A Rare Case of Clinically Silent Giant Pheochromocytoma

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Citation

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Abstract

We report the case of a 70-year-old male patient who was firstly admitted for a pathologic fracture in the left scapula in 1998, which was attributed to metastatic neuroendocrine carcinoma. The CT presented a soft tissue mass (9x7x9 cm) in the left adrenal characterized as adrenal tumor. MIBG scanning was positive for pheochromocytoma, despite the lack of any other clinical or laboratory finding. The excised tumor proved histologically to be malignant pheochromocytoma. A year later, a new CT showed relapse of the tumor. Again no clinical or laboratory findings towards pheochromocytoma were present. The tumor removed once again. The patient's follow-up has reached 5 years with no obvious metastases or local recurrence. Although malignant, large, non-secreting pheochromocytoma in patients older than 60 years is very uncommon, it should be considered in cases of metastatic neuroendocrine tumor of unknown primary lesion.

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We report the case of a 70-year-old male patient who was firstly admitted to “Agii Anargiri” hospital for a pathologic fracture in the left scapula in 1998. His father died at the age of 75 from prostate cancer. His own medical history included appendectomy (in 1967), nephrolithiasis and congenital abnormality of the left kidney, prostate hyperplasia (from 1995) and a pathologic fracture in the head of the right femoral bone (in 1996). The fracture was attributed to tumor metastasis. Resection of the tumor and bone reconstruction followed. The histological examination of the tumor showed extended metastases of a low-grade carcinoma that could not be identified. The cytological examination of the tumor indicated metastatic adenocarcinoma. As the primary tumor has not been identified by laboratory and imaging control, the patient underwent radiotherapy and entered a clinical follow-up.

Laboratory examinations on admission were normal except slightly elevated serum Ca^{++} (2.65 mmol/L) and a-FP (4.68 μg/L). CEA and Ca 19-9 were within normal limits. The chest x-ray, the ECG and the cardiological examination were normal. The tumor of the left scapula removed surgically. The histological examination of the tumor revealed metastatic neuroendocrine carcinoma most probably originating from lung. Chest CT scan did not reveal any lesion in lungs or enlarged lymph nodes in the mediastinum. Upper abdomen CT presented a soft tissue mass (9x7x9 cm) in the left adrenal characterized as adrenal tumor and a hyperplastic prostate gland. Ultrasonography of the left adrenal revealed tumor, probably incidentaloma. IV pyelography showed version of the left kidney and pachynsis of the urinary bladder wall and prostate hyperplasia.

Plasma, urine catecholamines (adrenaline, noradrenaline and VMA) and dopamine had normal values, and showed no evidence of pheochromocytoma. MIBG scanning was positive for pheochromocytoma. No clinical indications for MEN I or MEN II were present.

No preoperative therapy was necessary. The tumor removed through a flank incision with removal of the 11th rib. No intraoperative or postoperative complications were recorded. The histological examination of the tumor showed malignant pheochromocytoma of the adrenal (dimensions12x8x10cm) with central necrosis and smooth fibrous capsule envelope. Biopsies taken from the tissues surrounding the tumor were negative for neoplasia.

The patient did not receive chemotherapy, as follow-up was preferred. A year later, the patient had a new chest-upper and low abdomen CT scan that showed a new mass in the site of the operation (dimensions 3.5x2x3). Ultrasonography showed local recurrence of the tumor. Urine and blood catecholamines and their products were once again normal

and no clinical symptoms of pheochromocytoma were present. The tumor removed once again with flank incision and no postoperative complications observed.

The patient's follow-up has reached 5 years and no metastases or local recurrence observed. Hypertension unrelated to tumor was developed 1.5 year after the second operation.

Pheochromocytomas are usually benign unilateral tumors (only 5% are malignant) that follow familial distribution and are usually observed between the 20-50 years of age. In our case, the tumor was malignant, unilateral (right adrenal), there were no signs of familial distribution of the disease and our patient was older than 50 years. These epidemiological data did not primarily support the diagnosis of pheochromocytoma[1].

Pheochromocytomas produce a number of other biologically active peptides that may cause vascular and visceral disturbances. The most common manifestation of the disorder is sustained or paroxysmal hypertension. Headaches, palpations, and chest discomfort are also common[2,3,4,5]. Our case presented no evidence or reference of any of these symptoms; neither had increased arterial blood pressure at any time.

The diagnosis of pheochromocytomas is assisted by the elevated plasma values of adrenaline (>100ng/L) and noradrenaline (>400ng/L). Increased values of adrenaline and noradrenaline indicate malignant pheochromocytoma in adrenal or in Zuckerkandl's organ[2,3,4,5]. Notably, in our case, the disease was misdiagnosed two times, although metastases were present. The inability of imaging examinations to contribute at early stages of the disease, the paradoxical growth of the tumor and its non-secreting nature was the probable causes. Finally, the diagnosis was made from the histological examination of the last metastasis.

Pheochromocytoma that is malignant, large, non-secreting in patients older than 60 years is very uncommon[6], but should be considered especially in cases of metastatic neuroendocrine tumor with no profound primary lesion.

References

1. Lucon AM, Pereira MA, et al. Pheochromocytoma: study of 50 cases. J Urol Apr 1997;157(4):1208-12.
2. Bouloux P, Fakkeh M. Investigation of pheochromocytoma. Clin Endocrinol 1995;43:657-64.
3. Bravo EL. Evolving concepts in the pathology, diagnosis and treatment of pheochromocytoma. Endocrinol Rev 1994:15:356-68.
4. Sheeps SG, Jiang NS, Klee GG, et al. Recent developments in the diagnosis and treatment of pheochromocytoma. Mayo Clin Proc 1990:65:88-95.
5. Bravo E, Gifford RW. Pheochromocytoma: diagnosis, localization and management. N Engl J Med 1984;311:1298-303.
6. Basso L, Lepre L, et al. Giant Phaechromocytoma: Case Report. Ir J Med Sci Jan-Mar 1996;165(1):57-9.

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