Somatostatin-Receptor Scintigraphy Identifies a Cardiac Pheochromocytoma

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A 17-year-old woman had paroxysmal hypertension. Very high levels of urinary catecholamines suggested a diagnosis of pheochromocytoma (urinary norepinephrine 27 600 nmol/24 hours, normal <485 nmol/24 hours; urinary normetanephrine 37 607 nmol/24 hours, normal <2000 nmol/24 hours), but no adrenal tumor was found on the abdominal CT scan. Metaiodobenzylguanidine (MIBG) whole-body scintigraphy was performed, and no abnormal uptake was observed. To locate the tumor, 111In-labeled pentetreotide somatostatin-receptor scintigraphy was performed. An intense focal uptake was seen in the thoracic area (Figure 1). An intravenous injection of 99mTc microspheres was used to label the lungs. The subsequent double-isotope tomoscintigraphy located the tumor beside the right lung in the inferior mediastinum (Figure 1).

ECG-gated MRI and transesophageal echocardiography gave further information about the anatomic extent of the tumor. The pheochromocytoma was located adjacent to the right atrium (Figure 2).

The patient underwent surgical removal of the tumor and had an uneventful recovery, except for an atrial fibrillation that was resolved after a few weeks. This case emphasizes the value of pentetreotide scintigraphy to localize pheochromocytomas when MIBG scan and abdominal CT scan are negative.

Figure 1. Whole-body scintigraphy 48 hours after the administration of 185 MBq of 111In-pentetreotide. Anterior and posterior views reveal a focal uptake above the liver. A transaxial slice acquired by double-isotope single photon emission CT shows that the tumor labeled with 111In pentetreotide (red) is in the mediastinum beside the right lung, which is labeled with 99mTc microspheres (green).
Figure 2. A $T_1$-weighted black-blood spin-echo axial MRI slice (left) shows the pheochromocytoma as a bright tumor adjacent to the right atrial free wall and to the right lung. A breathhold bright-blood cine-MRI oblique slice (right) shows the tumor extending behind the superior vena cava.
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