Paget's Disease of Bone in a Patient on Hemodialysis

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Paget's disease is a disorder of bone remodeling. Its occurrence in hemodialysis patients is very rare. Here, we report a case of Paget's disease in a 77-year-old patient on hemodialysis who presented with elevated serum level of alkaline phosphatase without any clinical or laboratory findings of secondary hyperparathyroidism. To our best knowledge, this is the first reported case of Paget's disease in a patient with end-stage renal disease from Iran.

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INTRODUCTION

Paget's disease of bone is a chronic affliction of the adult skeleton featuring one or more areas of aggressive osteoclast-mediated bone resorption preceding imperfect osteoblast-mediated bone repair.[1] The resultant deranged skeletal remodeling causes bone expansion and softening, sometimes with pain, fracture, or deformity, and rarely, with neoplastic transformation.[1] The occurrence of Paget's disease in patients receiving maintenance hemodialysis is a very rare entity, and so far, there are 2 reported cases of this condition.[2,3] Here, we describe a patient on hemodialysis that was presented with a highly elevated serum level of alkaline phosphatase (AP) and finally diagnosed as a case of Paget's disease of the bone. To our knowledge, this is the first reported case of Paget's disease in a patient with end-stage renal disease from Iran.

CASE REPORT

A 77-year-old woman on hemodialysis for 2 years was referred to our nephrology clinic with an elevated level of AP in serum, while she did not have any clinical or laboratory findings of secondary hyperparathyroidism. Her chronic kidney failure was secondary to hypertensive nephroangiosclerosis diagnosed in 2003 and long-term hemodialysis was started in 2004. On physical examination, the patient had no organ-specific symptom except for mild generalized bone pain. Complete blood count showed hemoglobin of 11.4 g/dL, hematocrit of 35%, white blood cell count of 3.8 × 10^9/L, and platelet count of 153 × 10^9/L. Serum biochemistry results were as follows: creatinine, 6.9 mg/dL; blood urea nitrogen, 77 mg/dL; calcium, 9.4 mg/dL; phosphorous, 3.7 mg/dL; total AP, 6336 U/L; heat-label AP (bone fraction), 5675 U/L; alanine aminotransferase, 10 U/L; aspartate aminotransferase, 16 U/L; lactate dehydrogenase, 561 U/L; creatine phosphokinase, 122 U/L; and intact parathyroid hormone, 45.1 pg/mL. Serum and urine protein electrophoresis did not reveal any abnormality. Abdominal ultrasonography

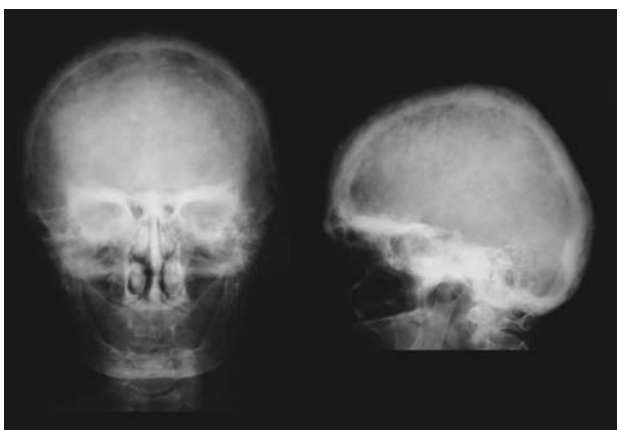

Figure 1. Multiple hazy sclerotic areas are seen in the skull of the patient. This is the typical "cotton wool" appearance of the sclerotic phase of Paget's disease in the skull.

Case Report

demonstrated bilateral small-sized kidneys and normal liver and biliary tract. Radiographic evaluation showed multiple hazy sclerotic areas seen in the calvaria (Figure 1). Whole-body bone scan by technetium Tc 99m methylene diphosphonate showed diffusely intense uptake in the calvaria (Figure 2). The patient's skull radiography taken 2 years earlier showed the same changes with a milder intensity (Figure 3). At presentation, she was receiving oral calcitriol, calcium carbonate, folic acid, and atenolol.

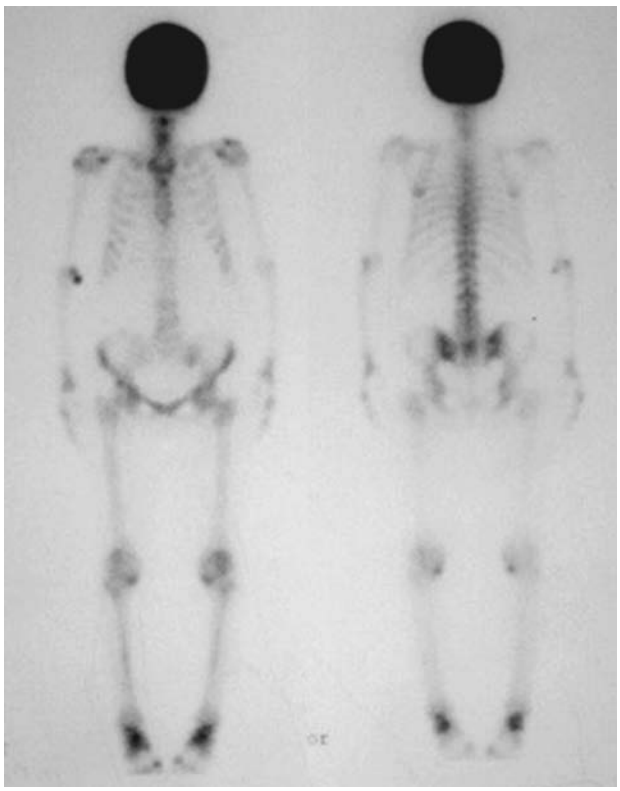

Figure 2. Whole-body bone scan showed diffusely intense uptake in the calvaria (hot spot), which is most likely suggestive of Paget's disease.

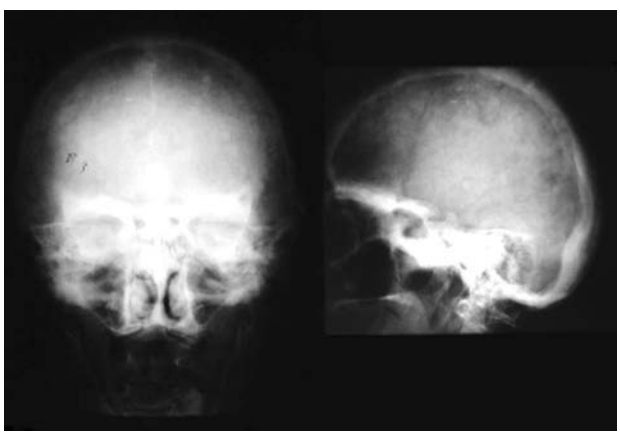

Figure 3. Skull radiography of the patient taken 2 years earlier was suggestive of the Paget's disease in its initial phase.

Based on the clinical and laboratory profiles, a diagnosis of Paget's disease was made, and alendronate at a dosage of 70 mg/w was started on. Six months later, she was free of symptoms and the total AP was 600 U/L.

DISCUSSION

In our patient, a high serum level of the AP bone fraction suggested a persistent high turnover bone remodeling disease, while the intact parathyroid hormone was within its normal range. Bone diseases involving high-turnover bone remodeling include: hyperparathyroidism, bone metastases of malignancies, and the Paget's disease.[2] It is sometimes difficult to differentiate Paget's disease from metastatic bone disease. Previous laboratory tests and radiographies of the patients can be helpful. If, for example, laboratory and radiographic studies are uneventful during the past year, diagnosis of the Paget's disease is unlikely.[4] In this case, abnormal skull radiography 2 years earlier suggests the indolent course of the bone disease and rules out metastatic bone disease.

Paget's disease is found more in Anglo-Saxon people. A survey in Johannesburg revealed a prevalence of 1.3% among the black population and 2.4% among the white,[5] and a report on radiological examination of the pelvis showed an estimated prevalence of 1% to 2% in the USA.[6] It is rarely encountered in China, Japan, Iran, India, Scandinavia, Africa, and the Middle East.[7] One important point of this article is extreme rarity of Paget's disease in Iran, as Soltanpur claimed that the 3 cases that he reported in 1978, were the only documented cases of Paget's disease in Iran.[8]

Reduced hemopoietic capacity as a result of Paget's disease-associated fibrosis has been reported,[9,10] which could be the cause of leukopenia in these patients. We did not check the serum level of 1,25-dihydroxy cholecalciferol in our patient, because he was receiving calcitriol at that moment. The serum level of the AP reduced to its reference range after administration of alendronate, which argues against osteomalacia as a possible cause of high AP. In the 2 reported cases of Paget's disease in patients on hemodialysis in the literature, it was the remaining of high serum bone AP after parathyroidectomy that led to the

diagnosis of Paget's disease. In our patient, the serum level of parathyroid hormone was within the normal limits. This urged us to the diagnosis of Paget's disease without any need to decide on parathyroidectomy.

It is not unusual for hemodialysis physicians to be faced with an elevated AP in patients on hemodialysis, because of secondary hyperparathyroidism. But, as we describe in this case, this phenomenon may be due to other simultaneous diseases. Thus, in the evaluation of patients with kidney failure with elevated AP without clinical and laboratory findings of secondary hyperparathyroidism, Paget's disease of bone should be considered in differential diagnosis for patients older than 40 years.

CONFLICT OF INTEREST

None declared.

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