Primary Hyperparathyroidism from an Ectopic Retrosternal Parathyroid Adenoma

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Abstract

Primary hyperparathyroidism is one of the common endocrine abnormalities. It is estimated that 0.3-8% of patients with primary hyperparathyroidism are originated from ectopic parathyroid adenomas(s). Medical management is recommended if patient is asymptomatic. However if the patient has clinical manifestations of bone disease, nephrolithiasis, hypophosphatemia, renal dysfunction, neuropsychological symptoms etc, a surgical intervention is guaranteed. Here we reported a patient presented with neuropsychological symptoms, possibly related to hypercalcemia induced by an ectopic parathyroid adenoma in a relatively rare location of retrosternum. After parathyroidectomy, patient’s neuropsychological symptoms resolved.

Keywords: Primary hyperparathyroidism; Ectopic adenoma

Introduction

The diagnosis of primary hyperparathyroidism is usually characterized by hypercalcemia and elevated Parathyroid Hormone (PTH) concentration or inappropriately elevated hypercalcemia with normal range of PTH [1]. It is estimated that 0.3 - 8 % cases of primary hyperparathyroidism originate from ectopic parathyroid adenoma(s) including the mediastinum, thymus, tracheoesophageal groove, thyroid, and behind the sternum, submandibular triangle, retropharyngeal space, carotid sheath etc [2,3]. Here we report the case of a 66-year-old male patient who presented with neuropsychiatric symptoms possibly secondary to an ectopic parathyroid adenoma in a rare retrosternal location. Surgical excision of the adenoma reversed the patient’s clinical symptoms.

Case Presentation

This is a 66- year- old gentleman with past medical history of diabetes mellitus type 2, hypertension and COPD who was traveling from Oregon to Florida. He had to stop by a county hospital in Texas due to severe shortness of breath. The O2 saturations were in the low 70s in the emergency department and patient was diagnosed with COPD exacerbation. He then required endotracheal intubation after failure of conservative treatments of oxygen, serial nebulizer treatments, antibiotics and steroids. The abnormalities in the initial laboratory studies included elevated bicarbonate of 34 meq/l (normal range 20 - 29 meq/l), hyperkalemia of 5.2 meq/l (normal range 3.4 - 4.9 meq/l), and hypercalcemia of 12.3 mg/dL (normal range 8.1-10.5 mg/dl). The subsequent studies showed elevated intact parathyroid hormone (PTH) of 376 pg/ml (normal range 15 - 65 pg/ml) and low 25-hydroxy vitamin D of 12 ng/ml (normal range 13 - 96 ng/ml). Primary hyperparathyroidism was diagnosed and patient was placed on the adequate normal saline intravenous hydration. The plan was to stabilize patient’s respiratory status, then discharge him home with observation and medical management. However, the patient developed a duodenal ulcer which perforated on day 7 of hospitalization. General surgery performed duodenectomy and antrectomy on the same day. Afterwards the patient developed multiple complications and had fluctuating mental status and depression. The serum calcium was slowly trending up with a peak of 15.5 mg/dl. Calcitonin and aggressive hydration was started followed by the addition of Sensipar. However, none of these had significant effect on...
the serum calcium level. The patient’s mental status was deteriorated which was likely related to uncontrolled hypercalcemia. Initial CT scan showed a substernum mass (Figure 1A-F). The sestamibi scintigraphy showed positive uptake from this unknown substernum mass (Figure 2). Overlapped CT and scintigraphy scan (SPECT) indicated that this is a 2.3 x 1.3 x 0.9 cm parathyroid adenoma in the retrosternal region behind the brachiocephalic vein, a very unusual location of the parathyroid gland (Figure 3A-D). An ENT specialist was consulted and decided to proceed with parathyroidectomy. The incision was made above the clavicular margin. After identification of both lobes of the thyroid, the upper mediastinum was inspected with a thoracic surgeon standby. Parathyroid adenoma was noted in the anterior superior mediastinum measuring about 3.5 cm in length. The blood supply was clipped and divided and the entire specimen was excised, and then submitted to pathology for frozen and permanent section. Frozen section pathology was consistent with parathyroid adenoma. The intra-operative PTH level dropped to 40.8 pg/ml instantly. Calcium slowly trended down in the first 12 hours postoperatively and the patient was placed on calcitriol 0.25 mcg twice daily. The corrected calcium level normalized 48 hours post operation. Meanwhile the patient’s altered mental status significantly improved.

Discussion

Though medical management is recommended for asymptomatic patients with primary hyperparathyroidism, surgical intervention of parathyroidectomy is still adequate in certain cases due to its benefit of curing the disease, decreasing the risk of renal stones, improving bone mineral density, and cost efficient etc [4-6]. Patients with symptomatic Primary Hyperparathyroidism (PHPT), ectopic or not, should have a parathyroidectomy. This patient presented with neuropsychiatric symptoms which led to the decision of surgical intervention. His mental status was significantly improved after the surgery. There are similar reports of symptoms relief after correction of hypercalcemia and/or elevated PTH level [7-13].

Although most ectopic adenomas can be removed by a cervical approach, a transthoracic dissection is necessitated when the adenoid tissues are located in a deep and complicated anatomic position in
the thoracic cavity. The first case of excision of ectopic mediastinal parathyroid adenoma was reported in an American sea captain who required 6 operations in 1932 [14]. Today, diagnosis and operation of ectopic parathyroid glands, especially mediastinal adenoma, continue to be a challenge. 99mTc-sestamibi scintigraphy are used to localize abnormal parathyroid gland(s) preoperatively which has led to a significant change in the operative approach for parathyroidectomy [15,16]. The sensitivity and specificity rates have reached 84.4% and 95.9% for sestamibi scintigraphy in a recent meta-analysis of 1297 patients by Castellani et al. [17]. In current report, the pre-operative sestamibi scan showed a 2.3 x 1.3 x 0.9 cm parathyroid adenoma located in anterior aspect of the left brachiocephalic vein. The intra-thoracic exploration was performed by an ENT and thoracic surgeon. They identified a 3.5 cm parathyroid adenoma in the anterior superior thoracic exploration was performed by an ENT and thoracic surgeon. In addition an intra-op PTH analysis showed more than 50% drop of PTH level which ruled out extra ectopic adenomas. The patient successfully recovered from surgery. Follow up serum calcium level normalized quickly after the excision of ectopic parathyroid gland.

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References