Recurrent pathological fractures in osteitis fibrosa cystica: a case of undetected primary hyperparathyroidism due to parathyroid adenoma

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Introduction

Osteitis fibrosa cystica (OFC) is a skeletal disorder which results in exposure to high levels of parathormone (PTH), mainly due to untreated primary hyper-parathyroidism (PHPT). This is a case of primary hyperparathyroidism detected in a patient investigated for pathological fractures. Since this is a treatable condition early detection and treatment by routine assessment of serum calcium and phosphate levels in the aged population must be emphasised especially in developing countries.

Key Learning Points

Early detection and treatment in Primary Hyper-parathyroidism will prevent the disease progression into Osteitis Fibrosa Cystica

Serum calcium and phosphate levels should be done as routine investigations in elderly patients presenting with fractures

Case

A 58 year old female presented with a left supracondylar fracture of the humerus (Figure 1) following an accidental fall on her outstretched hand. She had had a sub-trochanteric fracture of the right femur five years back, which had been fixed with a K nail. (Figure 2). She had a goiter which was only visible on neck extension.

X rays of the left arm and the right lower limb revealed large osteolytic bony lesions in the left distal humerus (Figure 1) and the left middle third of the tibia (Figure 3). In the hand X-ray, subperiosteal resorption of phalangeal bones was evident (Figure 4).

Biochemical investigations revealed elevated levels of alakaline phosphatase (ALP), high serum total and ionised calcium levels, increased parathyroid hormone (PTH) levels and low phosphate levels were noted. Her thyroid functions were normal. Ultrasound scan of the neck revealed a multi-nodular goitre with a hyperechoic nodule in the infero-posterior aspect of the left lobe of thyroid raising the possibility of a parathyroid adenoma and contrast CT scan of the neck confirmed the left parathyroid adenoma and the multiple heterogenous enhancing thyroid nodules. Osteitis fibrosa cystica due to primary hyperparathyroidism was diagnosed.

She underwent a left hemithyroidectomy with left parathyroidectomy. Histology revealed a parathyroid adenoma. Following surgery the serum calcium level demonstrated a rapid drop and calcitriol with calcium lactate supplements were needed to maintain normal serum calcium levels.

Discussion

PHPT is a condition with a female preponderance and the peak incidence will be at age 30-50 years. One third of PHPT patients are asymptomatic. The annual incidence is 2-10/10000 and nearly 60% of them will be identified during routine biochemical investigations [1].

Majority of the PHPT cases are due to solitary or multiple parathyroid adenomas. Only 20% would be due to parathyroid carcinoma, hereditary causes like MEN type I/IIa/IIb or renal osteodystrophy [2].

Elevated PTH levels will stimulate bone resorption via high osteoclastic activity, mobilising large amounts of calcium in to the blood stream. Thus, the altered calcium metabolism will lead to renal calculi, peptic ulcers, loss of appetite, loss of weight and weak bones which is referred to as 'bones, stones, abdominal groans and psychic moans' [3].

Elevated serum levels of calcium, PTH and ALP, low phosphate levels with radiological evidence of bone

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resorption are characteristic findings in PHPT. The first bones to show subperiosteal erosions are the bones of the hand. Characteristic X-ray changes in the skull are described as ground glass or salt and pepper appearance [4].

Osteitis fibrosa cystica also known as osteitis fibrosa, osteodystrophia fibrosa, Von Recklinghausen's disease of bone, is a skeletal disorder caused by prolonged exposure of bone to elevated PTH levels in PHPT. Gerhard Engel in 1864 and Friedrich Daniel von Recklinghausen in 1890 described this condition for the first time [5].

The progression of PHPT to OFC is relatively uncommon in developed countries during the past few decades due to the improved techniques in early detection and treatment of the condition [3].

But still it is not uncommon in developing countries as a result of failure in early diagnosis and treatment or poor patient compliance.

OFC is evident in X-ray films as multiple cystic lesions in bones. These are known as 'brown tumours'. Brown tumours are benign hypervascular focal lesions in bones caused by increased osteoclastic activity and fibroblastic proliferation [6]. They are seen commonly in facial bones, mandible, sternum, pelvis, ribs, femurand rarely in vertebrae [6]. Even though pathological fractures in PHPT are relatively uncommon [7], areas of bone with brown tumours are vulnerable to fractures with trivial trauma.

The treatment of choice would be total para-thyroidectomy. It has been shown to result in the reversal of bone resorption and the complete regression of brown tumours [8]. According to some reports, total para-thyroidectomy, even in parathyroid carcinoma will be the remedy to control the symptoms of hyperparathyroidism and OFC [9]. Bone transplants can also be used to fill the lesions (cavities) caused by OFC [10].

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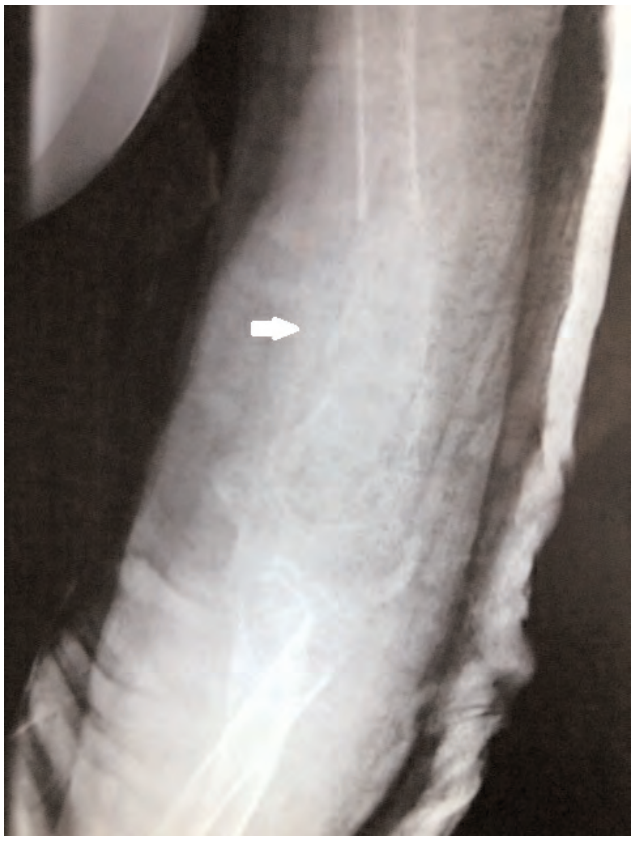

Figure 1. White arrow indicates a left supracondylar fracture of the humerus at the site of a large cystic lesion of the bone (brown tumour), managed with manipulation under anesthesia and plaster cast.

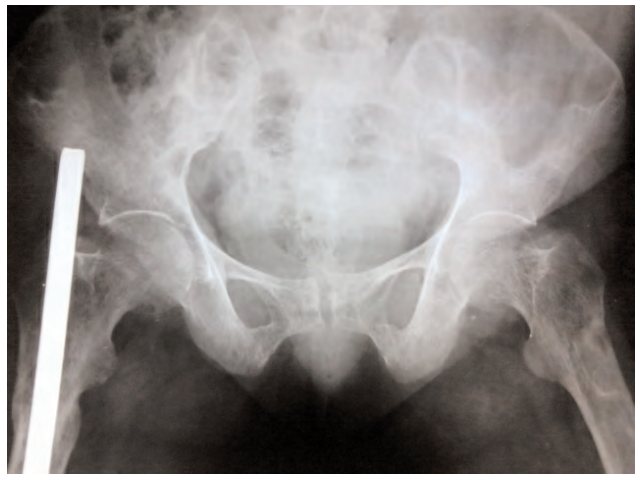

Figure 2. Right femur sub-trochanteric fracture fixed with a K-Nail

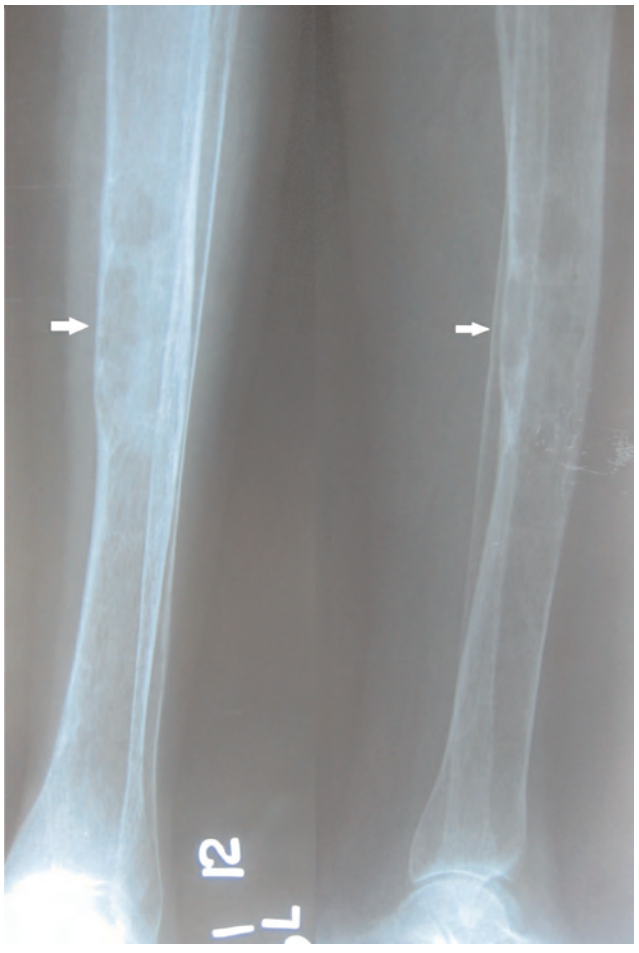

Figure 3. White arrows indicate large osteolytic bony lesion in the left middle third of the tibia (brown tumour) in AP and lateral X-ray views of the left leg.

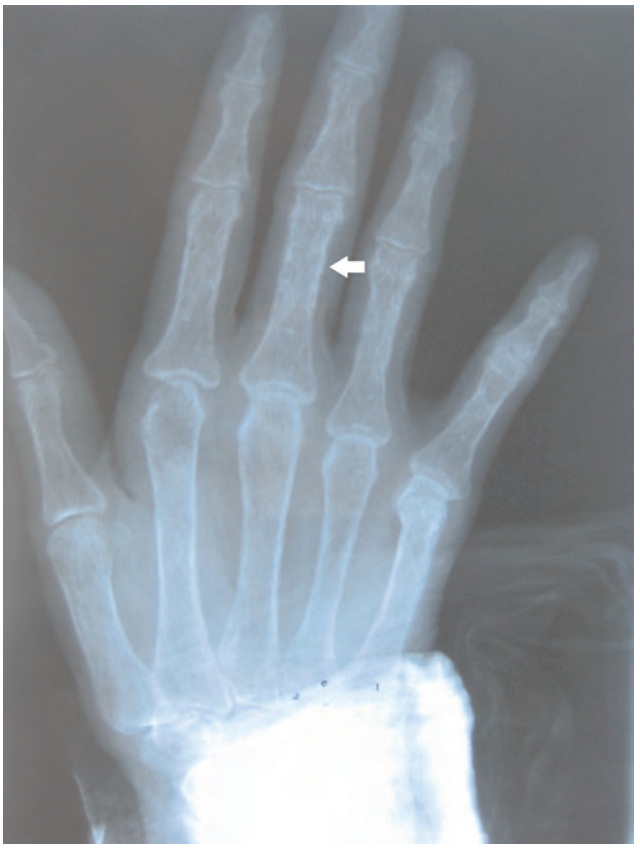

Figure 4. White arrow indicates subperiosteal resorption of phalangeal bones.