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CONTINUING PROFESSIONAL EDUCATION

Paget's: The neglected bone disease

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Summary After osteoporosis, Paget's disease is the next most common metabolic bone disease in the UK where it occurs in approximately 2% of adults over the age of 55, rising to 8% of men and 5% of women by the eighth decade of life. Patients with this disease are managed in varied clinical settings including orthopaedics with some complaining about lack of awareness amongst health professionals, with delays in diagnosis and starting appropriate treatment being the major problems. Paget's disease which is characterised by focal increases in bone turnover affects one or multiple bones in the skeleton and occurs most commonly in the skull, spine, hip and pelvis. Whilst many people have few symptoms in others it can result in pain at the affected site, marked bone deformity, deafness, neurological symptoms, pathological fractures and may be associated with an increased risk of developing osteoarthritis. Both viral and genetic factors have been implicated in the causation of Paget's disease with most recent research tending to favour genetic causation. At present Paget's disease cannot be cured, but early recognition and treatment with either oral or intravenous bisphosphonates will help to control its progress and minimise complications. On occasions however a small number of patients will require surgery, commonly for joint replacement, fracture fixation and correction of bone deformities.

Editor's comments
This article explores a number of very relevant issues related to Paget's disease and aims to inform front line staff about the many facets of this condition. Including epidemiology, diagnosis and treatment this paper is sure to update and inform. BS

Introduction

In 1877 Sir James Paget, an eminent British surgeon, described ''osteitis deformans'' in an elderly man with progressive skeletal deformities (Paget, 1877).

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''. . . I think we may believe that we have to do with a disease of bones of which the following are the most frequent characteristics:- It begins in middle age or later, is very slow in progress. May continue for many years without influence on the general health and may give no other trouble than those which are due to the changes of shape, size and direction of the diseased bones''.

This condition which subsequently became known as Paget's disease of bone is characterised by rapid bone remodelling and the formation of bone that is structurally abnormal. Although many people remain symptom free it can result in pain, deformity, increased risk of fracture and other complications including osteoarthritis, hearing problems and nerve compression syndromes (see Tables 1 and 2). Currently this disease cannot be cured but there are effective treatments that can control its progression and minimise complications. Whilst Paget's disease is a common disorder, initial diagnosis is often missed and many healthcare professionals including nurses fail to recognise early symptoms and are uncertain about ongoing clinical management. Patients with this condition present in both primary and secondary care where they are managed in varied disciplines including care of the elderly, rheumatology and orthopaedics. It is, therefore, important that nurses working in these diverse specialities have an understanding about the symptoms, consequences and treatment of this condition.

- Common disease of the skeleton.
- Initial diagnosis often missed.
- Lack of awareness amongst healthcare professionals about clinical management.
- Patients present to various specialities in primary and secondary care.

Epidemiology

Paget's disease occurs in 1—2% of white adults over the age of 50 years with it being more common in males. The prevalence increases substantially with age and by the eighth decade of life it has been suggested that it may be present in approximately 8% of men and 5% of women (van Staa et al., 2002). There is marked ethnic and geographical clustering of the disease with it being common in some parts of the world but relatively rare in others. Clinical observations indicate that it presents frequently in Europe, North America, Australia and New Zealand but is rarely seen in Scandinavia, the Indian subcontinent, China, Japan and other countries in South East Asia. The UK has the greatest prevalence of Paget's disease in the world (Detheridge et al., 1982) with a large British survey, performed 20 years ago showing a marked geographical variation in occurrence with higher rates clustering in six Lancashire towns (Barker et al., 1980). Over the past 25 years the prevalence and severity of disease has reduced substantially in the UK and New Zealand, remained fairly stable in the USA with an increase in severity of the condition being noted in southern Italy (Poor et al., 2006). The reasons for these recent changes are poorly understood but possible explanations include modifications of environmental triggers for the disease in some countries and not others or changes in ethnic makeup of the population in some countries due to an influx of migrants from low prevalence regions such as the Indian subcontinent.

- Occurs in 1—2% of white adults, aged over 50 and becomes more common with advancing age.
- Marked ethnic and geographical clustering on a global basis.
- Highest European prevalence in Great Britain.
- Over the past 25 years marked decrease in disease in Great Britain and New Zealand.

Pathophysiology

Bone is a living dynamic tissue that is constantly being renewed through a remodelling process that provides a mechanism for self-repair and adaptation to stress. This takes place at discrete sites on the bone surface and is largely regulated by two types of bone cells, osteoclasts and osteoblasts. Osteoclasts degrade bone by attaching to bone surface and secreting acids and enzymes into the bone cavity with the osteoblasts then synthesising and mineralising new bone matrix within the cavity created. In Paget's disease the osteoclasts are increased in size and number and contain many more nuclei than is normal causing increased osteoclastic bone resorption. This leads to a rapid rate of bone turnover resulting in the production of bone that is larger than normal but with disorganised architecture and reduced mechanical strength, leaving people at greater risk of developing fractures and deformities.

- Osteoclasts increased in size and number.
- Rapid rate of bone turnover.
- Larger bone but with a disorganised structure and lack of mechanical strength.