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A case report of pseudohypoparathyroidism with bilateral fracture of neck of femur

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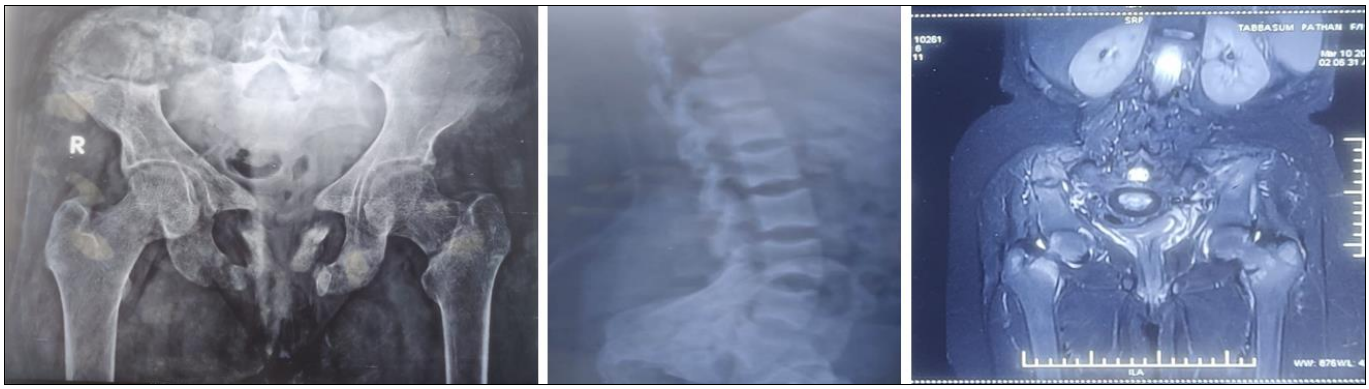
Abstract
A 19 year old female presented with a spontaneous onset of progressive non-traumatic b/l hip pain with inability to walk since 5 weeks. P/h/o pulmonary TB 2 years back present. 5 drug AKT taken for 9 months. Local tenderness was elicited and xray and MRI pelvis with both hips and LS spine was done which showed pathological fracture of basal neck of femur both side with severe listhesis of lumbosacral spine. Diagnosis of Pseudohypoparathyroidism was made on the basis of blood investigations which showed hypocalcaemia, hyper- phosphatemia, raised PTH, low Vit D3 level. Fixation of bilateral of neck femur with cancellus screw done to provide compression both side along with calcium and Activated Vitamin D3 supplimentation. Patient showed excellent response.

Keywords: Non traumatic, hypocalcemia, hyperphosphatemia, raised PTH, cancellous screw, calcium, vit D3

Introduction
Pseudohypoparathyroidism is uncommon metabolic disorder in which body fails to respond to parathyroid hormone characterized by hypocalcemia and hyperphosphatemia but in contrast to Hypoparathyroidism there is a significantly raised PTH level [1]. In 1942 Fuller Albright introduced the term pseudohypoparathyroidism for patients presenting with PTH resistant hypocalcaemia, hyperphosphatemia and raised PTH along with a constellation of developmental and skeletal defects, collectively termed Albright Hereditary Osteodystrophy (AHO). These includes: short stature, rounded face, obesity, shortened fourth metacarpals, dental hypoplasia, soft tissue ossification. On the basis of AHO it is subdivided into type Ia and type Ib, where PHP type Ia shows AHO, while type Ib lacks AHO.

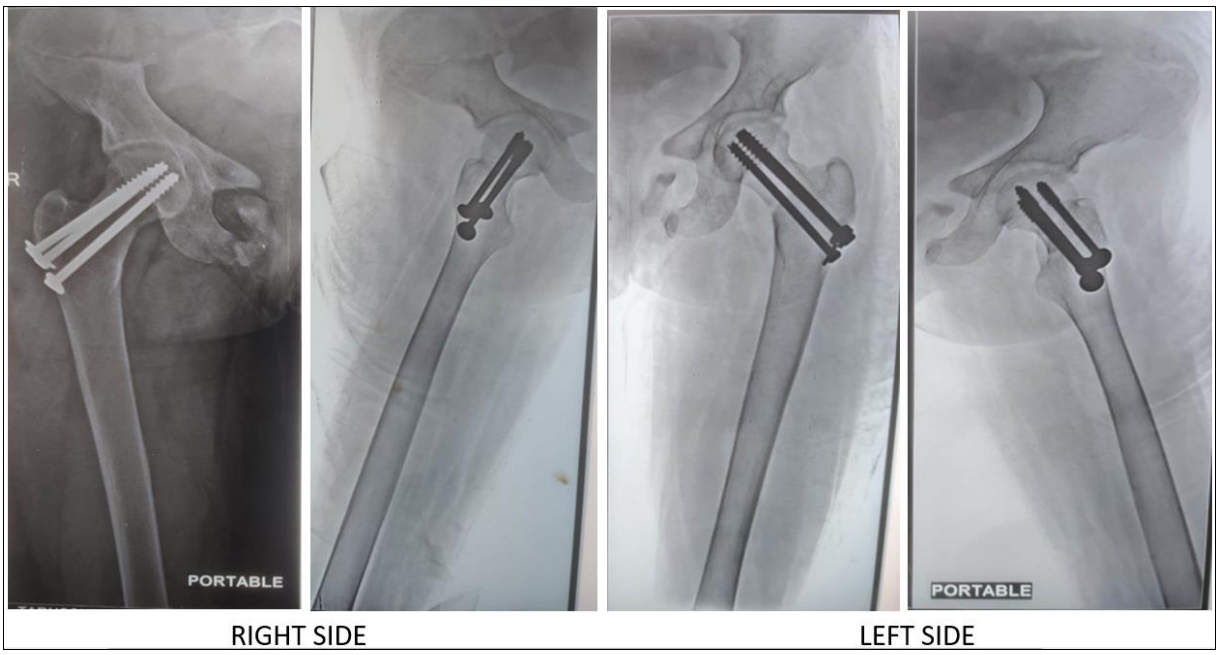
Case report:
A 19 year old female presented with a complain of spontaneous and gradual onset of progressive b/l hip pain since 5 weeks with inability to walk. No history of trauma was present. Physical examination showed tenderness on both hips. P/h/o pulmonary TB 2 years back was present. 5 drug AKT taken for 9 months.no c/o fever present. No significant family h/o. Sputum CBNAAT & AFB performed to rule out active tuberculosis. X ray and MRI pelvis with both hips and LS spine was done which showed pathological undisplaced fracture of basal neck of femur both side with severe listhesis of lumbosacral spine. Diagnosis of Pseudo-hypoparathyroidism was made on the basis of blood investigations which showed hypocalcaemia, hyper- phosphatemia, raised PTH, low Vit D3 level with normal thyroid levels. Endocrinologist reference was done & Treatment initiated with calcium and Activated Vitamin D3 supplimentation once in a day. The goal of treatment is to maintain serum total and ionized calcium within reference range to avoid hypercalciuria and to suppress PTH levels to normal. This is important because elevated PTH levels in patients can cause Increased remodelling and lead to hyperparathyroid bone disease. calcium and alpa-1 hydroxylated vitamin D3 is a mainstay of treatment [1]. Patient was managed with bilateral skin tractions till normalization of biochemical profile which took around 1 month after starting vit D3 and calcium. After correction of biochemical profile fracture was managed with three partially

threaded screws. Under spinal anaesthesia both fractures were reduced on traction table under fluoroscopic guidance & three 6.5 mm partially threaded cannulated cancellous screws were inserted in inverted triangle fashion to provide compression on both side. Patient discharged after uneventful post operative period with continuing medical management. Both hips were kept in abduction with help of abduction bar. Weight bearing was started after 6 weeks postoperatively. Medical management was continued for 6 months postoperatively.



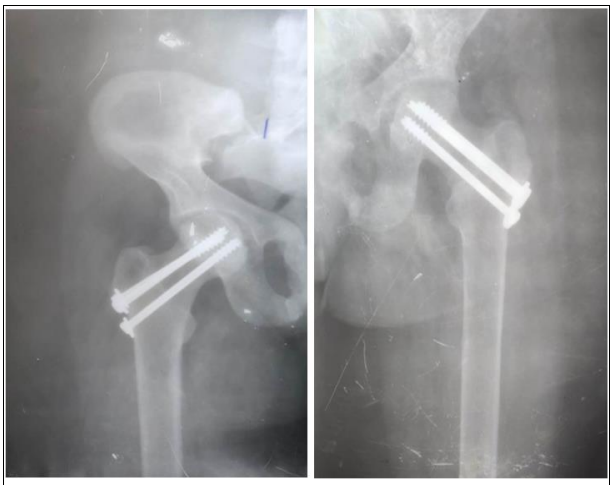
Parameters	Value	Normal
S. Calcium (mg/dl)	8.2	8.5 - 10.2
S. Phosphate (mg/dl)	5.5	3.4-4.5
S.ALP (IU/L)	208	Up to 117
S.PTH (pg/ml)	505.30	10- 65
S.25 Hydroxy Vitamin D3(ng/ml)	10.1	30-100
T3 (nmol/l)	1.71	1.2-2.8
T4 (nmol?l)	82.9	77-155
TSH (mU/L)	2.29	0.3-4

Immediate postoperative x-rays



At 6 month follow up

Parameters	Value	Normal
S. Calcium	8.8	8.5-10.2
S. Phosphate (mg/dl)	4.4	3.4 -4.5
S.PTH	121.1	10-65
S.ALP (IU/L)	161	up to 117
25 Hydroxy VIT D(ng/ml)	32	<10



RIGHT SIDE LEFT SIDE

Discussion

- Pathological fracture of neck of femur uncommon in young adults & the treatment of choice is early fixation [2]. As delayed fixation of these fractures can lead to nonunion, osteonecrosis of femoral neck.
- Detailed investigations related to the cause in pathological fracture of neck of femur required to rule out or confirm a particular condition associated with such pathological fracture.
- Serum electrolytes, bone metabolism related endocrine levels including thyroid and parathyroid profile, vit. D3 levels, ALP levels and renal function tests should a form a standard protocol in diagnosing such condition.
- Due to its rarity high level of suspicion is reqiured on the part of clinician in dealing with such fractures.
- The finding of biochemical hypocalcaemia associated with hyperphosphataemia could raise the possibility of hypoparathyroidism. Furthermore, the lytic lesions on radiograph could be explained on the basis of increased production of parathyroid hormone (PTH) presumably as a result of chronic hypocalcaemia. The absence of specific dysmorphic features or mental retardation initially described by Albright's *et al.* and a normal thyroid function, therefore, should raise the possibility of pseudohypohyperparathyroidism the so-called pseudohypoparathyroidism type Ib.
- Different subtypes of pseudohypoparathyroidism (PHP) seem to be related to different pathophysiologic mechanisms1. In PHP type I, PTH is unable to elicit cyclic AMP (c AMP) production in target cells and administration of exogenous PTH does not increase urinary c AMP production. PHP type II is the least common form. This variant of PHP is typically a sporadic disorder, although one case of familial PHP type 2 has been reported. Patients do not have features of AHO. Renal resistance to PTH in PHP type 2 is manifested by a reduced phosphaturic response to administration of PTH, despite a normal increase in urinary c AMP excretion [3]. In 60% of patients with PHP type I, there is a 40% to 50% reduction in the N protein of erythrocytes, platelets or cultured fibroblasts, the N protein complies a number of membrane receptors, including that for PTH to adenylate cyclase. These patients classified as PHP type Ia, usually present with dysmorphic features described by Albright. The N protein appears to be present universally in cells, and some patients with PHP type Ia are affected by other endocrinopathies (presumably due to end-organ resistance) such as abnormal thyroid response to thyroid-stimulating hormone (TSH) and hypothyroidism, hypogonadism, and decreased cyclic AMP generation in response to glucagon. In these patients, end organ resistance appears to be progressive over the first 2-3 years of life, with dysmorphic features and possible migratory calcification preceding hypocalcaemia and elevation of serum PTH concentrations. In the PHP type IB patients the N protein contents of cells is normal and no dysmorphic features are found. In these patients, the exact molecular basis for PTH resistance is yet undetermined. A wide spectrum of skeletal or renal end-organ resistance to PTH has been reported in some patients with PHP type I. End-organ resistance is found in the kidney but the bone is normally responsive. These patients, classified as PHP type I with osteitis fibrosa or pseudohypohyperparathyroidism have a combination ofhypocalcaemia and hyperphosphataemia with skeletal signs of hyperparathyroidism. Patient with skeletal but no renal resistance (pseudo-pseudohyoparathyroidism) present the constitutional features of PHP without hypocalcaemia or hyperphosphataemia. In these patients, skeletal defects may be more severe in the females as a result of early epiphyseal closure. In PHP type II, cyclic AMP production in urine is normally elicited by PTH, but phosphaturic response is profoundly decreased. The end-organ resistance is presumed to be due to defective tubular response to cyclic AMP. Interestingly, in these patients, restoration of normocalcaemia by treatment with Vitamin D and calcium also restores the phosphaturia response to PTH.

Conclusion

We had extremely rare presentation of pseudohypoparathyroidism with b/l femoral neck fractures. Early diagnosis with medical therapy for normalization of biochemical profile followed by close or open reduction and fixation with cannulated cancellous screw provide excellent results. Patients was able to squat and walk without support and carry out daily activities at 6 month follow up. So, early diagnosis combined with early fixation of fracture provides excellent outcome.

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