Case Report

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Cystic Parathyroid Adenoma: An Unusual Cause of Hypercalcemic Crisis

Kistik Paratiroid Adenomu: Hiperkalsemik Krizin Nadir Bir Nedeni

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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

Paratiroid krizi, primer hiperparatirodizmi (PH) olan hastalarda hayatı tehdit eden hiperkalsemi ile ani olarak ortaya çıkan bir klinik durumdur ve nadiren kistik adenoma bağlı olarak görülür. Bu yazıda şiddetli gastrointestinal semptomlar ile başvuran akut hiperkalsemik kriz tanısı koyduğumuz 53 yaşında bir olgu sunulmuştur. Hastada ciddi hiperkalsemi (kalsiyum seviyesi, 18,6 mg/dl) ile birlikte parathormon seviyesi 1063 pg/ml olarak yükselmiş bulundu. Boyun ultrasonografisinde sağ tiroid lobunun üst kısmında 25x15 mm boyutunda bir kısmı kistik kitle lezyonu saptandı. Hiperkalseminin ilaç tedavisi ile kontrolü sağlanamadığı için acil olarak cerrahi operasyon uygulandı. Patolojik tanı kistik paratiroid adenomu olarak konuldu. *Turk Jem 2014; 2: 64-66*

Key words: Primer hiperparatiroidi, kistik adenom, hiperkalsemik kriz

Introduction

Patients with primary hyperparathyroidism (pHPT) 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 pHPT, usually a solitary single adenoma occurs in approximately 80% of the patients (3). Rarely, cystic parathyroid lesions may cause pHPT. 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

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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 anasymmetric 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 pHTP, 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

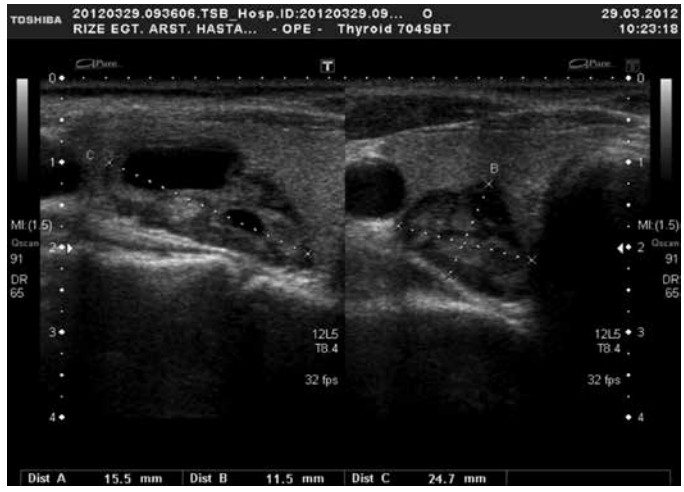

Figure 1. a) Sagittal and b) transverse ultrasound images of thyroid gland show solid parathyroid adenomain the right lobe of thyroid, with an eccentric cystic area (line arrowpointing tosolid portion of the parathyroid adenoma and dashed arrow pointing to an eccentric cystic area)

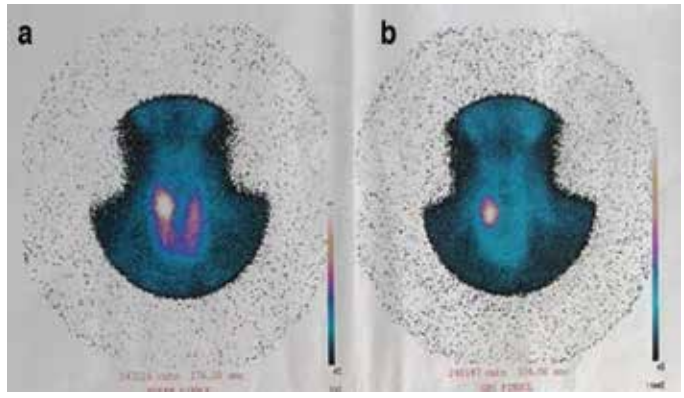

Figure 2. Technetium-99m-sestamibi SPECT shows an asymmetric uptake in superior aspect of right thyroid lobe
a) Early 3D sestamibi SPECT image b) Delayed 3D sestamibi SPECT image

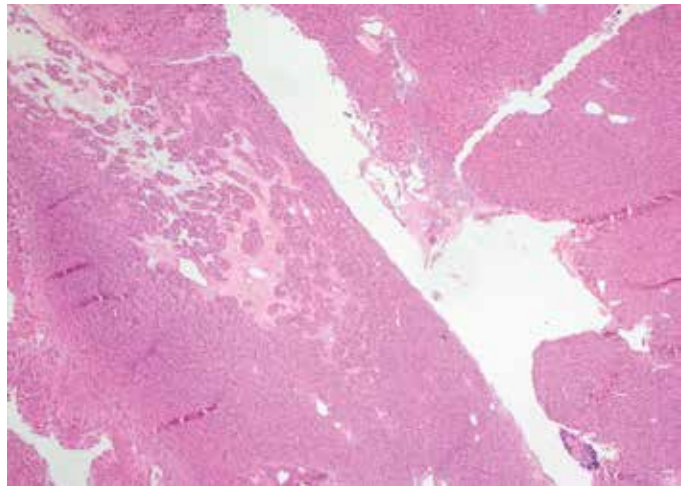

Figure 3. Histopathological findings showed a parathyroid adenomawith cysticchanges (arrow pointing to the cystic area) (H&EX40)

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 hypocalcemia 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 hypercalcemic 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.

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