

Hormonally Inactive Malignant Pheochromocytoma: Role of Multislice CT Reconstructions in the Preoperative Differential Diagnosis and Therapy Planning

T Franiel, C Enzweiler, P Rogalla

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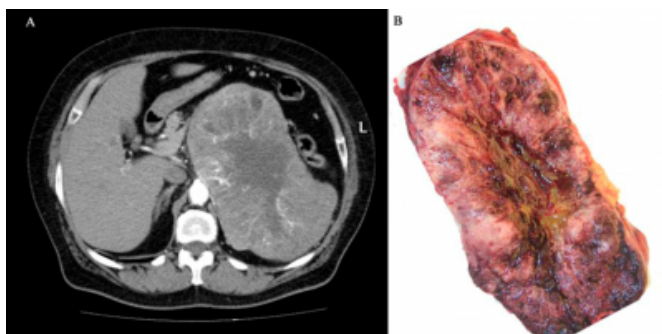
Abstract

An ultrasound examination performed by her general practitioner demonstrated a retroperitoneal tumor in a 64-year-old woman with arterial hypertension. The tumor appeared to arise from the left kidney. The patient was in good general condition and her laboratory values were normal except for a slight increase in serum creatinine (130.0 mol/l) and an elevated serum urea level (11.5 mmol/l).

Renal cell carcinoma was suspected and the patient was referred for computed tomography (CT) to evaluate local tumor extent. The axial scans suggested a renal cell carcinoma with a maximum axial diameter of 170 mm (Fig. 1 A).

Figure 1

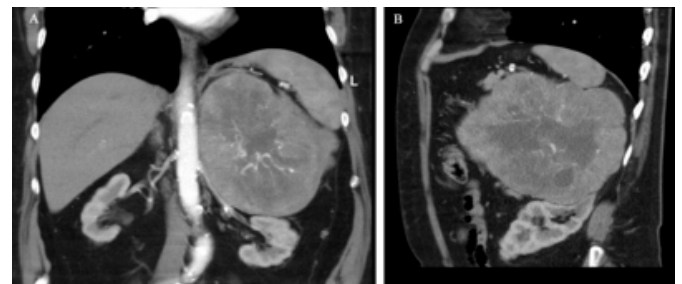
Figure 1: Contrast enhanced axial CT scan (A) and the corresponding cut section of gross specimen (B) of hormonally inactive malignant pheochromocytoma.



Moreover, the basal lung segments included in the scan volume contained multiple round lesions. Only the paracoronal and parasagittal views reconstructed from a multislice spiral CT dataset excluded a tumor origin in the kidney (Fig. 2 A&B).

Figure 2

Figure 2: Paracoronal (A) and parasagittal (B) views reconstructed from a contrast enhanced multislice CT.



There was no tumor extension into the inferior vena cava or renal vein. The fact that the left adrenal gland was not demarcated suggested an adrenal tumor and the suspicious pulmonary lesions a malignancy. Endocrinologic laboratory tests did not identify any abnormal hormonal activity. A metastasis from an unknown primary was considered to be highly unlikely on the basis of the size of the adrenal tumor since adrenal metastases have an average size of 2 cm.¹ Thus, the entities to be considered in the differential diagnosis were an occult malignant pheochromocytoma and cancer of the adrenal cortex. These two tumors cannot be differentiated on the basis of their morphologic appearance and enhancement pattern on CT scanning.^{2, 3} Moreover, a pheochromocytoma may mimic many other adrenal tumors.² Additional entities to be considered therefore also included rare adrenal neoplasms such as angiosarcoma and primary malignant melanoma.⁴ The pulmonary lesions made benign tumors such as large degenerative adenoma of the adrenal cortex or ganglioneuroma highly unlikely.^{4, 5} The kidney-sparing operative approach was planned and performed on the basis of the reconstructed MSCT data. A lobulated and

encapsulated tumor weighing 1238g and measuring 170 mm x 140 mm x 110 mm was removed (Fig. 1 B). Histology and immunohistochemistry of the surgical specimen yielded the diagnosis of a hormonally inactive malignant pheochromocytoma with a proliferation rate of about 2% (MIB-1) based on the demonstration of vascular invasion with strong expression of Chromogranin A, Synaptophysin and S-100.

CORRESPONDENCE TO

Franiel, Tobias, M.D. Department of Radiology Charité
Campus Mitte Schumannstraße 20/21 D- 10098 Berlin
Germany e-mail: tobias.franiel@charite.de

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Author Information

T. Franiel, M.D.

Department of Radiology, Charité

C. Enzweiler, M.D.

Department of Radiology, Charité

P. Rogalla, M.D.

Department of Radiology, Charité