

Paget's disease

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Abstract

Sir James Paget's seminal description of the clinical findings of the bone disease that now bears his name in 1877 holds true today, but his reference to the disease as *osteitis deformans*, implying an inflammation of the bone, is not accurate, and it is now called *osteodystrophia deformans*.

Keywords

Paget's disease.

Introduction

Sir James Paget's seminal description of the clinical findings of the bone disease that now bears his name in 1877 holds true today, but his reference to the disease as *osteitis deformans*^[1], implying an inflammation of the bone, is not accurate, and it is now called *osteodystrophia deformans*.

Paget's disease

Paget's disease of bone (PDB) is now believed to result from an abnormality of bone remodelling due to an increase in osteoclastic activity. Its aetiology remains controversial. A slow-virus infection of the osteoclasts has been suggested but not proven. The lack of an animal model for PDB makes it difficult to apply Koch's postulate. Genetic susceptibility may be necessary for the development of PDB, because between 15 and 40% of individuals have a positive family history of the condition^[2].

Most patients with PDB are asymptomatic. The diagnosis is usually made incidentally by finding an elevated serum alkaline phosphatase level on a routine blood test or by plain radiograph requested for another indication. When symptomatic, the two main features of PDB are pain and deformity in the affected area of the body. Sir James Paget correctly observed that the disease 'begins in middle age or later... affects most frequently the long bones of the lower extremities and the skull'. He also accurately described the clinical features: 'The bones enlarge and soften, and those bearing weight yield and become unnaturally curved and misshapen'. The increase in skull size is carefully documented: 'The skull became gradually larger... his hat and the helmet he wore... needed to be enlarged. In 1844, he wore a shako measuring twenty two and a half inches inside; in 1876 his hat measured twenty seven and a quarter inches inside'.

Patients with PDB have a significantly increased risk of developing osteoarthritis^[3]; '... when the disease had existed sixteen years, the left knee joint was, for a time, actively inflamed and its cavity

was distended with fluid. But the inflammation soon subsided, only leaving the joint stiffer and more bent'.

Diagnosis and treatment

The diagnosis of PDB is made on finding an elevated serum alkaline phosphatase level with radiological features of abnormal bone remodelling (osteosclerosis, osteolysis and bone expansion). Radionuclide bone scanning is useful in evaluating skeletal involvement. Paget's description preceded the development of these tests by many years.

Paget 'advised iodide of potassium and Liquor Potassae; but they did no good'. The introduction of bisphosphonates in 1973 provided effective treatment for PDB, reducing pain and bone turnover^[4], but there is currently no evidence that intensive treatment with bisphosphonates can help prevent long-term complications of the disease^[1].

Paget's original description of the disease, as published, did not contain any illustrations, probably for technical reasons. However, there exists, in the pathology museum at St Bartholomew's Hospital, a photograph of Paget's first case, which is linked to the publication by the addendum of the correct reference. The photograph has been exhibited in broad daylight for many years and is in excellent condition, with no evidence of fading. This photograph is now published together with the original case report in this electronic journal and we believe that this is the first time that the photographs and the case report have been published together.

References

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