Paget Disease of Bone
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After completing this article, the reader should be able to:

- Discuss the etiology of Paget disease of bone.
- Identify the clinical characteristics of the disease.
- Recognize the radiological manifestations of Paget disease of bone.
- Describe the treatment for Paget disease of bone.

On January 11, 1814, a man was born who would influence the science of medicine like Leonardo da Vinci influenced Renaissance art. This man, Sir James Paget, is considered to be one of the founders of modern pathology and one of the surgical “greats” of 19th century England (see Figure 1). Paget began his apprenticeship in medicine at the age of 16. At the age of 20, he began his medical studies at St. Bartholomew’s Hospital in London. Paget had not even graduated from medical school when he made his first important medical discovery: the pathogen for trichinosis. Trichinosis is a parasitic disease caused by miniscule roundworms that infest the muscles of the human body and usually is contracted by eating infected pork. When describing how he discovered this pathogen, *Trichina spiralis*, he said, “All the men in the dissecting rooms, teachers included, ‘saw’ the little white specks in the muscles, but I believe I alone ‘looked at’ them and ‘observed’ them.”

Paget wrote several important works, including *Lectures on Tumors* and *Lectures on Surgical Pathology*. He received many rewards and achieved knighthood in 1871. He also was the first to describe osteochondritis dessicans and the first to recognize that the median nerve could be compressed at the wrist, which was confirmed in the early 20th century. Additionally, Paget was the first to describe several nonrheumatologic conditions, including Paget disease of the nipple, extramammary Paget disease, maxillary vein thrombosis and recurrent abscesses (Paget residual abscesses).1

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However, he is best known for his description of osteitis deformans, or Paget disease of bone, the clinical symptoms of which include enlarged bones, progressive deafness, bowed legs and chronically inflamed bone (see Figure 2).³

Paget disease of bone is a localized disorder of bone remodeling, with both active and inactive phases, that results in large and deformed bones in various parts of the skeleton.⁴ Paget first described the disease in 1877 in reference to a small group of patients. These patients were described as having overly large heads and enlarged or deformed extremities with a higher likelihood of fracture.⁵ Dr. Paget believed it was a new disease; however, today abundant evidence indicates that it was not a new development in 1877. This has been confirmed by histologic and radiographic studies of ancient skeletal remains.¹

Skeletons unearthed in medieval burial grounds in England showed radiographic and histopathologic evidence of Paget disease.⁴ Skeletons unearthed in a cemetery in Torre de Palma, Portugal, a Roman villa established in the late 1st century AD, also showed signs of Paget disease. The report of this exhumation and research, presented in 2002 to the Paleopathology Association by Drs Cook and Powell stated, “This adult male skeleton showed extensive new bone formation on the medial and lateral surfaces of the right ilium. There is no remaining normal bone cortex, and the underlying trabecular bone is coarsened. There is fine, new nodular bone on the recovered acetabulum. Radiographic and histologic analysis helps to differentiate between Paget disease of bone, fibrous dysplasia, and metastatic carcinoma in this interesting specimen.”⁵ It even has been suggested by some that the painting of a grotesque old woman with an enlarged head, hanging in the National Gallery in London, indicates Paget disease of the skull.¹

**Etiology**

The exact cause of Paget disease remains uncertain.⁶ However, genetic factors certainly are involved, and viral factors are suspected.⁷ Research from several different areas of investigation provides useful hypotheses. One hypothesis, although still controversial, proposes that changes in bone remodeling are caused by a viral infection in the bone. These viruses include the measles virus, the canine distemper virus and the respiratory syncytial virus.³

A case-control study with dog owners indicated the risk of Paget disease was nearly 3 times higher in owners of unvaccinated dogs compared with those who owned dogs vaccinated against the distemper virus.⁸ Additionally, a study of 13 patients with Paget disease found that the osteoclasts and peripheral blood mononuclear cells from 9 of these patients expressed measles virus transcripts, but none of the 10 control patients expressed any measles virus at all. In 1 patient, a novel mutation that converted lysine to glutamic acid in the measles virus transcript was identified.⁹

No theory about the cause of Paget disease has been proven definitively, and all remain controversial; however, studies suggest that Paget disease could result from a virus in the osteoclasts of genetically susceptible people. The belief is that the viral infection lies dormant.