Clinical Case Report

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Giant malignant pheochromocytoma in an elderly patient

A case report

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Abstract

Rationale: Malignant pheochromocytoma is a rare disease and surgical resection is the only curative treatment.

Patient concerns: An 81-year-old man of Chinese ethnicity was found to have a giant retroperitoneal tumor.

Diagnoses: B-scan ultrasonography and CT scan presented a mass above the left kidney, measuring 13.5 × 10 .6 × 9.8 cm. Subsequent analysis of 24-h urinary catecholamines and vanillylmandelic acid, as well as of blood catecholamines and blood cortisol, showed no elevated levels.

Interventions: The patient was treated with surgery.

Outcomes: The result from immunohistochemical staining confirmed the presence of malignant pheochromocytoma. After three months follow-up, the blood pressure and serum potassium were all within normal limits, no post-operative complications, no tumor recurrence and metastasis were found.

Lessons: This is the oldest patient known to have histologic documentation of this disease. Giant malignant pheochromocytomas are rare entities requiring clinical suspicion coupled with strategic diagnostic evaluation to confirm the diagnosis, personalized therapeutic treatment is required, particularly among elderly population.

Abbreviations: CT = computed tomography, PHEO = pheochromocytoma.

Keywords: elderly, giant, malignant pheochromocytoma

1. Introduction

Giant malignant pheochromocytomas (PHEOs) are rare, especially in elderly patients.[1] Differing from benign PHEOs, which produce catecholamines and present adrenergic syndrome without metastasis, malignant PHEOs may develop secondary malignant growths at a distance from the primary site. A preoperative diagnosis and therapeutic treatments may be difficult for malignant PHEO patients, particularly in the elderly, octogenarians, and older patients. In this report, we describe a case of a giant malignant PHEO, currently the largest known to be published was in an 81-year-old male patient, for which a retroperitoneal laparoscopic adrenalectomy was performed. The malignancy was confirmed by the result of postoperative pathology, biochemical, and radiological reports and examinations. Three months after curative resection, the patient has been in good health since with no recurrence or metastasis.

2. Case report

An 81-year-old man presented with a 29-year history of hypertension and hypokalemia. Blood pressure was maintained at 200/120 mm Hg and serum potassium was 3.0 to 3.2 mmol/L. Antihypertensive medication is ineffective for him who arrived in the emergency department. The physical examination showed a cystic mass in the lower left abdomen. B-scan ultrasonography and computed tomography (CT) scan presented a mass above the left kidney, measuring 13.5 × 10.6 × 9.8 cm (Figs. 1 and 2). Subsequent analysis of 24-hour urinary catecholamines and vanillylmandelic acid, as well as of blood catecholamines and blood cortisol, showed no elevated levels.

Our presumptive diagnosis based on imaging results was a PHEO. Then we conducted a retroperitoneal laparoscopic adrenalectomy and no lymphatic metastasis was noted. Before surgery, patient was initiated on a regimen of 10-mg dose phenoxybenzamine administered twice a day for preoperative 7 days, and the blood pressure was about 165/85 mm Hg in these days. During surgery, after the superior pole of the kidney was exposed, mass became more visible. Surrounding fat or adjacent tissue was used to manipulate the mass to minimize blood pressure fluctuations and bleeding. The operative duration was 84 minutes and the blood loss was <100 mL, and therefore no need of blood transfusion. Blood pressure fluctuated within the range of 60/40 mm Hg during surgery. The blood pressure

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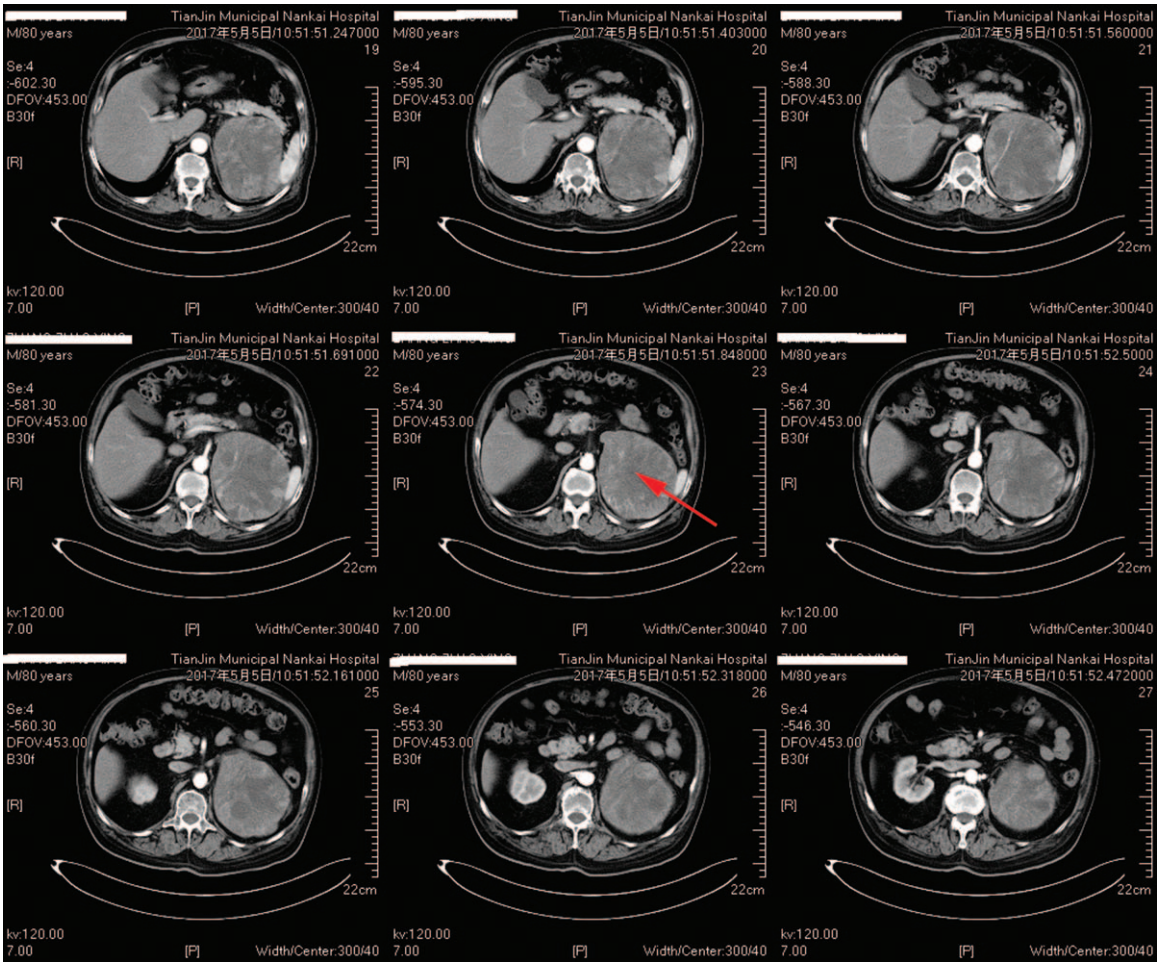

Figure 1. 13.5 × 10.6 × 9.8 cm mass (red arrow) in the left adrenal gland of a computed tomography (CT).

was 145/80 mm Hg and serum potassium 3.7 mmol/L on the first day after operation. The result from immunohistochemical staining confirmed the presence of malignant PHEO (Fig. 3). Immunohistochemical staining was positive for CD56 and 12% of Ki-67, it's negative for CgA, Syn, CK, S-100, GFAP, PAS, Des, CD99, and Vim. After 3 months follow-up, the blood pressure and serum potassium were all within normal limits, no postoperative complications, no tumor recurrence, and metastasis were found. This study was approved by the Ethics Committee of the Tianjin Nankai Hospital and the patient signed an informed consent form for publication of the case to be included in this study.

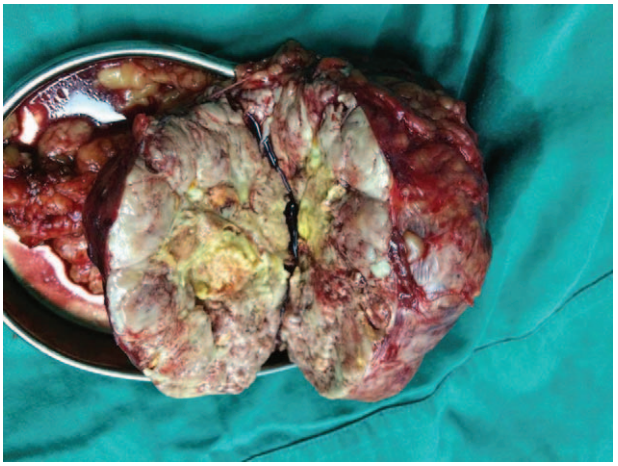

Figure 2. Macroscopic view of adrenal mass (13.5 × 10.6 × 9.8 cm).

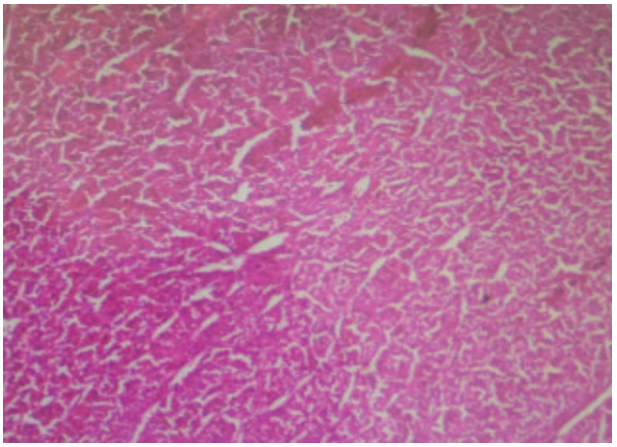

Figure 3. Pathological view confirmed the presence of malignant pheochromocytoma.

3. Discussion

The classic presentations of PHEO are included paroxysmal headaches, sweating, palpitations, and anxiety. Preoperative diagnosis is usually made by the presence of clinical signs and the determination of catecholamines and their metabolites in blood and urine. When within the range of 8% and 12.5%, PHEOs are classed as malignant. Previous study showed the incidence of metastatic PHEO was approximately 15% to 26%.[2] It is identified by the presence of local invasion or metastatic. The giant asymptomatic and nonsecreting PHEOs are rare. The larger the size of the tumor, the excessive excretion of dopamine, and the local extension are arguments in favor of the malignancy. According to Sturgeon et al,[3] the size >6 cm could be a predictor of malignancy. We would like to emphasize the importance of preoperative diagnosis that can establish the accurate diagnosis in elderly giant PHEO patient. According to above criteria, we can reduce the mortality rate and the complications related to the disease. The basic principles in the treatment of malignant PHEOs are to surgically resect the primary tumor and to prevent its recurrence or metastasis whenever possible, as well as to treat hypertensive symptoms by catecholamine blockade.

Our patient presents with an abdominal mass without any clinical signs except a 29-year history of hypertension and hypokalemia, which is similar to the classic triad presentation of aldosteronism. Our case indicated that the radiological findings of PHEO were useful for the diagnosis; an ultrasound could be employed as a primary investigation; abdominal CT has high sensitivity to suggest malignancy and can reliably localize most PHEOs, with accuracy approaching 100%.[4] Tumors enhanced markedly and internal density is not uniformed on CT scanning in our patient. With regard to the current treatment of patients with malignant PHEOs, treatments were usually not effective and most medical cure are palliative; tumor and metastasis resections are conducted for some resectable cases, it could prolong the lifespan and reduce risk of the damage to the cardiovascular system.

4. Conclusion

In conclusion, malignant PHEO and nonsecreting PHEO are very rare and aggressive neoplasms in the elderly, and there are often many other comorbidities worsening the survival.[5] A single case is not a guideline in the management of elderly patients with this rare disease. The therapy treatment plans for malignant PHEO are not standardized and must be personalized, particularly among elderly patients.

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Investigation: Chengquan Ma.
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