

A Case of Primary Hyperparathyroidism due to Ectopic Mediastinal Parathyroid Adenoma Presenting With Pathological Fracture

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Abstract: Primary hyperparathyroidism (PHPT) may cause skeletal deformities with pathological fracture in symptomatic patients if untreated. We present a case of 23 years old male patient presenting with recurrent pathological fracture from trivial trauma. Subsequent biochemical, radiological and scintigraphic findings revealed PHPT from an ectopic mediastinal parathyroid adenoma and concomitant vitamin-D deficiency. Initially the patient was treated conservatively with hydration and forced diuresis for hypercalcaemia. Finally the tumor mass was resected by sternotomy and histopathology report of the excised tumor was consistent with parathyroid adenoma.

Keywords: Hyperparathyroidism, Scintigraphic, Mediastinal, Hypercalcaemia, Sternotomy

1. Introduction

Primary Hyperparathyroidism may present with unusual clinical features like pathological fracture due to resorption of bone and rarely it may be due to ectopic location of parathyroid gland. Inability to locate the adenoma in ectopic parathyroid gland often delay the diagnosis and treatment [1]. Mediastinal locations of parathyroid glands are uncommonly encountered which presents diagnostic difficulty. Nuclear imaging scintigraphy accurately localizes the tumor in 90% of cases [1,2] and simplifies the surgical management. We present such a case of mediastinal parathyroid adenoma which initially created diagnostic dilemma due to ectopic location

and unusual clinical presentation. Preoperative scintigraphy determined the location and helped us to select surgical approach.

2. Case Report

A 23 years old male patient presented initially in the orthopedic department with subtrochanteric fracture of left femur and fracture of left clavicle on trivial trauma due to fall. The fracture of femur was operated for internal fixation and clavicle fracture was managed conservatively.



Subtrochanteric Fracture Left Femur



Fracture Left Clavicle

Afterwards the patient was thoroughly investigated to determine the cause of pathological fracture. The blood is

biochemically examined for metabolic bone disease and it revealed-

parameters	value	Normal value	remarks
Serum calcium	11.2 mg/100ml	8.8 – 10.6 mg/100ml	Hypercalcaemia
Serum Vitamin D total (25-hydroxy vitamin D2 & D3)	10.90 ng/ml	Deficiency <20ng/ml Insufficiency 20-30ng/ml Sufficiency 30-70ng/ml Toxicity >70ng/ml	Elevated
Serum Alkaline Phosphatase	1396 Units/L	40-130 Units/L	Elevated
Serum Parathormone intact	3216 pg/ml	15-65pg/ml	Hyperparathyroidism

All these features point towards diagnosis of primary hyperparathyroidism. Radiological investigations are also supportive of primary hyperparathyroidism. Radiograph of skull shows multiple lucent areas at the skull vault. Xray of both knee joints also revealed severe osteopenia with multiple

lucent areas. Finally X-ray of both hands also revealed subperiosteal bone resorption of middle and distal phalanges which is very much characteristics of primary hyperparathyroidism.



X-Ray Skull



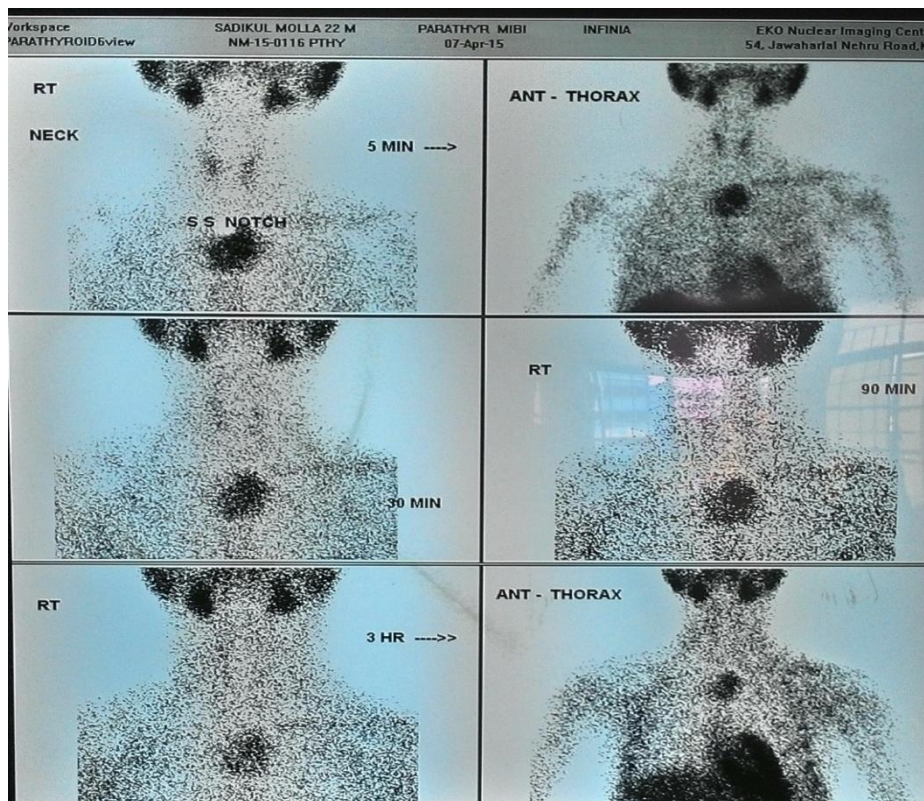
X-Ray Knee Joint



X-Ray Hand

After that in search of the site of hyperparathyroidism an ultrasonography of neck was performed but it did not revealed enlargement of any neck glands. Subsequently a radionuclide parathyroid scan was performed with 99m Tc MIBI, which shows a fairly large oval shaped area with moderately increased uptake of radiotracer over retrosternal region immediately below the sterna notch. The washout of radiotracer from the thyroid gland is uniform except from the

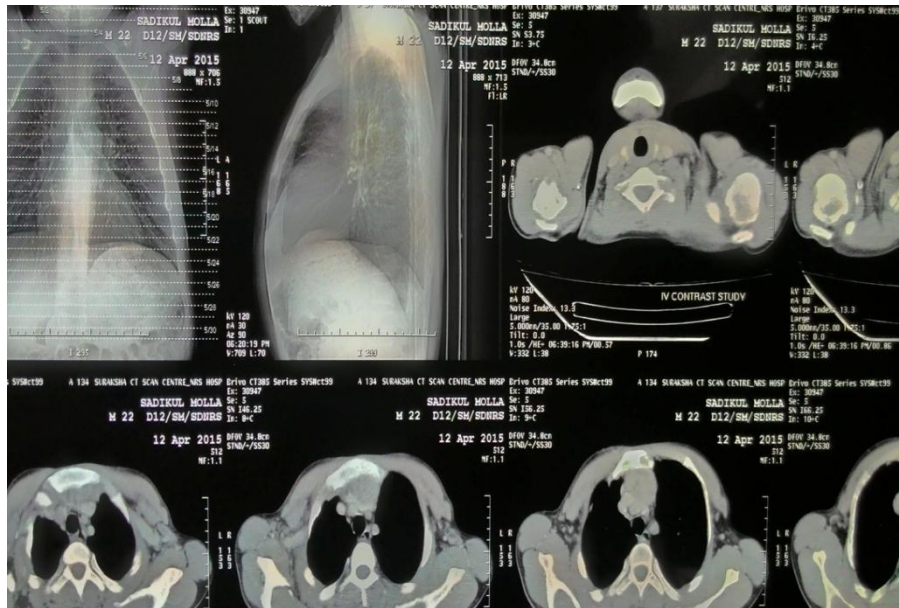
above mentioned oval shaped area which remained persistently high till three hours of study. No other abnormal extrathyroidal focal radiotracer concentration is observed over the neck or mediastinum. So the scintigraphic findings appear to be consistent with mediastinal extrathyroidal parathyroid adenoma.



TC 99M SESTAMIBI SCAN

Next to delineate the actual position of the mass and its relation with surrounding structures a CECT chest was done. A 3.9x2.9 cm size heterogeneously enhancing mass lesion is noted in the anterior mediastinum causing compression on left

brachiocephalic trunk along with erosion of posterior margin of sternum.



CECT CHEST

Operative intervention was decided and the median sternotomy was done to access the anterior mediastinum. The mass is found to be adhered with surrounding structure and there is no other lesion in the mediastinum. The mass is

carefully dissected out from the surrounding structures. Special care was taken during dissection from underlying innominate vein to avoid injury of the vasculature. Finally hemostasis was secured and the sternum is closed.



Resection of Mediastinal Tumor



Tumor Bed after Resection

The post operative recovery was uneventful without any episode of hypocalcaemia. Biochemical examinations were repeated at 2 days interval and the serum calcium and parathormone level gradually normalized over next 3weeks.

3. Discussion

Primary hyperparathyroidism is a common endocrine condition causing metabolic bone disease characterized by hypercalcemia and diffuse bone resorption. A large number of cases are usually asymptomatic and are incidentally detected with hypercalcemia [1,2]. In the clinically manifested disease, it is known to present with subtle and protean manifestation,

leading to misdiagnosis in the early stage of the disease. Excess PTH leads to involvement of skeletal system and the kidneys in the majority of the cases. Skeletal involvement is mainly as a result of increased bone resorption leading to characteristic manifestations like osteitis fibrosa cystica (OFC), subperiosteal resorption of distal phalanges, bone cysts and brown tumors. Renal involvement is seen in more than 15% of the cases of primary hyperparathyroidism and is mainly due to hypercalciuria leading to nephrocalcinosis and nephrolithiasis (renal stone)[1,2].

Diagnosis of primary hyperparathyroidism in a clinically suspected case is suggested by hypercalcemia, hypophosphatemia, raised levels of bone specific alkaline phosphatase and raised intact parathyroid hormone (PTH) levels. Anterior neck mass may occasionally be palpable in a case of parathyroid tumor[1].

The superior and the inferior parathyroid glands originate from the 4th and 3rd branchial pouches respectively and migrate caudally to occupy their normal positions in relation to the thyroid gland. Any aberrancy during this descent may lead to ectopic locations of these glands. They may be located in posterior mediastinum behind the cervical esophagus, retrosternally in the anterior mediastinum, within the thymus (intrathyroidic), in the tracheo-esophageal groove or unusually within the thyroid parenchyma (intrathyroidal)[1,2].

Ectopic location of parathyroid gland is an important albeit, uncommon reason for failure to locate the gland during surgery. Pre-operative ^{99m}TcSestamibi scan helps in localizing the tumor accurately in almost 90% of patients[1,3]. It has a greater role in localizing ectopic glands which helps the surgeon in planning the surgical approach. In cases of recurrence of the disease or failed surgery, localization of adenoma by sestamibi scan is mandatory[[5]. Other modalities to localize and delineate the mass preoperatively include CT scan and MRI and FNAC guided by imaging modalities[1,6,7].

Open surgical excision remains the standard treatment for mediastinal parathyroid adenoma. Thoracoscopic removal and angiographic ablation could be considered in selected cases.

4. Conclusion

Primary hyperparathyroidism with its varied manifestations and indolent course is a condition well known to pose a diagnostic dilemma to the clinician. An ectopic location of the parathyroid gland, albeit uncommon, may further complicate the issue. Preoperative scintigraphy helps in confirming the location of the adenoma and simplifies the surgical management.

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