Brief Communication

A challenging case of an ectopic parathyroid adenoma

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ABSTRACT

The occurrence of ectopic parathyroid adenomas is not uncommon (3-4% of all parathyroid adenomas). A 42-year-old female diagnosed as having GH secreting pituitary adenoma presented with an ectopic mediastinal parathyroid adenoma located between left (Lt) pulmonary artery and Lt main bronchus. The aim of presenting this case is not to appreciate the rarity of the condition but to rather discuss some of the vital practical problems faced during its management. Patient presenting in endocrine OPD with nausea, vomiting, drowsiness and chronic constipation was investigated biochemically and with various imaging modalities and accordingly managed. Patient was also investigated from the perspective of MEN 1 syndrome. Results: Baseline routine investigations revealed hypercalcemia (corrected S. Ca- 16.9 mg/dl) due to primary hyperparathyroidism (PHP, PTH-1190 ng/L) with adenoma located between Lt main bronchus and Lt pulmonary artery. Patient was medically managed and after proper preoperative preparation, surgical excision by open thoracotomy was planned but two days before surgery she developed pulmonary embolism and was shifted to ICU where she died after 20 days. An accurate preoperative localization by various imaging procedures plays a decisive role in case of ectopic adenomas in the chest. Ectopic parathyroid adenomas are frequent cause of failed initial surgery. The best surgical approach to these ectopic adenomas is still controversial. Equally effective newer medical treatment modalities are also required in patients who are awaiting or are unfit for surgery. Lastly combination of MEN 1 with ectopic parathyroid adenoma is rare.

Key words: Parathyroid, ectopic adenoma, hypercalcemia

Ectopic parathyroid adenomas, although not so common, are occasionally seen in clinical practice. Here we discuss a case of a middle-aged female who presented with primary hyperparathyroidism (PHP) with ectopic parathyroid adenoma. The aim of presenting this case is not to appreciate the rarity of the condition but rather to discuss some of the vital practical problems faced during the management of ectopic parathyroid adenoma in the general population.

A 42-year-old female diagnosed with growth hormone (GH)-secreting pituitary adenoma was referred to the endocrine outpatient department (OPD) with hypercalcemia and hyperparathyroidism [parathyroid hormone (PTH): 1190 ng/mL]. The relatives complained of nausea, vomiting, and drowsiness on and off since 15 days. There was a two-year history of bone pains. There was no history of calcium disorders, kidney stones, fractures, osteoporosis, or ingestion of any drug that could be responsible for her hypercalcemia. Her family history was not contributory. She had already undergone a negative neck imaging. Physical examination including the neck was unremarkable. Routine laboratory investigations were normal except for severe hypercalcemia (corrected serum calcium: 16.9 mg/dL); phosphorous was 1.3 mg/dL. Urinary calcium was 879 mg/24 hours; 25-hydroxyvitamin D [25(OH)D] was 29 ng/mL. Hypercalcemia was urgently managed with fluid therapy, diuretics, and calcitonin.

Once the patient stabilized, a repeat neck ultrasound was done with a higher resolution but it failed to detect enlarged parathyroids. A technetium (99mTc) sestamibi scan was planned, considering the possibility of ectopic parathyroid adenoma which revealed a 2.5×1.5 cm sized lesion in the middle mediastinum just near the aorta and pulmonary trunk as shown in Figure 1. For further anatomical specification, a computed tomography (CT) angiogram was done which confirmed a 2.5×1.9 cm sized lesion between
the left main bronchus and left pulmonary artery [Figure 2a and b]. Fluorodeoxyglucose (18F) (18FDG) positron emission tomography (PET) and DOTANOC scans were done as a part of the multiple endocrine neoplasia-1 (MEN 1) workup but were negative for any neuroendocrine tumor. The patient was medically managed and after proper preoperative preparation, surgical excision by open thoracotomy with neck exploration for coexisting hyperplastic parathyroid glands was planned, but two days before surgery she developed pulmonary embolism and was rendered unfit for surgery and shifted to the intensive care unit (ICU).

In the ICU, hypercalcemia worsened and the patient developed acute kidney injury (serum creatinine: 4.9 mg/dL) with severe sepsis (total leukocyte count: 54,000 cells/dL with no blasts or band forms). Pamidronate was planned as an interim control measure but doses >45 mg/dL could hardly be tolerated in the face of the worsening renal function. We had also planned for bronchoscopic ethanol injection, but the deteriorating condition of the patient could not permit the same. Hemodialysis was undertaken but without much effect on the hypercalcemia. Finally the patient died after 15 days. The diagnosis provisionally remained ectopic parathyroid adenoma with the possibility of MEN 1 syndrome.

Ectopic parathyroid glands arising due to abnormal embryological migration are found in 1–3% of the population and these combined together account for 20–25% of the cases of parathyroid adenoma and clinically present as PHP. Majority of them are located in the anterior mediastinum near the thymus, but, rarely, some are present in the visceral compartment of the mediastinum or paraesophageal position or in the aortopulmonary window or close to the right pulmonary artery near the tracheal bifurcation which requires open thoracotomy or thoracoscopic removal.

The approach to ectopic parathyroid adenoma is faced with multiple challenges. First, the varied locations of ectopic glands make radiological support pivotal for accurate diagnosis. However, even the best of imaging modalities sometimes fail to identify the lesion accurately or even if successful, they consume a lot of time and expense. Second, neglecting the possibility of these ectopic adenomas results in a failed initial cervical exploration of PHP on several occasions. Accurate preoperative localization is the key to successful surgical removal. Coupling anatomical imaging like thin-slice contrast CT with functional scan (sestamibi) is most essential.

The third important aspect is the paucity of a systematic approach to the management of such lesions. Unfortunately, given the rarity of ectopic mediastinal parathyroid adenomas, existing guidelines do not include any official recommendations for ectopic parathyroid adenomas. Conditions wherein a conservative approach is preferred are poorly defined, apart from established indications like serious comorbidities and contraindications to surgery. No consensus exists for incidentally diagnosed asymptomatic ectopic parathyroid adenomas.

Another problem faced by clinicians managing such cases is the morbidity associated with various surgical approaches. The advent of modern imaging modalities, minimally invasive surgeries, and intraoperative PTH assays have rendered traditional cervical exploration obsolete. But even the most modern techniques are not completely devoid of debilitating thoracic and systemic complications which add a lot of morbidity to the prevailing clinical condition.
Medical management of PHP has not been satisfactory because no agents exist that can produce either sustained blockage of PTH release or suppress hypercalcemia. Hence, we need to expand our existing medical armamentarium for better control of hypercalcemia and thereby avoid its long-term complications.

Calcitonin, although safe and nontoxic, is relatively weak and is faced with tachyphylaxis. Although diuretics are also not completely devoid of their metabolic side effects, renal compromise precludes the use of fluid therapy, bisphosphonates, gallium nitrate, and plicamycin. Calcimimetics often tried in PHP fail to affect urinary calcium and bone mineral density (BMD) and hence are not medical surrogates of surgery. Conversely, denosumab although increasing BMD, does not affect serum or urinary calcium in PHP. Other strategies in the pipeline are specific vitamin D analog [paricalcitol, 22-oxa-calcitriol (OCT)]-induced selective PTH suppression, PTH receptor antagonism, and immunotherapy.

Therefore, in view of the restricted medical options, surgery still remains the best option. The potential for malignancy in ectopic parathyroid adenomas is unknown except in a few cases of parathyroid carcinoma in intrathyroidal parathyroid tissues which are managed surgically.

To conclude, the incidence of ectopic parathyroid adenomas is increasing with the increasing use of modern imaging modalities and we need to formulate a systematic approach to deal with such clinical scenarios.

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