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Review Article

Primary Hyperparathyroidism with Parathyroid Carcinoma

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Abstract

Parathyroid carcinoma is a rare endocrine malignancy, accounting for less than 1% of cases of primary hyperparathyroidism. We report the case of a female patient hospitalized for acute kidney failure associated with hypercalcemia, parathyroid hormone levels 8 times above the normal value, and a nodular mass behind the thyroid gland on magnetic resonance imaging. The patient underwent en bloc resection of the parathyroid glands and the right thyroid lobe.

Keywords: carcinoma; parathyroid; primary hyperparathyroidism; renal failure; calcium

Introduction

Parathyroid carcinoma is a rare endocrine malignancy, accounting for less than 1% of cases of primary hyperparathyroidism with an indolent but progressive behavior (1) and can constitute an endocrine emergency when it presents with severe hypercalcemia, as in the present case.

We report the case of a 54-year-old female patient who sought medical care due to asthenia, dyspepsia, vomiting and weight loss (6 kg) within the past 6 months.

On admission, the laboratory tests showed creatinine levels of 1.95 mg/dL, BUN 99 mg/dL, hypercalcemia 11.2 mg/dL and parathyroid hormone 842 pg/mL. The patient was admitted with a diagnosis of acute kidney failure. A contrast-enhanced computed tomography (CT) scan of the abdomen and pelvis reported lytic lesions in both iliac bones that were not present in a previous scan performed in 2011. A technetium-99m (Tc-99m)-sestamibi parathyroid gland scintigraphy showed diffuse increased uptake in the right thyroid gland (Fig. 1). She was treated with intravenous fluids and bisphosphonates, with partial recovery. An ultrasound of the neck showed a 1-mm solid lesion in the right thyroid lobe; fine needle aspiration (FNA) was unsatisfactory for diagnosis. Calcium levels remained elevated (13.5 mg/dL) with ionized calcium of 1.92 mmol/L, 25-OHvitamin D3 of 18.70 ng/mL, calciuria of 285 mg/24 hours and parathormone (PTH) of 1036 pg/ mL. A magnetic resonance imaging (MRI) of the neck and thorax reported a rounded lesion of 10 × 15mm, with well-defined borders, behind the right thyroid lobe (Fig. 1.B). With a presumptive diagnosis of malignant hypercalcemia versus metastatic lesions, a CT-guided percutaneous biopsy of the lytic lesions in the iliac bone was performed, which confirmed osteitis fibrosa cystica consistent with hyperparathyroidism.

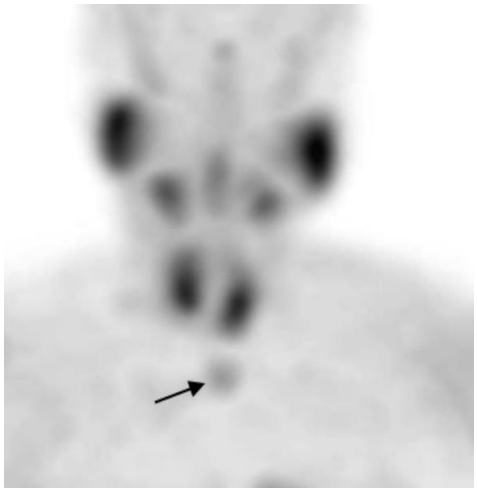

Figure 1. A Tc-99m-sestamibi parathyroid gland scintigraphy in the anterior projection showing high uptake area in the right thyroid lobe (small black arrow).

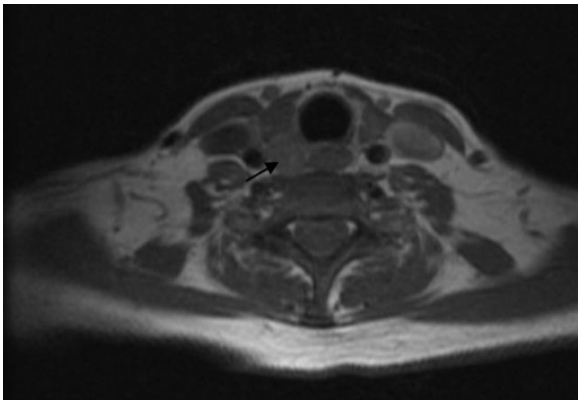

Figure 1 B. MRI of the neck, sagittal section. A 10 × 15 mm mass is observed behind the thyroid gland (small black arrow).

The patient underwent surgical exploration with en bloc resection of the parathyroid glands and the right thyroid lobe (Figure. 2). The pathological examination reported the presence of parathyroid carcinoma. Postoperative PTH level was 6 pg/mL.

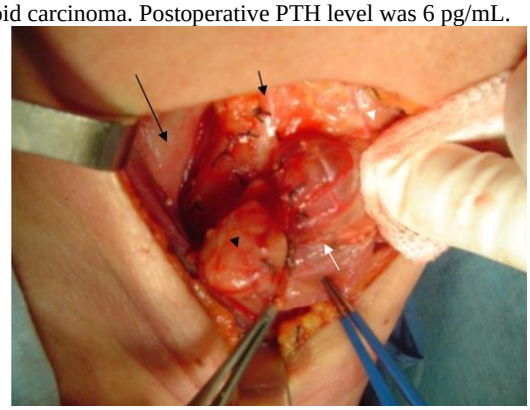

Figure 2: Bloc resection of the parathyroid glands and the right thyroid lobe. Recurrent laryngeal nerve (small black arrow); parathyroid gland (black arrowhead), internal jugular vein (long black arrow), thyroid gland (white arrow) and trachea (white arrowhead).

The patient evolved with a favorable outcome. One month after surgery, the laboratory tests showed PTH 71.6 pg/mL, calcium 9.5 mg/dL and ionized calcium 1.11 mmol/L. So far, these tests have remained within normal levels and ultrasound scans are normal.

Of 183 patients with primary hyperparathyroidism undergoing surgery in the Department of Head and Neck Surgery of Hospital Universitario Austral between 2001 and 2020, 84% were adenomas, 13.1% had hyperplasia of the parathyroid glands, 0.7% (3 cases) were atypical adenomas and one case corresponded to a parathyroid carcinoma (0.5%). In 19 patients the resection of the parathyroid glands was associated with surgery of the thyroid gland.

Parathyroid carcinoma is a rare cause of hypercalcemia associated with very high PTH levels. Unlike benign tumors, it is not more common in women and occurs in younger patients (5th decade of life); symptoms are

nonspecific and has slow growth, which explains why it is usually diagnosed later (1).

The clinical manifestations are due to the overproduction of PTH that can reach levels 10 times above the normal value (2). Most patients present signs of renal and bone involvement at the time of diagnosis, as in this case. Other symptoms include anorexia, nausea, vomiting, abdominal pain, peptic ulcer, weakness, myalgias, arthralgias, pancreatitis, weight loss and fatigue (3). On some occasions, patients may present with a parathyroid crisis with calcium levels >16 mg/dL, a clinical event that is difficult to manage.

A palpable neck mass is a common finding (40- 70%) and 15-30% of cases (4) have cervical lymph node metastases. Distant metastases are unusual in the early stages; the most common sites are lung (40%), bone (20%) and liver (10%) (1).

The preoperative localization of the lesion is crucial to plan the correct treatment; the combination of two tests, Tc-99m-sestamibi parathyroid gland scintigraphy and ultrasound of the neck, offers the best results. If parathyroid carcinoma is suspected, CT scan and MRI should be performed to evaluate local extension for disease staging. Flourine 18 fluorocholine positron emission tomography helps to localize the tumor when the results of scintigraphy are inconclusive or negative, with sensitivity of 90% and positive predictive value of 100% (6).

The definitive diagnosis requires histopathological confirmation, which is complex as with other endocrine neoplasms, given the difficult differentiation from atypical adenoma. The development of tumor cells beyond the parathyroid capsule, and the presence of lymph nodes and vascular involvement confirm malignancy (3).

Surgery is the treatment of choice of parathyroid carcinoma. En bloc resection of the parathyroid gland with the ipsilateral thyroid lobe is recommended, as in this case. Parathyroid hormone is a very useful tumor marker to follow up these patients; persistent elevated values of PTH or calcium are associated with local recurrence, incomplete resection or distant disease.

Its prognosis is variable and is influenced by delayed diagnosis, local invasion or distant metastases at the time of the initial consultation (5). Morbidity and mortality in these patients are more related with the management of hypercalcemia and its renal and cardiovascular complications than with the local progression of the disease.

After 30 months of follow-up, our patient has no clinical or biochemical evidence of disease.

References:

1. Ramírez Tejeda S, Sosa Eroza E, Ferreira Hermosillo A. (2016). Crisis hipercalcémica por cáncer paratiroideo: reporte de un caso. Revista Mexicana de Endocrinología, Metabolismo y Nutrición. 3:182-188.
2. Wilttveen JE, van Thiel S, Romjin JA, Hamdy NA. (2013). Hungry bone syndrome: still a change in the post-operative management of primary hyperparathyroidism: a systematic review of the litera- ture. Eur J Endocrinol. 168(3):45-53.
3. Mohebati A, Shaha A, Shaha J. (2012). Parathyroid carcinoma: challenges in diagnosis and treatment. Hematol/Oncol Clin Norht Am. 26:1221-1238.
4. Wei CH, Harari A. (2012). Parathyroid carcinoma: update and guidelines for management. Curr Treat Options Oncol. 13:11-23.
5. Witteveen JE, Haak HR, Kievit J, et al. (2010). Challenges and Pitfalls in the Management of Parathyroid Carcinoma: 17-Year Follow-Up of a Case and Review of the Literature. Horm Canc. 1:205-214.
6. Wouter P. Kluijfhout, MSc Jesse D. Pasternak, MD Jessica E. Gosnell, MD Shen WT, Quan-Yang Duh, Vriens MR , et al. 18F Fluorocholine PET/MR Imaging in Patients with Primary Hyper- parathyroidism and Inconclusive Conventional Imaging: A Prospective Pilot Study.

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