

Case Reports

Coincidence of Angiomyolipoma and Pheochromocytoma

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Introduction

Angiomyolipoma is a benign clonal neoplasm with a prevalence of 0.13% when screened by ultrasonography.⁽¹⁾ Pheochromocytoma is the causative factor of hypertension in less than 1% of the hypertensive population.⁽²⁾ Each of these neoplasms may be accompanied by other tumors; however, to best of our knowledge, concomitant angiomyolipoma and pheochromocytoma has not been reported yet and our report is the first in this matter.

Case Report

A 40-year-old woman presented with hypertension, resistant to treatment for several years. The results of complete blood cell count, serum electrolytes laboratory tests, renal and liver function tests, and urinalysis were normal. On ultrasonography, a mass in the left adrenal gland and a hyperechoic mass, approximately 4.5×3.5 cm, in the right kidney were reported (Figure 1). The 24-h urinary excretion of vanillylmandelic acid and homovanillic acid were in the reference range. A CT scan with oral and intravenous contrast media showed a left adrenal mass and a nonenhanced 5×5 -cm mass with a small amount of fat in the lateral surface of middle lobe of right kidney (Figure 2). Magnetic resonance imaging revealed a low-signal-intensity mass in the left adrenal gland (Figure 3) and a low-signal-intensity mass in the lateral surface of the middle lobe of the right kidney.

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FIG. 1. Ultrasonography: a hyperechoic mass in the right kidney

The patient underwent left adrenalectomy through a left 12th costal incision. The patient's position was then changed to the right flank and partial nephrectomy was also performed.

Histopathologic examination showed pheochromocytoma of the left adrenal gland and angiomyolipoma of the right kidney.

Discussion

Angiomyolipoma is a benign clonal neoplasm. This tumor is found in 0.3% of all autopsies and in 0.13% of the population when screened by ultrasonography.⁽¹⁾ Approximately 20% of angiomyolipomas are found in patients with tuberous sclerosis syndrome (an autosomal-dominant disorder characterized by mental retardation, epilepsy, and adenoma sebaceum).^(1,3)

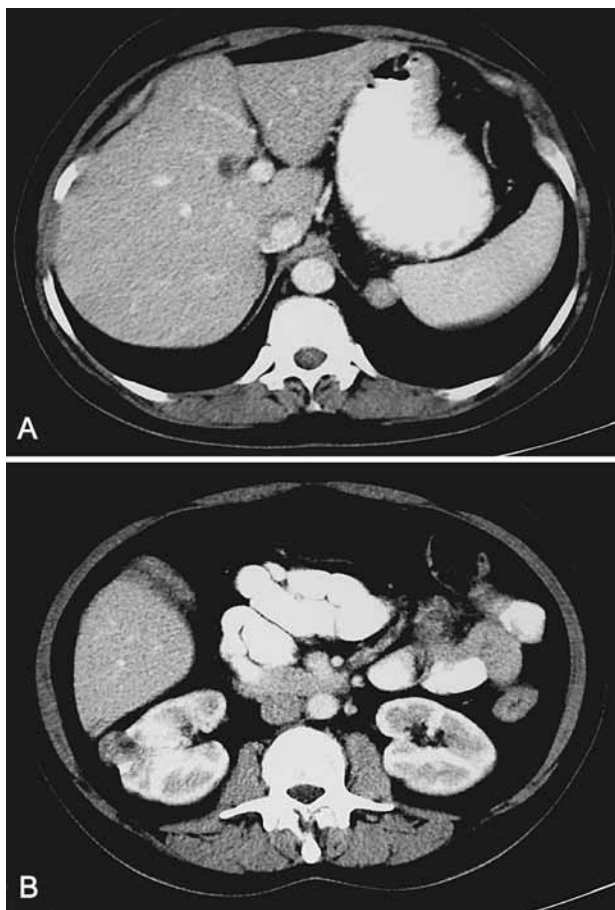


FIG. 2. CT scan with contrast-medium. **A.** a 2-cm tumor in the left adrenal gland, **B.** a mass on the external surface of the middle lobe of the right kidney

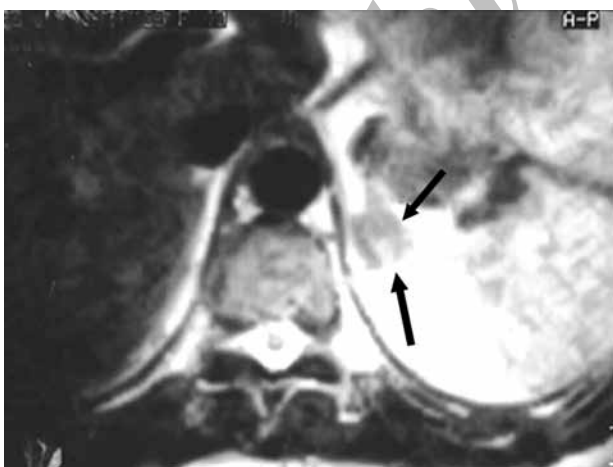


FIG. 3. Magnetic resonance imaging revealed a hypoechoic mass in the left adrenal gland (arrows).

Angiomyolipoma's common signs and symptoms include flank pain, hematuria, palpable mass, and hypovolemic shock. The presence of even a small amount of fat within a renal lesion on CT scan (confirmed by Hounsfield units ≤ 10) is characteristic for angiomyolipoma.⁽⁴⁾ The typical but not diagnostic finding on ultrasonography is

a well-circumscribed, highly echogenic lesion, often associated with shadowing.⁽⁵⁾

Pheochromocytoma is the causative factor of hypertension in less than 1% of the hypertensive population.⁽²⁾ Additional signs and symptoms are numerous but not specific. Among these are headaches, sweating, pallor or flushing, palpitations, tachycardia, abdominal or chest pain, and postural hypotension. Also common are weakness, nausea, emesis, and anorexia.⁽²⁾ About 10% of pheochromocytomas are found in normotensive patients.⁽⁶⁾ Diagnosis is confirmed by demonstrating elevated levels of catecholamines in the blood or urine, which occur in 95% to 99% of patients with pheochromocytoma.⁽⁷⁾ Because of the severe consequences of the undiagnosed pheochromocytoma, it is recommended that hypertensive patients be screened. Measurement of urinary catecholamines and metanephrines is adequate in most patients. Rarely, the plasma and urinary concentrations of catecholamines and their metabolites are not elevated, especially if the patient is normotensive at the time of study,⁽²⁾ as in our patient.

To our knowledge, the coexistence of angiomyolipoma and pheochromocytoma has not been reported previously and our report was the first in this matter. It seems that the patient has had one of the following conditions: first, a renal tumor in the right kidney has developed metastases to the left adrenal gland—adrenal metastasis can be found in 9% of autopsies.⁽⁸⁾ Specifically, the adrenal glands have been found to be a site of metastasis in 40% of patients with renal cell carcinoma.⁽⁸⁾ Hypertension may be one of the paraneoplastic syndromes of renal tumor.^(7,9) Second, a pheochromocytoma in the left adrenal gland has given metastasis to the right kidney; hypertension is due to catecholine release from pheochromocytoma. And, third, it has been the presence of 2 separate neoplasms in the right kidney and the left adrenal gland.

The preferred procedure might be laparoscopic resection of tumors. However, after excision of adrenal mass, the patient's hemodynamic condition might become unstable; thus, we decided to perform adrenalectomy through the left 12th costal incision, and then if the patient's condition was appropriate, change the position and perform partial nephrectomy through a contralateral incision.

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Editorial Comment

The authors describe a case of coexisting contralateral adrenal pheochromocytoma and renal angiomyolipoma. This has been reported in at least 2 other patients to date.^(1,2) Both pheochromocytoma and angiomyolipoma are uncommon but not rare entities in the general population, approximating 1 per 100 in incidence. Both are much more common in the setting of multiple endocrine neoplasm (MEN) type 1.⁽³⁾ Coexistence of these two entities is hence statistically expectable and has been pathologically documented by this and previous cases. One must also keep in mind that the conventional histologic appearance of angiomyolipoma, normal adrenal tissue, and adrenal pheochromocytoma plus normal cortical tissue, is mistakably similar. Therefore, such findings must be correlated with the exact site of origin and whenever possible, with results of liberal immunostaining.⁽⁴⁾ This propensity for multifocal extrarenal (including adrenal) involvement by angiomyolipoma must be borne in

mind in coexisting lipid laden renal and adrenal masses.⁽⁵⁾ Finally, limitations imposed by availability and practice setting in this case are understandable. However, when conditions allow, one should approach the asymptomatic renal angiomyolipoma and symptomatic lesions less than 4 cm in diameter by angioinfarction.⁽⁶⁾

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Reply by Author

Two reports are available on the coexistence of angiomyolipoma and pheochromocytoma,^(1,2) but some differences exist between them and our report: in both previous reports, patients had multiple endocrine neoplasm (MEN). In one of them (an unusual combination of primary multiple apudomas and malignant angiomyolipoma of the kidney),⁽¹⁾ the authors have suggested that pheochromocytoma represented metastases of a malignant carcinoid tumor. In the second one (pheochromocytoma-ganglioneuroma of the adrenal gland), the patient had a composite adrenal tumor composed of both pheochromocytoma and ganglioneuroma in association with elevated urinary and serum catecholamine levels. In this regard it seems that the presence of these tumors (pheochromocytoma

and angiomyolipoma) without other tumors and without elevated urinary and serum catecholamine levels (that suggest the presence of MEN or other syndromes) was seen in our case as first.

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