THE SECOND CASE OF PAGET'S DISEASE (OSTEITIS DEFORMANS) IN A CHINESE LADY

K M H Yip, Y L Lee, S M Kumta, J Lin

ABSTRACT

Paget's disease of the bone is extremely rare in the Chinese population. We report a case of a 91-year-old female patient who presented with a leg ulcer after an injury. On examination, she was incidentally noted to have bowing of the tibia and facial features suggestive of Paget's disease which was later confirmed with plain radiograph. An extensive search of the international literature over the last thirty years, including the use of Medline, has revealed only one other case report of Paget's disease in a Chinese patient in Thailand.

Keywords: Chinese, Paget's disease, skull

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INTRODUCTION

Paget's disease of the bone is a disease of unknown aetiology characterized by osteoclastic resorption of bone followed by osteoblastic regeneration of primitive woven bone. It is more prevalent in those of Anglo-Saxon origin. Schmorl reported the incidence to be 3% in those over the age of 40[1,2]. It is generally perceived to be rare in the Chinese population[3], and this is confirmed after an extensive search of the literature, which has revealed only one other reported case in a Chinese patient in Thailand[4].

We report a case of a Chinese female patient who presented with Paget's disease after trauma and was noted incidentally to have bowing of the tibia with facial features suggestive of Paget's disease. Her diagnosis was confirmed later with pathognomonic features on plain radiography.

CASE REPORT

A 91-year-old Chinese lady presented to our hospital with an ulcer on her left leg which was the result of an injury. Apart from the site of injury, she was not suffering from any pain in the rest of her body. Past medical history was unremarkable in that she is a pure Chinese with no mixed marriage in the family history. During her hospital admission, it was noticed that she had facial features compatible with Paget's disease. These included marked enlargement of the skull, a broad forehead and a wide jaw (Fig 1). She was almost completely deaf. General physical examination revealed kyphosis but no bowing of legs. There were no other neurological signs present and she was not in cardiac failure.

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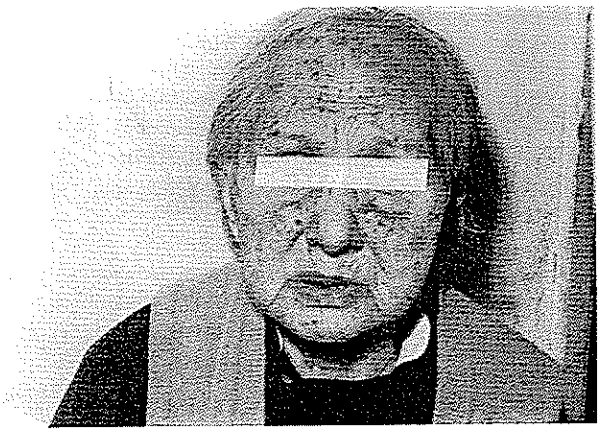
Fig 1 – Anterior view illustrating the features of Paget's disease which include a broad forehead and a wide jaw.



Plain radiography of her skull showed features of classical Paget's disease (Fig 2 and 3), with wool or cotton ball spots and loss of distinction between the inner and outer cortical tables. There was marked cortical thickening with thick coarse trabeculae.

Blood investigations revealed a raised alkaline phosphatase of 384 IU/L (normal = 45-145 IU/L), albumin adjusted calcium of 2.27 mmol/L, phosphate of 1.4 and an ESR of 23 in the first hour. Bone scan and biopsy were not performed as they were not indicated in this case. She was 91 years of age and was asymptomatic apart from her deafness. The diagnosis of Paget's of the skull was made based on the clinical, blood biochemistry and plain radiography.

DISCUSSION

There are marked racial and geographic variations seen in Paget's disease. It occurs much more frequently in the populations of Europe, North America, Australia and New Zealand. The highest prevalence rates are in England (4.6%) and France (2.4%). Its prevalence is low in Scandinavia (0.3% in Norway and Sweden). It is distinctly rare in Asia, particularly China, India and Malaysia[5].

Based on Caucasian data, Paget's disease occurs predominantly in the elderly, and its incidence increases with age. Men are more likely than women to contract the disease.

Clinically, many patients are detected incidentally, as in our case. Many patients present because of trauma, and their radiographs are then noted to have features characteristic of Paget's. Of those who do present with symptoms, these are usually bone pain, changes in skin temperature, pathological

Fig 2 – Lateral view of the patient with Paget's disease. Note the marked enlargement of the skull.

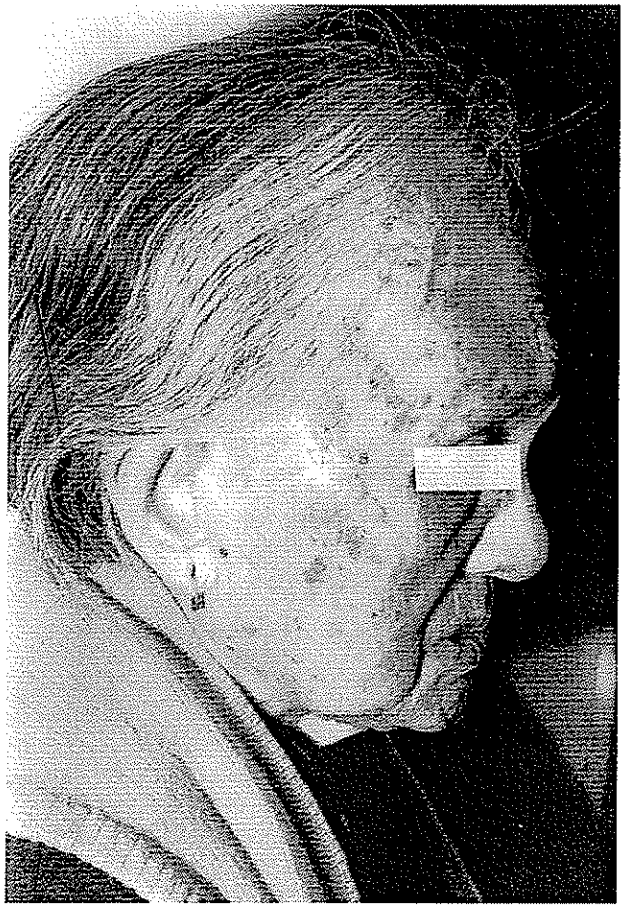
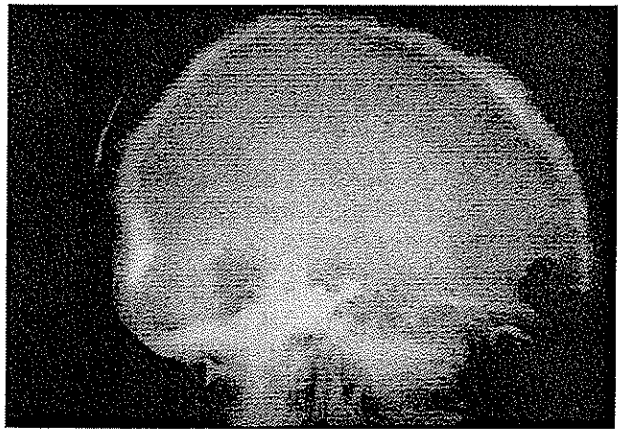


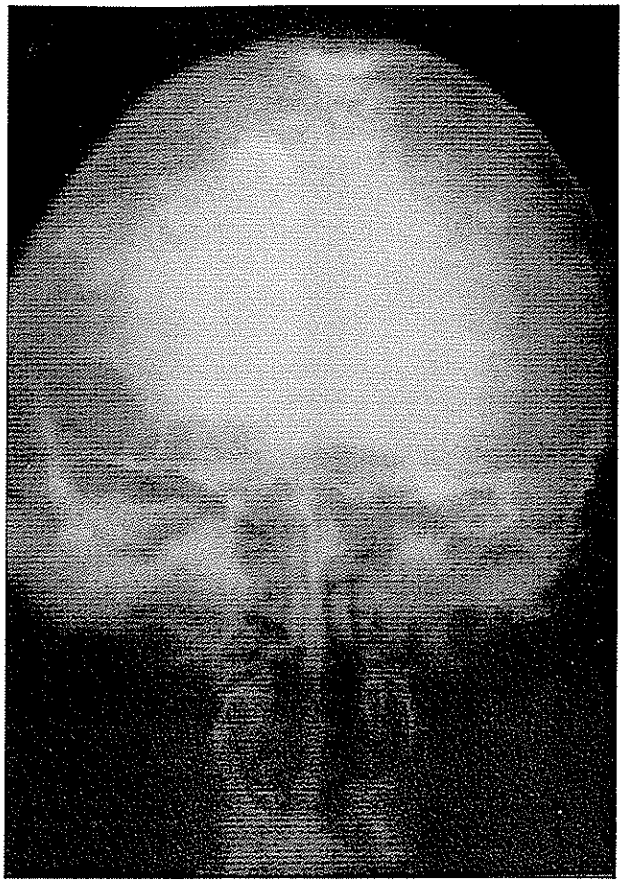
Fig 3 – AP view of the plain radiograph of the skull demonstrating features of classical Paget's disease, with wool or cotton ball spots and loss of distinction between the inner and outer cortical tables.



fractures, skeletal deformities, and symptoms related to nerve compression. The most common of these is bone pain, which is probably due to several causes. One theory is that recurrent microfractures frequently occur causing persistent pain, and another is that the skeletal deformities, such as bowing, leads to abnormal gait and therefore abnormal mechanical stresses. Another more uncommon clinical manifestation is high output congestive cardiac failure, which can occur in patients who have disseminated and active Paget's disease[6].

Diagnosis of Paget's disease is based on the clinical findings,

Fig 4 – Lateral view of the plain radiograph of the skull of a Paget's disease patient illustrating the thickened cortex with thick coarse trabeculae.



with radiological and biochemical investigations. The radiological manifestations of Paget's disease is often pathognomonic. The impression is one of disordered bone resorption and formation. Overall bone size is enlarged, with thickened cortices and coarse and irregular trabeculae. In the latter stages of the disease, the bone becomes more sclerotic and enlarged. Deformities such as bowing of the tibia occur. Incomplete pseudofractures and frank pathological fractures may occur. There may be secondary arthritic changes in joints adjacent to pagetic bone.

The technetium bone scan is important in the diagnosis and management of Paget's disease[7]. It is less specific than plain radiography for diagnosis, but is more sensitive in that up to 30% of lesions noted on scintigraphy cannot be recognised on plain radiographs[8-10]. This makes bone scanning ideal for monitoring and evaluating the extent of disease.

Most routine blood investigations are normal in Paget's disease. The only consistent abnormal finding is usually a raised alkaline phosphatase. It has relatively low specificity and sensitivity in the diagnosis of Paget's disease[7], as the contribution of one bone to the total amount of alkaline phosphatase is small. However, it is of proven value in monitoring disease activity and the response to treatment[11].

The behaviour of Paget's disease is unknown in the Chinese population. The clinical presentation in our patient was classical, however it cannot be concluded that the prognosis and eventual clinical outcomes of the Chinese or Asian patient with Paget's disease will be the same as in the Caucasian. The incidence of Paget's disease in the Chinese population is probably underreported as it is a relatively rare entity and a high index of suspicion is essential in order to make a diagnosis of the disease.

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