Primary Hyperparathyroidism due to Ectopic Mediastinal Parathyroid Adenoma

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Abstract: Parathyroid Adenoma is the commonest cause of primary hyperparathyroidism. Majority of adenomas arise from eutopically positioned parathyroid glands. However in 10% of the cases they can be from ectopic parathyroid glands. Aberrant migration during development may lead to ectopic locations of parathyroid glands. Proper pre operative localization is crucial for successful surgical outcome. We describe a case of primary hyperparathyroidism due to parathyroid adenoma in mediastinum, which was excised using sternotomy.

Keywords: Parathyroid gland, Aberrant migration, Mediastinum, Parathyroid adenoma.

INTRODUCTION
Primary hyperparathyroidism (PHPT) is due to adenoma in majority of the cases. About 85% of these cases are due to eutopic adenoma [1] but multigland disease can occur in 10-15% of cases [2]. However in 10% of the cases these adenomas can be in ectopic position. The ineffectiveness of surgical exploration without preoperative localization is known [3]. Various imaging modalities like USG, CT, MRI, MIBI scintigraphy are commonly used [4], where as PET and SPECT with CT fusion imaging are performed in difficult cases [4-6], to localize parathyroid adenoma either eutopic or ectopic.

However use of TC99 MIBI scan with CT scan is sufficient in majority of the cases to localize ectopic adenoma [3, 7].

Here we are reporting one such case of primary hyperparathyroidism due to ectopic mediastinal adenoma. The purpose of this case report is to demonstrate the usefulness of preoperative imaging.

CASE REPORT
A 35 yrs old woman presented with symptoms of epigastric pain, occasional episodes of vomiting, muscle aches and bony pains of six years duration. She gradually developed anorexia and insomnia. She also had difficulty in walking and getting up from squatting position one year prior to presentation. She was admitted for these complaints elsewhere and was found to have hypercalcemia and renal calculi, hence had been referred to our centre for further evaluation and management. She was not on any medication which can cause hypercalcemia like lithium or excess doses of vitamin D. None of the family members had similar problem. She was treated for pulmonary Koch’s 8 yrs ago. Clinical examination revealed mild pallor, had no goiter, no other obvious neck swelling, no skeletal deformities. She was normotensive and except for painful proximal myopathy other systemic examination was normal.

Investigations- Routine biochemistry was normal except for anemia with hemoglobinof 9 gm/dl. Her serum calcium (corrected) 12.4 mg/dl, phosphorus 2.4 mg/dl, alkaline phosphatase-PTH 338 pg/ml (15-65), 24 hr urinary calcium 424 mg. USG abdomen showed nephrocalcinosis, cholelithiasis, and no pancreatic calcifications. CECT chest revealed small enhancing lesion in anterior superior mediastinum, which was confirmed by TC99 sestamibi (Fig. 1).

Fig. 1: Radionuclide imaging of the parathyroid glands
Median sternotomy was done and pea nut size yellow colored tumor found, which was excised without difficulty. The histopathological examination showed the presence of a well circumscribed nodule comprising of highly cellular monomorphic tumor cells with round nuclei, abundant granular eosinophilic cytoplasm-chief cells arranged in acini (Fig. 2, 3). Post operatively her calcium levels normalized but parathormone levels failed to demonstrate adequate fall. Hypovitaminosis D was found (vit D-8ng/ml), accordingly replacement was advised. One month follow up showed normal calcium, phosphorus, alkaline phosphatase and parathormone levels.

**DISCUSSION**

Primary hyperparathyroidism with its non-specific varied manifestations and indolent course, poses a diagnostic challenge to the clinician [8]. Though uncommon, detection of ectopically located hyperfunctioning parathyroid gland is equally challenging and if done pre-operatively, is associated with successful surgical outcome.

Diagnosis of primary hyperparathyroidism in a clinically suspected case is suggested by hypercalcemia, hypophosphatemia, elevated bone specific alkaline phosphatase and intact parathormone levels [8].

During embryogenesis the superior and inferior parathyroid glands originate from the 4th and 3rd branchial clefts respectively and migrate caudally to their normal positions in relation to the thyroid gland. Any aberrancy during this descent may lead to ectopic location of these glands [8, 9]. The ectopic parathyroid glands are more often derived from inferior glands, because of their long migration path, providing them a higher probability of being ectopic from the angle of mandible to pericardium [10]. They may be located in the mediastinum either anterior or posterior, in the thymus (intra thymic), in the trachea esophageal groove and very rarely in the thyroid parenchyma [8, 9].

Pre operative 99m TC sestamibi scan helps in localizing the tumor in almost 90% of cases [11]. It has an important role for the localization of ectopic gland that helps the surgeon in order to plan the surgical approach as in our case. MIBI scintigraphy is one of the most widely used investigating technique for preoperative localization [4, 12]. MIBI localizes in both thyroid and parathyroid glands initially. On the delayed images, the MIBI washes off from the thyroid and the normal parathyroid glands allowing persistent radioactivity in the hyperfunctioning parathyroid gland [4].

MIBI is less sensitive in detecting hyperplastic parathyroid glands and also multigland disease [13].

**CONCLUSION**

We described a case of primary hyperparathyroidism due to parathyroid adenoma in mediastinum, which was excised using sternotomy.

Preoperative scintigraphy helped in confirming the location of the adenoma in our case and simplified the surgical management which would have been a challenging task otherwise.

**REFERENCES**


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