

# PRIMARY HYPERPARATHYROIDISM PRESENTING WITH MULTIPLE PATHOLOGICAL FRACTURES AND NORMOCALCAEMIA

I. A. Mungadi, \*A. O. Amole and \*\*U. H. Pindiga

Departments of Surgery, \*Radiology and \*\*Pathology, Usmanu Danfodiyo University Teaching Hospital, Sokoto, Nigeria

Reprint requests to: Dr. I. A. Mungadi, Department of Surgery, Usmanu Danfodiyo University Teaching Hospital, Sokoto, Nigeria. E-mail [imungadi@yahoo.com](mailto:imungadi@yahoo.com)

## Abstract

The diagnosis of primary hyperparathyroidism (PHPT) is a rarity in developing countries. We report a 30-year old Nigerian farmer seen at the Usmanu Danfodiyo University Teaching Hospital, Sokoto with multiple pathological fractures. The diagnosis of PHPT was made based on these bone changes and the elevated parathyroid hormone level. The patient however had normocalcaemia. Computerised tomography localised a left inferior parathyroid adenoma. He had uneventful parathyroidectomy but developed hungry bone syndrome that was successfully treated with active vitamin D and oral calcium. The differences in presentation between patients from developed countries as well as the apparent rarity of PHPT in tropical countries are stressed.

**Key words:** Hyperparathyroidism, parathyroid adenoma, normocalcaemia, pathological fracture

## Introduction

Primary Hyperparathyroidism (PHPT) is an unstimulated and inappropriately high secretion of parathyroid hormone for the concentration of plasma ionised calcium.<sup>1</sup>

Primary hyperparathyroidism is estimated to be prevalent in approximately 1% of adult population and the usual causes are parathyroid adenoma, hyperplasia and, rarely, parathyroid carcinoma.<sup>2</sup> The diagnosis of PHPT is a rarity in developing countries<sup>3</sup> probably because of the difficulty of making a diagnosis in an environment where there are limited facilities for serum calcium estimation, parathyroid hormone assay and parathyroid gland imaging. The diagnosis can be very perplexing especially because the expected hypercalcaemia associated with PHPT may be masked by calcium, protein or vitamin D deficiency.<sup>4-6</sup> This is a report of a case of PHPT presenting with multiple fractures secondary to parathyroid adenoma but who had normocalcaemia. It is the only documented case of PHPT from the North-Western Nigeria with such an unusual presentation.

## Case report

A thirty year old peasant Nigerian farmer presented on the 11<sup>th</sup> of February 1999 with painful swellings of the right hip, knee and ankle of eighteen months duration. The ankle pain and swelling followed a sprain while working on his farm. Two weeks later, the painful

swellings of the hip and knee developed on falling down while climbing a staircase due to his ailing ankle. He had no other symptoms and no positive family history of similar illness.

On examination, he was chronically ill looking and pale. His weight could not be taken with the available standing scale due to multiple lower limb fractures. He had tender swellings around the right hip, knee and ankle joints. The knee swelling was particularly gross, irregular and warm, with a combination of firm and bony hard areas. All the joint movements around the affected knee were limited. The distal femoral shaft was fractured. Plain radiographs of the pelvis and right knee revealed centrally located intramedullary, expansile radiolucent lesions in the subtrochanteric and supracondylar regions of the femur with pathological fractures respectively (Figures 1 and 2).

The serum calcium level was 2.4 mmol<sup>-1</sup> (normal: 2.2 - 2.6 mmol<sup>-1</sup>), with phosphate level of 1.6 mmol<sup>-1</sup> (normal: 0.8 - 1.5 mmol<sup>-1</sup>) and alkaline phosphatase level of 134 IU/l. The serum albumin was 34g/l and serum calcium corrected for observed albumin was 2.52 mmol/l (corrected calcium in mmol/l = observed calcium in mmol/l + (40 - albumin in g/dl) x 0.02).<sup>1</sup> The parathyroid hormone level was elevated to 132 pmol/l (normal <105 pmol/l). The packed cell volume was 21%; the total leukocyte count was 3.0 x 10<sup>9</sup>/l with an ESR of 150 mm/hour (Westergreen). Serum urea, electrolyte and creatinine were within normal limits. There was no radiological evidence of nephrolithiasis. A contrast enhanced computed tomographic (CT)

scan showed a 1.8 x 1.9 cm non-enhancing discrete lesion in the inferior pole of the left thyroid lobe, consistent with a left inferior parathyroid adenoma (Figure 3). Urinary calcium and acid base status were not assessed.

The patient had a successful neck exploration and parathyroidectomy. The tumour weighed about 18g and showed parathyroid adenoma on histology. He developed hungry bone syndrome with serum calcium level dropping to  $1.7 \text{ mmol}^{-1}$  on the fourth post-operative day. The serum calcium normalised on active vitamin D and oral calcium. The patient was being considered for osteosynthesis by the orthopaedic surgeon when he requested for a discharge against medical advice probably for traditional bone setting. He had since been lost to follow-up.

Figure 1: Radiograph of the right hip showing an expansile centrally located intramedullary radiolucent lesion with a pathologic fracture at the subtrochanteric region

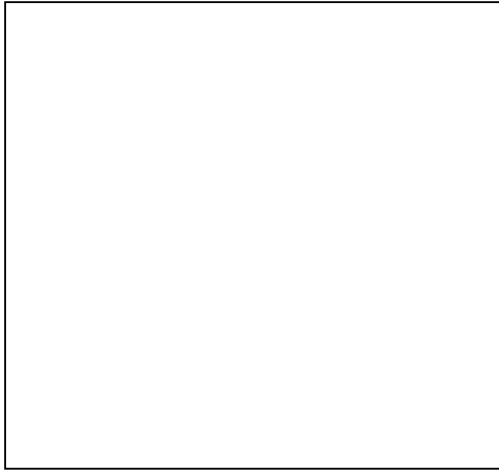


Figure 2: Radiograph of the right knee showing an expansile centrally located intramedullary radiolucent lesion with a pathologic fracture

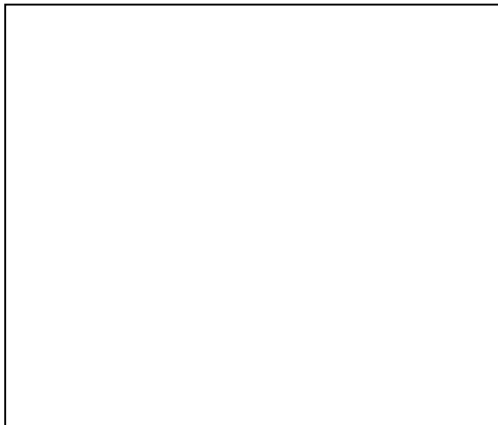
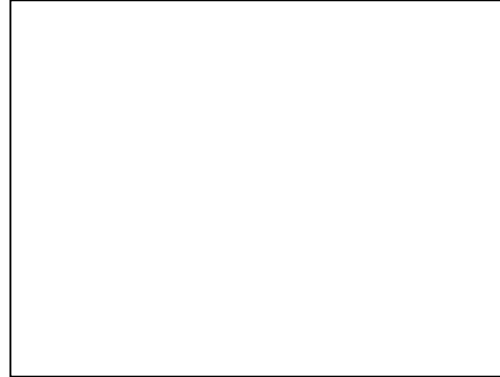


Figure 3: Contrast enhanced computed tomographic image of the thyroid gland showing a round hypodense mass in the posteroinferior aspect of the left lobe (arrow heads)



## Discussion

Primary hyperparathyroidism (PHPT) is a disease commonly due to solitary parathyroid adenoma.<sup>2</sup> It was considered rare in developing countries but recent experience has shown that its apparent rarity may be due to paucity of reports from these countries and also to limited diagnostic facilities.<sup>3,7</sup> Our case has shown that with adequate facilities, more reports of the disease may soon be emanating from developing countries.

Primary hyperparathyroidism (PHPT) has protean manifestations. The advent of automated serum biochemical analysis has highly augmented its diagnosis and has made asymptomatic hypercalcaemia its commonest presentation in the developed countries.<sup>8</sup> This is in contrast to our experience, as in many developing countries, where patients usually present with metabolic bone disease and multiple fractures.

The incidence of metabolic bone disease in patients with PHPT in developing countries is very high.<sup>6,9</sup> The reasons advanced for this is the high prevalence of protein, vitamin D and dietary calcium deficiencies and the high dietary phytate and phosphates in some cultures. The protein deficiency can further reduce total serum calcium since 50% is bound to albumin. These patients therefore, tend to be normocalcaemic even after the correction of hypoalbuminaemia. Our patient is a typical example and similar findings have been noted in previous reports.<sup>5,6</sup> It is therefore pertinent for clinicians practicing in the developing countries to note that nutritional deficiencies can be seen in their patients, unlike those from the developed nations, making hypercalcaemia irrelevant in the diagnosis of PHPT. Hence, serum calcium level may not be part of the criteria for surgical intervention contrary to what was earlier suggested.<sup>10</sup> Multiple fractures, although uncommon, have been described as some of the indicators for pathological fractures in PHPT.<sup>3</sup> They suggest a late presentation of the disease and the severity of the disease. With an increased awareness and

knowledge of the presentation of PHPT in developing countries and the availability of diagnostic facilities, late presentation could be avoided. However, since patients with fractures in Northern Nigerian commonly opt for traditional bone setting as evidenced by our patient, many cases of PHPT may still present late to the orthodox doctors practicing in this area. Furthermore, patients with multiple fractures secondary to a parathyroid adenoma in our environment may find it difficult to appreciate the role of parathyroidectomy in the management of multiple fractures located, for instance, in the lower limbs. They may therefore opt for traditional bonesetters. This unusual situation therefore makes aggressive health education of the populace a necessary priority.

Parathyroidectomy is the treatment of choice in PHPT. Success of the surgery is determined largely by early diagnosis and localisation of the adenoma before surgery.<sup>11, 12</sup> Despite the late presentation of our case, the diagnosis was assisted by radiographic studies including CT and the assay of the parathyroid hormone. These pre-operative investigations made decisions for early surgery possible.

Sudden post-operative hypocalcaemia may be a major complication of parathyroidectomy as shown by our patient. The incidence of this hungry bone syndrome is likely to be high in our environment due to the associated pre-operative dietary calcium and vitamin D deficiency. Therefore, this potential complication should be anticipated and aggressive nutritional support to address these deficiencies must be instituted appropriately.

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