We report a case of a single intrahepatic pheochromocytoma in the absence of an adrenal lesion and no evidence of metastatic disease. The patient had strong clinical and biochemical evidence of a pheochromocytoma. A CT scan was abnormal but nondiagnostic for pheochromocytoma. An $^{123}$I-metaiodobenzyl guanidine (MIBG) scan was falsely negative, but an MRI scan showed a definitive hepatic abnormality. After confirmation of endocrine activity by venous sampling, the tumor was surgically removed. The patient's symptoms have resolved and her plasma catecholamine levels as well as her 24-h urine catecholamine excretion have normalized. The case shows an unusual location of an isolated pheochromocytoma and provides an example of a false negative $^{123}$I MIBG scan. Am J Hypertens 1996;9:1040–1043

KEY WORDS: Pheochromocytoma, hepatic tumor, magnetic resonance imaging, radioiodinated metaiodobenzyl guanidine scan.

We have encountered a case of an intrahepatic pheochromocytoma without evidence of any other tumor, indicating that the lesion was likely to be a primary tumor. All previously reported cases of intrahepatic pheochromocytoma have been metastatic. Our patient underwent surgical removal of the tumor with complete resolution of clinical and biochemical abnormalities.

CASE REPORT

A 42-year-old woman presented to her internist with a history of episodes of palpitations, sweating and occasional headaches. She first noted such episodes at age 30 when she was also found to be hypertensive. The episodes were initially tolerable, but increased in frequency over the ensuing years. During the three months prior to her visit, her blood pressure had become difficult to control and reached levels of 250/130 mm Hg. She lost seven pounds. Two 24-h urine collections for catecholamines and metabolites showed markedly increased amounts of norepinephrine, metanephrines, and vanillylmandelic acid, with normal epinephrine excretion. There was no family history of pheochromocytoma or related endocrinopathies. An abdominal computed tomography (CT) scan was done and read as negative, and the patient was referred to the Hypertension Section at the University of Cincinnati Medical Center for further workup (Figure 1).

Our initial evaluation revealed a blood pressure of 190/94 mm Hg supine and 168/76 mm Hg upright, with pulses of 72 and 92 beats/min, respectively. The patient appeared to be in good general condition and the remainder of the physical examination was unremarkable. Her eye grounds showed no signs of hypertensive vascular changes. Laboratory evaluation showed normal blood count, renal and hepatic function. A clonidine suppression test was performed and showed lack of suppression of plasma catecholamines. The results were, in pg/mL (baseline/3 h post clonidine 0.3 mg): norepinephrine 5504/6281, epinephrine 46/56, dopamine <10/<10. A $^{123}$I metaiodobenzyl guanidine (MIBG) scan done the following week was negative. The CT scan done at another hospital was reviewed and a
subtle $4.5 \times 3$ cm lesion in the liver was detected. A magnetic resonance imaging (MRI) scan confirmed the presence of a mass in the medial segment of the left liver lobe on the T-2 weighted image (Figure 2). To confirm that this liver lesion was indeed catecholamine producing, selective vena cava sampling for catecholamines was done.

In preparation for surgery, a CT portogram and hepatic angiography were performed with pharmacological blocking. This revealed a highly vascular tumor in the medial segment of the left lobe with no other lesions in the remainder of the liver (Figure 3). The tumor extended to the right hepatic vein. The patient was started on 20 mg phenoxybenzamine twice daily, 50 mg metoprolol four times daily, and nifedipine pm. Six weeks later she underwent left hepatic lobectomy and cholecystectomy. Intraoperatively, the liver tumor was found to be completely encapsulated. There was a small area near the right hepatic vein where the capsule was entered during surgery to free the tumor from the right hepatic vein without damaging the vessel, and a small amount of tumor tissue had to be left in place.

The cut surface of the left lobe showed a $4.5 \times 5.0 \times 3.8$ cm, dark red, soft to firm, well circumscribed encapsulated mass (Figure 4). Light microscopic review of multiple hematoxylin and eosin stain sections showed the classic histologic features of a paraganglioma (extraadrenal pheochromocytoma). Stains were positive for serotonin but negative for somatostatin and neurofilament.

The patient tolerated the procedure well. All antihypertensive medications were discontinued. Supine plasma catecholamine levels were obtained two weeks post surgery with the following results (in pg/mL): norepinephrine 417, epinephrine 43, dopamine < 10. On her last visit with her internist 14 months following surgery, she was on no medication and feeling well. Her blood pressure was 120/72 mm Hg sitting. A 24-h urine obtained at that time contained normal amounts of catecholamines, metanephrines and vanillylmandelic acid.

**COMMENT**

This patient presented with a typical clinical picture of pheochromocytoma and the diagnosis based on urinary catecholamine excretion was straightforward. The CT scan was initially misread as negative, but the clonidine suppression test confirmed the presence of a pheochromocytoma. The negative $^{123}$I-MIBG scan was surprising, given the high specificity of this test (approximately 95% true negative results). $^{123}$I is superior to $^{131}$I in identifying pheochromocytomas, but in this
case the high background activity of the liver tissue obscured the tumor. The information provided by the MRI scan was accurate and confirmed by subsequent surgery. Pheochromocytomas are hyperintense on T-2 weighting, allowing one to distinguish such lesions from paragangliomas or other nonfunctioning adenomas.

The mass was in the medial segment of the left lobe, and we elected to confirm the endocrine activity of the tumor by venous sampling. The preoperative hepatic angiogram demonstrated a highly vascular tumor. The patient tolerated the surgery well and had an excellent postoperative result. We have not found another case of an isolated intrahepatic pheochromocytoma in the peer-reviewed literature. Reference to a personal communication is made in a report of the Armed Forces Institute of Pathology. All well-described cases of intrahepatic pheochromocytoma described so far have been metastatic.

In summary, our case shows that: 1) pheochromocytoma in the liver can occur in the absence of a detectable adrenal lesion or other metastases; 2) $^{123}$I-MIBG scanning may give false negative results in this situation due to high background activity; and 3) surgery can produce a good immediate result. Only long-
term follow-up will show if a permanent cure has been achieved in our patient.

REFERENCES

