reverend William Guthrie, was a Minister of the United Free Church, and educated at Kirkcaldy High School and the Royal High School, Edinburgh. He graduated with honours in the Faculty of Medicine at Edinburgh in 1907 and was awarded the McCosh Travelling Scholarship which enabled him to continue his studies in Berlin, Hamburg, Jena, Vienna and Paris, where he served as a clinical assistant in the Hôpital Louis. While building up a general practice at Lanark he proceeded to his M.D. in 1909 and the Fellowship of the Royal College of Surgeons of Edinburgh in 1913. During the first world war he served for two years with the R.A.M.C. and was then appointed commandant of a hospital for officers of the Royal Flying Corps. At the end of the war he decided to specialize in laryngology and otology and became a lecturer in the "extra-mural" school of the Royal Colleges at Edinburgh as well as Ear and Throat Surgeon to the