Cystic Parathyroid Adenoma: An Unusual Cause of Hypercalcemic Crisis

Kistik Paratiroid Adenomu: Hiperkalsemik Krizin Nadir Bir Nedeni

Abstract

Parathyroid crisis is a rare clinical entity characterized by life-threatening hypercalcemia of a sudden onset in patients with primary hyperparathyroidism (pHTP) and rarely results from a cystic adenoma. We describe a 53-year-old woman with acute hypercalcemic crisis who presented with severe gastrointestinal symptoms. She had severe hypercalcemia (calcium level: 18.6 mg/dl) in conjunction with an elevated parathyroid hormone level of 1063 pg/ml. Ultrasonography of the neck revealed a 25x15 mm partly cystic mass at the superior pole of the right thyroid lobe. The patient underwent surgery immediately because hypercalcemia could not be controlled with medical treatment. The pathologic diagnosis was a cystic parathyroid adenoma. Turk Jem 2014; 2: 64-66

Key words: Primary hyperparathyroidism, cystic adenoma, hypercalcemic crisis

Özet


Key words: Primer hiperparatiroidi, kistik adenom, hiperkalsemik kriz

Introduction

Patients with primary hyperparathyroidism (pHTP) usually present with asymptomatic hypercalcemia. In contrast to this presentation, 1%-2% of the patients, may present with an unusual and life-threatening form, parathyroid crisis [1]. Patients with this form are symptomatic and have extremely high serum calcium (>15 mg/dl) and PTH levels [2].

Parathyroid adenoma is the most common cause of pHTP, usually a solitary single adenoma occurs in approximately 80% of the patients [3]. Rarely, cystic parathyroid lesions may cause pHTP. Cystic lesions of parathyroid glands are uncommon, they account for a small proportion (1%-4%) of all parathyroid adenomas [4,5,6]. Here, we describe a patient with solitary parathyroid adenoma containing a giant cystic area who presented with acute hypercalcemic crisis.

Case Report

A 53-year-old female patient was admitted to the hospital with nausea and vomiting. She complained of progressive fatigue, generalized body aches, arm muscle weakness and heartburn for last four months. For a few days, she developed polyuria, increased thirst, constipation and severe vomiting. She had a history of acid peptic disease, kidney stone and osteoporosis. She had undergone cholecystectomy three months ago. On physical examination, she was conscious and oriented, a nodule was palpable at the right pole of the thyroid gland. Neurologic...
examination revealed muscle weakness (muscle strength: 4/5).
Laboratory examinations were as follows: serum calcium: 18.6
mg/dL (8.4-10.2), serum phosphorus concentration: 2.7 mg/dL
(2.3-4.7), serum albumin concentration: 3.7 gr/dL (3.5-5), blood
urea concentration: 25 mg/dL (15-43) and potassium: 2.8 mmol/L
(3.5-5). Her serum intact PTH (parathormone) level was 1063.7 pg/
ml (15-68.3). Thyroid hormone levels were as follows: free T3: 2.8
pg/ml (n=1.71-3.71), free T4: 1.4 ng/dl (n=0.7-1.48), and TSH: 0.28
µIU/ml (n=0.35-4.94). Her 24-hour urine calcium was 380 mg/dl
(100-300). Her symptoms and the laboratory findings suggested
parathyroid crisis. She was immediately treated with intravenous
fluids, furosemide and calcitonin.
Ultrasonography of the thyroid and parathyroid glands revealed
a 25x15 mm hypoechogenic solid lesion in the right lobe of the
thyroid, with an eccentric cystic area. A marked increase in blood
flow in comparison to the thyroid parenchyma was observed on
colored Doppler ultrasonography. An iso-hypoechogenic solid
nodule measuring 12x7 mm was observed in the left lobe at the
level of the isthmus intersection. In addition, a cystic nodule 4x2
mm in diameter was observed in the anterior part of the left lobe.
An isoechogenic solid nodule with a diameter of 8x6 mm and
with a calcific center was observed in the inferior part of the right
lobe (Figure 1).
Technetium-99m-sestamibi SPECT revealed an asymmetric uptake
in the superior aspect of the right thyroid lobe (Figure 2). Thyroid
scintigraphy showed toxic multinodular goiter.
After we had reduced the serum calcium concentration to 12.3 mg/
dl, she underwent total thyroidectomy and parathyroidectomy.
Pathological examination revealed a parathyroid adenoma
with cystic degeneration (Figure 3). After the surgery, her serum
iPTH and calcium values decreased to 1.5 pg/mL and 7 mg/dL,
respectively. Her serum calcium concentration was maintained
with enteral and parenteral calcium and calcitriol. She required
intravenous calcium substitution until the tenth postoperative
day. She was discharged on the 12th postoperative day taking
oral calcium and calcitriol supplements. Calcium and calcitriol
supplementation was gradually reduced and eventually
discontinued after six months. Serum calcium, phosphate, and
PTH concentrations were all within the reference ranges.

discussion
This case highlights the rare presentation of parathyroid cystic
adenoma presenting with parathyroid crisis. Cystic lesions of the
parathyroid glands, whether functioning or nonfunctioning, are
uncommon, and rarely cause parathyroid crisis (7).
Parathyroid cysts have been described in several surgical reports
(4,7,8,9). In a recent study, cystic parathyroid lesions were found in
48 of 1769 patients (3%). Functional parathyroid cysts were more
common than nonfunctional parathyroid cysts, arising in 41 of
48 patients (85%) (5). In another study, more than 1700 patients
were operated on for primary hyperparathyroidism. The authors
presented six patients with cystic degeneration of a parathyroid
gland causing pHPT in five patients (8).
Functional parathyroid cysts commonly become evident with
symptoms of pHPT, such as fatigue, depression, osteoporosis,
nephrolithiasis, and rarely, they present with parathyroid crisis.
In a surgical report, eleven parathyroid cysts were found in 325
patients who underwent parathyroid operations. Two patients had

Figure 1. a) Sagittal and b) transverse ultrasound images of thyroid
gland show solid parathyroid adenoma in the right lobe of thyroid, with an
eccentric cystic area (line arrowpointing to solid portion of the parathyroid
adenoma and dashed arrow pointing to an eccentric cystic area)

Figure 2. Technetium-99m-sestamibi SPECT shows an asymmetric
uptake in superior aspect of right thyroid lobe
a) Early 3D sestamibi SPECT image b) Delayed 3D sestamibi SPECT image

Figure 3. Histopathological findings showed a parathyroid adenoma with
cystic changes (arrow pointing to the cystic area) (H&EX40)
acute parathyroid crises, which required emergency operations (9). Rapidly growing large-sized adenomas are more commonly associated with marked hypercalcaemia. In our patient, the size of the cystic adenoma was 25x15 mm. Wang and Guyton reported that all the patients presenting with parathyroid crisis in their series had large parathyroid masses, measuring from 3 to 7 cm. (10). While 12 patients were diagnosed with parathyroid adenoma, 1 patient was diagnosed with parathyroid carcinoma and 1 patient was diagnosed with parathyroid hyperplasia. Hypercalcemic crisis is a rare, but potentially fatal complication of hyperparathyroidism and should be treated urgently. It is characterized by severe hypercalcaemia (>12.5 mg/dL) associated with signs and symptoms of multi-organ failure (1,2). Although most cases of severe hypercalcaemia are seen in malignancy, severe hypercalcaemia with raised PTH is pathognomonic for primary hyperparathyroidism. Patients with parathyroid crisis demonstrate gastrointestinal and neurological symptoms, renal failure and cardiac rhythm abnormalities. Our patient had very high serum calcium level of 18.6 mg/dL. She exhibited gastrointestinal complaints, such as nausea, vomiting and constipation.

Acute management of parathyroid crisis includes treatment with saline solutions, furosemide and biphosphonates. Early removal of the mass has been recommended in these patients (10). In our patient, when serum calcium level decreased to 12.3 mg/dl, she was operated on. The symptoms improve immediately in patients who undergo surgical treatment. The serum concentrations of calcium and PTH return to normal in most patients within one to four days after parathyroid surgery. Hypocalcaemia may develop after the removal of all the parathyroid tissue, after an ischaemic injury, or long-term suppression of the parathyroid tissue. In our patient, serum iPTH and calcium values decreased in the first day and hypocalcaemia occurred in the first week after the surgery. She required intravenous calcium substitution until the tenth postoperative day and was maintained oral calcium and calcitriol supplementation for six months.

Conclusion

Cystic lesions of parathyroid glands may present with hyperparathyroid crisis. We presented a rare case of parathyroid crisis secondary to a cystic adenoma, with very high serum calcium and iPTH levels.

Conflicts of Interest

There are no conflicts of interest.

References