

PRIMARY HYPERPARA THYROIDISM: A CASE REPORT

MD. FAIZUL ISLAM CHOWDHURY¹, M A K KHANDAKER², HOSSAIN M. ZAID³
BIPLAB KUMAR SAHA⁴, MD. MURAD HOSSAIN⁵

Introduction:

The old saying, 'Bones, stones, abdominal groans & psychic moans' is still relevant in primary hypo parathyroidism Here¹ –

Bones = bone pain & pathological fracture.

Stones = in Kidney & rarely in gall bladder.

Abdominal groans = peptic ulcer & pancreatitis.

Psychic moans = usually depression & rarely other psychiatric disorder.

The patient is usually asymptomatic (>70%). The patient may present with other multiple symptoms and signs including tiredness, malaise, Vomiting, loin pain due to nephrolithiasis(40% to 50% cases), Polyuria, thirst, Arthritis and less frequently extensive bone resorption & osteitis fibrosa cystica (Brown tumour). Bony lesions occur in 5% to 10% cases. Important signs are – muscle hypotonicity, band keratopathy & hypertension (less common). The manifestation may be subtle and the disease may have a benign course for many years or life time. This milder form is usually termed as Asymptomatic Hyperparathyroidism. Rarely the disease may progress abruptly and cause complications like severe dehydration and coma known as parathyroid crisis.

Primary Hyperparathyroidism is a generalized disorder of Calcium, Phosphate and bone metabolism due to increased secretion of PTH. The elevation of circulating hormone usually leads to Hypercalcemia and Hypophosphatemia. The elevated level of hormone is caused by autonomous secretion of PTH by a single parathyroid adenoma of varying size.

Case Report :

A 20 year old Muslim young chap came from Gabtoli, Dhaka, Admitted to DMCH on 25th Nov. 07 with the complaints of continued dull pain in the left loin,

sometimes reduced by taking drugs associated with burning micturation and severe unremitting vomiting for the last 2 years. Bouts of vomiting was accentuated after meal but persisted even after emptying all contents. After 2 months he visited a consultant, diagnosed as a case of left renal stone and underwent PCNI. After operation the pain reduced in intensity but vomiting and retching continued. For the last 8 months, he sustained fractures after trivial trauma on several occasions; first in right upper arm then in right thigh which was treated with skeletal traction. 4 months back there was another history of fracture in left thigh after trivial injury which was left untreated. During the period of illness he became crippled and bed ridden. He gave no history of diarrhoea, headache, visual disturbances, cough of haemoptysis.

On examination patient was ill looking, having below average body build, mildly anaemic and dehydrated, not icteric, thyroid gland was not enlarged, accessible lymph nodes were not palpable. Examination of alimentary system revealed nothing but mild tenderness in left lumbar region on deep palpation. There was no lump, no organomegally or ascitis. Examination of all other systems were normal except examination of limbs, which revealed wasting of muscles of all group of both limbs, bowing of both humeri, right thigh medially rotated, long slender fingers & toes, movement restricted with varying degree in both limbs. sensation intact, all peripheral pulses were palpable.

In investigations we have found:

ESR : 69mm in 1st hour, TC : 10200/cmm, DC:N-54%L-37% M-6% E-3%, Hb : 48%,

Urine R/E-Phosphate-Present Pus cell : 30-35/HPF, Amorphous Phosphate-present.

1. Associate Professor, Department of Medicine, Dhaka Medical College
2. Professor, Department of Medicine, Dhaka Medical College
3. Registrar, Department of Medicine, Dhaka Medical College Hospital
4. Intern Doctor, Dhaka Medical College Hospital
5. Assistant Registrar, Department of Medicine, Dhaka Medical College Hospital

S. Calcium- 14.5mg/dl , S. phosphate: 2.4mg/dl(2.5-5.0), S. Parathormone : 690 pg/ml (9-80) .RBS- 5.7mmol/l, Blood group – O positive, ECG- LVH, Echo- Normal

.USG of neck to see parathyroid gland: Hypoechoic mass in lower pole of left lobe of thyroid -Parathyroid adenoma/hyperplasia ,Parathyroid scanning.

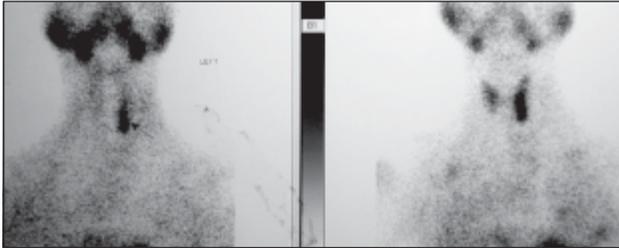


Fig. 1: *⁹⁹Tc-sestamibi scanning showing uniform uptake of the radioactive isotope by both lobe of thyroid gland. After 2 hours the isotope uptake is evident only in parathyroid gland. High probability of parathyroid adenoma /hyperplasia.*

Other investigations to see the effects of hyperparathyroidism:

X-ray skull & both phalanges both view:

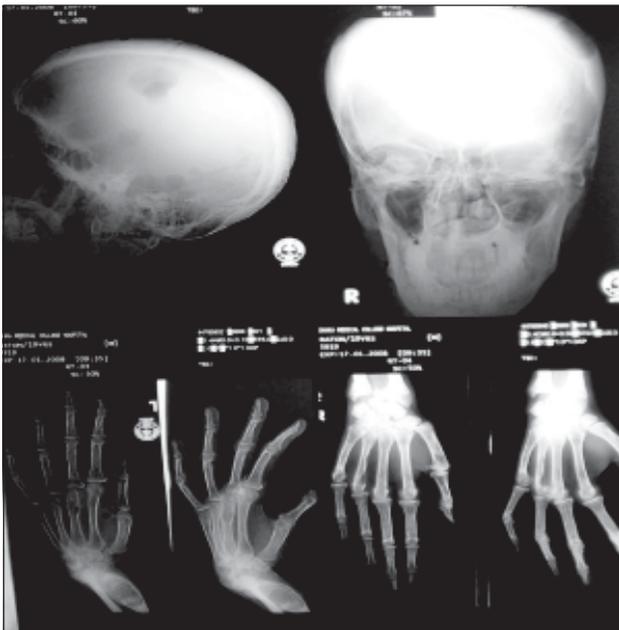


Fig.-2: *Multiple lytic lesion in skull & bone resorption in phalanges.*



Fig 3: *Left sided Renal stone*



Fig 4: *X-ray pelvis with both hip A/P view showing Generalized osteoporotic changes*

Multiple lytic lesions noted in left ileal blade and upper femoral shaft.

Discussion:

Primary Hyperparathyroidism is the most common cause of parathyroid disorder with a prevalence of 1 in 800. It is two to three times more common in women than in men and 90% of the patients are over 50 years of age. The disease has a pick incidence

between third and fifth decades but seen in young adults in rare occasions.

The cause of hyperparathyroidism is one or more hyper functioning glands. A single abnormal gland occurs in 80% of patients. The abnormality in the gland is usually a benign neoplasm or adenoma and a parathyroid carcinoma.

Hereditary hyperparathyroidism can occur without other endocrine abnormalities but is usually part of a Multiple Endocrine Neoplasia Syndrome (MEN).

Adenomas are most often located in the inferior parathyroid glands, but in 6-10% of patients, parathyroid adenomas may be located in the thymus, the thyroid, the pericardium, or behind the esophagus. Chief cells are predominant in both hyperplasia and adenoma. Hyperparathyroidism from a parathyroid carcinoma may be indistinguishable from other forms of primary hyperparathyroidism, a potential clue to the diagnosis is however provided by the degree of calcium elevation. Calcium values of 3.5 to 3.7 mmol/L (14-15 mg/dl) are frequently associated with carcinoma.

Two fundamental types of genetic defects have been identified in parathyroid gland tumours-

1) over activity of protooncogenes 2) loss of function of tumour suppressor genes. Mutations in MEN1 gene locus on chromosome 11q13 is responsible for causing MEN 1. There are two rare syndromes associated with hyperparathyroidism that involve one or more genes located on chromosome 1q. The hereditary hyperparathyroidism jaw tumor syndrome shows an autosomal dominant inheritance pattern, which may be associated with parathyroid carcinoma. Parathyroid carcinoma may also appear in the other syndrome familial isolated primary hyperparathyroidism. Allelic deletion has been identified in all parathyroid carcinomas. It is also seen in 10% of adenomas.

Skeletal and radiological changes – Osteitis fibrosa cystica is the classical hyperparathyroid bone disease resulting from increased bone resorption by the osteoclasts (Howship's lacunae) with fibrous replacement in the lacunae. This may present as bone pain and tenderness, fracture and deformity.

There are characteristic changes on plain x-rays. In the early stages there is demineralization with subperiosteal erosions and terminal resorption in

the phalanges. A 'pepper pot' appearance or loss of lamina dura may be seen on lateral x-rays of the skull. In nephrocalcinosis scattered opacities may be visible within the renal outline.

Reduced bone mineral density can be assessed by DEXA scan.

⁹⁹Tc-sestamibi scanning, USG, CT and selective neck vein catheterization with PTH measurement are the effective ways to locate the parathyroid tumour.

Management:

Hypercalcemia in patients with primary hyperparathyroidism responds less well to glucocorticoids and bisphosphonates than in those with malignancy. Currently the only long term therapy is surgery, with excision of a solitary parathyroid adenoma or debulking of hyperplastic glands. Postoperative hypocalcaemia is not uncommon during the first two weeks.

References:

1. Shonni J, Silverberg, MD and John P, Bilezikian, Evaluation & Management of Primary Hyperparathyroidism. The Journal of Clinical Endocrinology & Metabolism Vol 82, No 3983
2. Goshan O, Aviel Ronen S, Brown Tumour of Hyperparathyroidism in the mandible associated with atypical parathyroid adenoma. Journal of Laryngeal & otolaryngology 2000, 114:302-4.
3. Lloyl HM, Aitken RE, Primary Hyperparathyroidism resembling rickets of late onset BMJ 1965ii, 853-6.
4. Parveen Kumer, Michael Clark. Clinical Medicine, 6th ed. WB Saunders, Edinburgh 2007
5. Walker BR, Colledge NR, Boon NA. Davidson's Principles & Practice of Medicine, 20th ed. Churchill Livingstone, 2006.
6. JS Dinnen, RH Greenwood, JH Jones, DA Walker. Parathyroid carcinoma in familial Hyperparathyroidism, Journal of Clinical Pathology 1977.
7. Mitchell E Tublin John H. Yim. Recurrent Hyperparathyroidism secondary to parathyromatosis. Journal of Ultrasound Medicine 26: 847-851.
8. Yasuo Imanishi, Hideki Tahara. Clonal Chromosomal Defects in the Molecular Pathogenesis of Refractory Hyperparathyroidism of Uraemia. Journal of American Society of Nephrology 13: 1490-1498, 2002.