Primary Hyperparathyroidism Due to a Giant Parathyroid Adenoma: A Case Report

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Parathyroid adenomas account for most cases of primary hyperparathyroidism. Parathyroid adenomas rarely attain huge proportions and since they are usually small in size, preoperative localization using sestamibi scanning or ultrasonography is required to avoid unnecessary bilateral neck exploration. We report herein the case of a 47 year old woman who was diagnosed as having a parathyroid tumor after she presented with bone pain. Detected by clinical findings, ultrasonography and parathyroid scintigraphy by 99m Tc-MIBI, her tumor was found to be a huge adenoma of the parathyroid, measuring 5.5×3.5×2 cm and weighing approximately 30 grams. This size and weight of tumor is a very rare finding in parathyroid adenomas, and deserves documentation.

Keywords: Giant parathyroid adenoma, Scintigraphy

Introduction

After discovery of the parathyroid gland in 1850 and a later description of the gross histological appearance by Ivar Sandstrom, a Swedish student, early clinical observations suggested that parathyroid tumors are a compensatory effect for osseous abnormalities. Conversely, a theory considered that parathyroid tumors were primary occurrences and were responsible for secondary changes in the skeleton; this was confirmed 10 years later by Mendel, when he excised an enlarged parathyroid gland from a patient with hypercalcemia, hypercalciuria, and severe bone disease. Calused by one or more hyper functioning glands, the incidence of primary hyperparathyroidism (HPT) is approximately 25 per 100, 000 in the general population, and the incidence increases with age, being especially common in postmenopausal women.

Since these benign tumors are rarely evident on physical examination. Certain symptoms and biochemical abnormalities alert the surgeon to their presence. Solitary gland disease occurs in 80% of patients; it is usually a benign neoplasm or adenoma and rarely a parathyroid carcinoma. Giant parathyroid adenomas are very rare; the most frequent etiological association has been seen in survivors of the atomic bomb in Japanese studies. There are also some interesting non irradiation related case reports of huge parathyroid adenomas. Our case, in comparison was larger and heavier.

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Case Report

A 47 year-old woman, from the city of Gorgan, Iran, weight 45 kg and height 153 cm, presented at the endocrinology ward of Tehran’s Taleghani hospital with bone pain in the thoracic spinal column, extremities and the knee joints; for the past 6 months she had been having the pain, which had exacerbated since 2 weeks before hospitalization, when it was accompanied by nausea, vomiting, headache and obvious loss of weight (5 kg during 6 months). Her physical exam revealed otherwise normal findings, except for visible swelling on the right and middle part of the neck. The blood chemistry results were as follows: Serum calcium was 14.6 mg/dL with the normal range of (8.5-10.5), serum intact PTH (IRMA) was 1624 pg/mL, (normal range 10-65), serum phosphate, 1.8 mg/dL; (normal range 2.5-4.5) and serum alkaline phosphatase, 752 IU/L (normal range 37-147). Thyroid function tests revealed euthyroid status as follows: T3:1.1, T4:7.2 and TSH: 6.7, complete blood count was normal, and ESR was 52. Simple roentgenograms of the pelvis, chest, skull and hands revealed diffuse reduced bone density, some erosion in the distal phalanx of 2nd digit and somewhat subperiosteal bone resorption in radial aspect of middle phalanxes suggesting hyperparathyroidism (Fig.1).

The whole body scan showed hypertrophic osteoarthropathy of the long bones and "Hot Skull" was also seen, raising suspicions for a metabolic bone disease such as primary HPT. The neck ultra sonography revealed a heteroecho mass in the inferior portion of the right lobe of thyroid, measuring 20×48×30 mm, anatomically separated from thyroid (Fig.2).

Sonography of the kidneys, urethras and bladder was completely normal with no nephrolithiasis. Interestingly, without IV, the head and neck CT scans contrast were completely normal, showing no mass in the thyroid and parathyroid until finally a parathyroid scintigraphy, of a large area by 99 m Tc-MIBI revealed an abnormal collection of radiotracer on the right side of the neck, indicating a huge parathyroid adenoma, adjacent to the lower pole of the right thyroid lobe (Fig.3).

Fig.1. X-ray of the hand showing a brown tumor

Fig.2. Neck ultra sonography indicating a heteroecho mass measuring 20×48×30 mm in inferior portion of right lobe of thyroid, anatomically separated from thyroid
Fig. 3: Parathyroid scintigraphy, using 99m Tc – MIBI showing a large area of abnormal collection of radiotracer on right side neck, adjacent lower area of right thyroid, demonstrating a huge adenoma of the parathyroid.

Based on the above findings, the patient was transferred to the general surgery ward of the hospital for neck exploration and parathyroidectomy, and, following diagnosis of a parathyroid adenoma, surgery was performed. Using a collar neck incision, a large parathyroid tumor measuring approximately 5.5×3.5×2 cm and weighing approximately 30 g was found in the right retro-thyroid area (Figures 4 and 5).

There was no evidence of macroscopic malignancy, and the remaining glands appeared normal. The frozen section and permanent pathologic diagnosis were compatible with parathyroid adenoma (Fig. 6).
The patient was discharged 4 days after surgery without any complications; post-operation serum calcium was 8.7mg/dL, one month after discharge.

Discussion

Although primary hyperparathyroidism (HPT), in approximately 80% of cases, is caused by a single parathyroid adenoma, and rarely by multiple adenomas or hyperplasia, in 1-2% of cases the cause could be carcinoma. Based on the pathophysiology of primary hyperparathyroidism, increased secretion of PTH promotes a release of calcium by inhibition of osteoblasts and stimulating osteoclasts. In the kidneys, PTH causes a decrease in calcium clearance, and increases renal excretion of phosphate by inhibiting its reabsorption in the tubules. PTH stimulates hydroxylation of 25-hydroxy vitamin D to 1, 25-dihydroxy vitamin D in the kidneys; it is the latter which causes enhanced absorption of calcium in the intestine. In the last century, the typical presentation of primary HPT has changed from a severe, debilitating disease towards a disease with subtle symptoms and physiologic derangements. Some common symptoms and their prevalence in these patients are as follows: Nephrolithiasis (30%), bone disease (2%), peptic ulcer disease (12%), psychiatric disorders (15%), muscle weakness (70%), constipation (32%), polyuria (28%), pancreatitis (1%), myalgia (54%) and arthralgia (54%); the clue to the diagnosis of primary hyperparathyroidism is usually the laboratory finding of hypercalcemia. The diagnosis of primary hyperparathyroidism rests on the laboratory confirmation of increased serum calcium levels and inappropriately elevated intact parathyroid hormone concentrations. The main reason is that the disease has no specific symptoms and only a few cases present with clinical entities such as nephrolithiasis, osteoporosis-osteopenia, pancreatitis, hypertension, peptic ulcer disease, depression, etc. If surgical intervention is planned, cervical ultrasonography and parathyroid-scintigraphy are indicated for the exact localization of hyper functioning parathyroid gland(s). As mentioned in our case, despite the massive proportions of the adenoma, it did not result in gross clinical findings, other than some bone pain and arthralgia, which are common in primary HPT. CT scans and/or MRI are usually not necessary, except in cases of prior neck operations; the definitive cure of the disease can be achieved only by surgical means. Several approaches are described ranging from minimally invasive parathyroidectomy to standard bilateral neck exploration with a success rate of curing hypercalcemia of around 98%. The usual weight of a parathyroid adenoma ranges from 70 mg to 1 g, although there are sporadic reports of tumors weighing more than 30 g. Searching pubmed, of cases of giant parathyroid adenomas, 6 cases were not available; we found 16 cases of parathyroid adenoma, weighing more than 30 grams. In 2005, Power et al. described a super huge adenoma of parathyroid measuring 8×5×3.5 cm, weighing 110 g in an 85 year-old woman; despite its huge size, it had not caused many of the hypercalcemic symptoms usually associated with larger adenomas. In 1993, Tsuchia et al. reported a 56- year-old woman, with hip joint pain and gait disturbances; she had a large parathyroid adenoma weighing approximately 70 g. In another 2004 case report, Younes et al. describe a 48-year-old man, with double parathyroid adenomas associated with brown tumor in lower limb. The left superior gland was 15gm in weight and the left inferior gland was 920 mg in weight. In 2006, Kammori, et al. reported a 59-yr-old man, who presented with episodes of increasingly severe, left arm bone pain, due to “brown tumor” and a giant mediastinal cystic parathyroid adenoma, measuring 12×7×1.7-cm and weighing 116 g; however in comparison to ours, most operated parathyroid adenomas, reported so far were smaller and weighed less.

Documented evidence shows that adenoma weight has direct correlation with preoperative serum calcium and PTH and is inversely
related to serum phosphate level; there is also higher incidence of post operative transient hypocalcemia and lower mean serum Ca level on days 3-4, following the surgery in cases of larger adenomas. In our case, preoperative levels of serum PTH and calcium were definitely higher than normal. Some studies reported a higher risk of death in patients with larger adenomas. Fortunately, our patient was diagnosed and referred early for surgery in time and is still alive, with optimum follow up results. Hence increased adenoma weight may be a valuable aid in identifying those patients in early stages of primary hyperparathyroidism at risk for transient hypocalcemia after neck exploration and surgical treatment. Since giant parathyroid adenoma patients present with non-characteristic symptoms, routine laboratory measurements including serum calcium and phosphorous should be conducted for suspected individuals. Our case was that of a patient with a 30 gram parathyroid adenoma, presenting with no significant signs or symptoms but laboratory findings and nuclear study confirmed the diagnosis.

In conclusion, primary hyperparathyroidism should be kept in mind in all patients presenting with longstanding history of bone problems, from simple bone pain to severe bone disease and hypercalcemia. Elevated serum level of PTH confirms the diagnosis.

References