Unusual Ectopic Parathyroid Adenoma: A Case Report

Habibeh Taghavi Kojidi, Nazanin Vagharimehr, Shahrzad Mohseni, Mohammad Pajouhi, and Mohammad Reza Mohajeri-Tehrani

Endocrinology and Metabolism Research Center, Endocrinology and Metabolism Clinical Sciences Institute, Tehran University of Medical Sciences, Tehran, Iran

Received: 09 Jul. 2014; Accepted: 25 Dec. 2014

Abstract- An ectopically placed parathyroid in the mediastinum is a rare cause of persistent or recurrent primary hyperparathyroidism. They are rarely in a huge size. We report a case of a 70-year-old man with a history of total parathyroidectomy and thymectomy presented with a lack of appetite, nausea, and generalized bone pain, polydipsia and a calcium level of 14.4 mg/dl. ^{99m}Tc-sestamibi scintigraphy with single-photon emission computerized tomography (SPECT) showed a focal zone of radiotracer accumulation in the midline of anterior chest wall (xiphoid level). The mass excised from our patient surgically was 75 grams. This weight and location of the tumor is a very rare finding in parathyroid adenomas.

Acta Med Iran, 2016;54(8):547-550.

Keywords: Giant ectopic parathyroid adenoma; Recurrent primary hyperparathyroidism

Introduction

Primary hyperparathyroidism (PHPT) is a prevalent endocrine disorder affecting mostly middle-aged women, and its incidence is approximately 25 per 100,000 in the general population that increases with age (1).

PHPT is classified as being caused by a single adenoma (75%-85%), hyperplasia (10%-20%), multiple adenomas (4%-5%) or carcinoma (1%) giving the histopathological characteristics and the number of involved glands (1).

Operator inexperience aside, the usual cause of the persistent disease is asymmetric parathyroid hyperplasia or ectopic parathyroid tissue that the latter includes intrathyroidal, undescended, gastroesophageal and mediastinal glands (2). Up to one in five parathyroid glands may be ectopic and this is especially true of supernumerary glands. Disease occurring after 6 or 12 months of normocalcemia is called recurrent hyperparathyroidism and varies in incidence from 2% to 16%. The cause usually is unresected hyperplastic glands, but it may be caused by parathyroid carcinoma, a second adenoma or inadvertent local seeding of parathyroid tissue into the neck during previous parathyroid surgery which is called parathyromatosis (3,4).

The benign tumors are not always evident in physical examination and need specific biochemical alterations and symptoms to be diagnosed. Giant parathyroid that may be easier to diagnose are not very common on the other hand, and the most frequent etiological association is found in atomic bomb survivors of Japanese studies. Some studies also shown some none irradiation related huge parathyroid adenomas (5).

Ultrasonography (US), computed axial tomography, magnetic resonance imaging, and ^{99m}Tc-sestamibi scintigraphy is most common imaging tests for detecting abnormal parathyroid glands and ^{99m}Tc-sestamibiscintigraphy with single-photon emission computerized tomography (SPECT) has the highest sensitivity. One benefit of this test over the US is its ability to diagnose ectopic mediastinal parathyroid glands, but the US is less costly and radiations free (1).

The physiological asymmetric activity of submandibular glands, the presence of thyroid nodules, multinodular goiter, breast, lung, head and neck carcinomas and their lymph node, bronchial carcinoids, differentiated thyroid malignancies and PTH-secreting paraganglioma are false positive results with ^{99m}Tc-sestamibiscintigraphy. Thyroid nodules and multinodular goiter are ruled out with 123I or ^{99m}Tc-sestamibi scintigraphy subtraction test (1,6).

Parathyroid hyperplasia, multiple parathyroid

Corresponding Author: M.R. Mohajeri-Tehrani
Endocrinology and Metabolism Research Center, Endocrinology and Metabolism Clinical Sciences Institute, Tehran University of Medical Sciences, Tehran, Iran
Tel: +98 21 88220037, Fax: +98 21 88220052, E-mail address: mrmohajeri@tums.ac.ir

adenomas, calcium channel blockers, the small size of the adenoma, superior position and paucity of oxyphilic cells are false negative results with ^{99m}Tc-sestamibi scintigraphy (6).

Definite cure hypothesis for PHPT is parathyroidectomy and is recommended for symptomatic patients with skeletal and renal problems (1).

Localizing the parathyroid tissue in the neck or at the arm before surgery is one of the difficulties (7).

Case Report

A 70-year-old man with a lack of appetite, nausea and lower limbs pitting edema was admitted to the emergency ward of Shariati University Hospital. Lab findings demonstrated calcium level of 14.4 mg/dl. Other symptoms were constipation, generalized bone pain, and polydipsia which were worsened in the past 6 months. There was no history of exposure to radiation, and there were no weight loss and consciousness alteration. He had a history of parathyroidectomy and renal stone surgery 36 years and 20 years ago respectively. He also had another surgery 18 years ago due to hypercalcemia in which right lobectomy of the thyroid was performed. There was no evidence of parathyroid tissue in the neck area. Midsternotomy and thymectomy were done consequently, and resection of intra-thymus parathyroid adenoma was done, and pathology results confirmed the diagnosis reporting: parathyroid adenoma (10 g) with a predominance of chief cell types, no oxyphilic cell. Some of the tissue was inserted in patient's left arm. He had hypocalcemia postoperatively that was treated with intravenous calcium subsequently with oral calcium and Rocaltrol® for 4 weeks.

At the time of admission, the patient was pale, and several nodules were palpated in the left thyroid lobe, and pitting edema of lower limbs was seen. Laboratory findings are shown in the (Table 1). Serum calcium was elevated (14.4 mg/dl), and PTH level was 930 pg/ml.

24h Urine calcium and Cr were 341 mg and 1100 mg respectively.

Table 1. Patient's laboratory findings

Test	Result	Normal range
WBC	8300/mm^3	4500-1100/mm^3
Hb	9.5 g/dl	13.5-17.5 g/dl
Plt	153000/mm^3	150000-400000/mm^3
ESR	25 mm/h	0-15 mm/h
BUN	45 mg/dl	7-18 mg/dl
Cr	3.3 mg/dl	0.6-1.2 mg/d
TSH	4.2 µU/ml	0.5-5 µU/ml
T4	8.5 µg/dl	5-12 µg/dl
PTH	930 pg/ml	10-65 pg/ml
Ca	14.4 mg/dl	8.4-10.2 mg/dl
25(OH) vitamin D	62 ng/ml	30-100 ng/ml

During treatment with normal saline, furosemide and zoledronic acid (4 mg) intravenously, calcium levels decreased to 10.7 mg/dl and BUN and creatinine levels went down to 22 mg/dl and 1.7 mg/dl respectively.

In the neck ultrasound, multiple various sizes iso-echo nodules without microcalcification, no lymphadenopathy, and no parathyroid gland was seen, and the left arm ultrasound showed no parathyroid tissue. Multiple bilateral various sizes large stones in both renal systems severely in right one were seen in abdominal ultrasonography. Bone mineral density results are shown in (Table 2).

Table 2. Bone mineral density results of the patient

BMD	T- score	Z- score
Lumbar spine	-3.7	-2.6
Femoral neck	-4.2	-2.6

Fine needle aspiration (FNA) of thyroid nodules showed adenomatous colloid goiter. No malignancies were detected.

^{99m}Tc-sestamibiscintigraphy with SPECT parathyroid scan showed a focal zone of radiotracer accumulation in the midline of anterior chest wall (xiphoid level) (Figure 1)

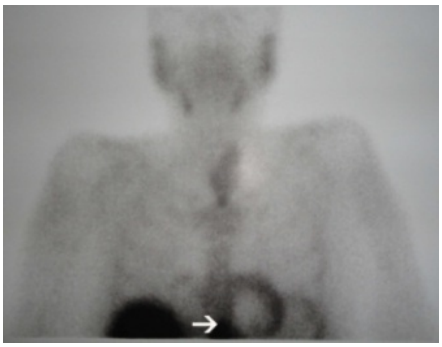

Figure 1. Sestamibi scan showing a large area of abnormal collection of radiotracer on xiphoid level

Spiral mediastinal computed tomography (CT) with contrast revealed soft tissue density mass with mild enhancement about 51 30 mm in anterior midline on the xiphoid level (Figure 2).

The mass was surgically removed, and pathology results confirmed active parathyroid lesion with mitotic activity in 5-10% of cells.

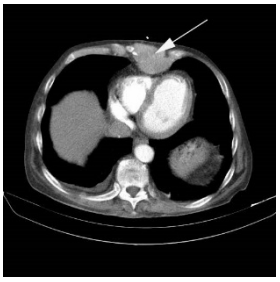

Figure 2. Computed tomography scan indicating soft tissue density mass on xiphoid level

Discussion

Approximately 80% of primary hyperparathyroidism is caused by a single parathyroid adenoma; 1-2% of cases are caused by carcinoma, and rarely by multiple adenomas or hyperplasia (5).

Our case had three recurrences episodes of hyperparathyroidism and all of them were in different origins and not related to each other. The latest one was at the midline of anterior chest wall on the xiphoid level.

The common weight of parathyroid adenoma varies between 70 mg to 1 g, but some sporadic cases of tumors were reported to be more than 30 g. Parathyroid tumor weighing more than 3.5 grams is referred to as" giant tumor" and adenoma weight has a direct relation with serum calcium and PTH levels and indirect relation with serum phosphate levels. A transient hypocalcemia and lower mean serum calcium level on day 3-4 post operation is commonly seen in cases of larger parathyroid adenomas (5,8). Ectopic parathyroid glands can be located at the aortopulmonary window, so for better recognition ^{99m}Tc-sestamibi SPECT/CT is a choice, and it can follow by thoracic and abdominal CT or MRI (1).

The mass excised from our patient was 75 g and according to the definition is considered giant adenoma (8). Also, the location of the mass was unusual.

Several findings help to distinguish parathyroid adenomas from parathyroid carcinomas. Adenomas usually have yellowish-brown color and have a round or oval shape and are soft in consistency whereas carcinomas usually have a grayish-white color capsule, are lobulated, firm to stony consistency and show gross infiltration of adjacent tissue. Their metastases sites include lungs (40%), cervical nodes (30%), and liver (10%). Metastases to bone and pleura, pericardium and pancreas have been reported occasionally (9).

(Table 3) shows the different features of parathyroid carcinomas and parathyroid adenomas (9).

Table 3. Comparison of different features of parathyroid carcinomas and adenomas

	Parathyroid carcinoma	Parathyroid adenoma
Female: male	1:1	3.5:1
Average age (year)	48	55
Asymptomatic	<5%	>80%
Serum calcium (mg/dl)	>14	=<1 above normal upper limit
PTH	Markedly elevated	Mildly elevated
Palpable neck mass	common	rare
Renal involvement	32-80%	4-18%
Skeletal involvement	34-91%	<5%
Concomitant skeletal and renal disease	Common	rare

Our case is unusual for a benign adenoma with PTH, Ca, and weight (75 g) that are all suggestive of parathyroid carcinoma. There was no invasion, metastases and no other features suggestive of malignancy. The patient recovered well postoperatively, with PTH level falling to 75 pg/ml and Ca 8.7 mg/dl on the postoperative day 4.

There were no signs of metastasis and the pathology confirmed the mass to be benign.

Serum calcium and PTH decreased dramatically after surgery. Laboratory test showed no abnormality after six months.

References

1. Gouveia S, Rodrigues D, Barros L, Ribeiro C, Albuquerque A, Costa G, et al. Persistent primary hyperparathyroidism: an uncommon location for an ectopic gland – case report and review. Arq Bras Endocrinol Metab 2012;56:393-403.
2. Akerström G, Rudberg C, Grimelius L, Johansson H, Lundström B, Rastad J. Causes of failed primary exploration and technical aspects of reoperation in primary hyperparathyroidism. World J Surg 1992;16:562-9.
3. Weber CJ, Sewell CW, McGarity WC. Persistent and recurrent sporadic primary hyperparathyroidism: histopathology,complications and results of reoperation. Surgery 1994;116:991-8.
4. Kollmorgen CF, Aust MR, Ferreiro JA, McCarthy JT, van Heerden JA. Parathyromatosis:a rare yet important cause of persistent or recurrent hyperparathyroidism. Surgery 1994;116:111-5.
5. Salehian MT, Namdari O, Mohammadi SS, Fazaeli Yousofabad H. Primary Hyperparathyroidism Due to a GiantParathyroid Adenoma: A Case Report. Int J Endocrinol Metab 2009;2:101-5.
6. Eslamy HK, Ziessman HA. Parathyroid Scintigraphyin Patientswith Primary Hyperparathyroidism: ^{99m}Tc Sestamibi SPECT and SPECT/CT. Radiographics 2008;28:1461-76.
7. Chou F, Lee C, Chen H, Chen JB, Hsu KT, Sheen-Chen SM. Persistent and recurrent hyperparathyroidism after total parathyroidectomy with autotransplantation. Ann Surg 2002:235:99-104.
8. Christakis I, Michas S, Mouziouras V, Augoustis C, Dimas S. Minimally Invasive Techniques for Giant Parathyroid Tumours; An oxymoron? Hellenic J Surg 2012;84:220-4.
9. Shane E. Clinical review 122 parathyroid carcinoma. J Clin Endocrinol Metab 2001;86:485-93.