순환기 : 제16권 제3호

Pheochromocytoma and Renal Artery Stenosis

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갈색세포종과 신동맥협착

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□ 국 문 초 록 □

신동맥성 고혈압과 갈색세포종은 각각 고혈압의 드문 원인 중 하나이다. 그러나 신동맥협착이 갈색세포종에 의해 나타날 경우 갈색세포종의 진단이 어려워질 수 있다.

갈색세포종은 수술로 완치할 수 있는 질병인 반면, 진단이 내려지지 않았을 때는 치명적 합병증을 수반할 수 있으므로 더욱 주의를 요한다.

저자들은 신동맥협착의 소견을 보이는 갈색세포종을 2례 경험하였다. 한 예에서는 신동맥성 고혈압의 수술 도중 갈색세포종이 발견되었고, 또 한 예에서는 신동맥협착의 혈관성형술을 시행한 후 발견되었다. 2례 모두에서 갈색세포종의 수술적 제거만으로 혈압은 정상화 되었다.

이처럼 흔하지는 않지만, 갈색세포종과 신동맥협착간의 연계가 있음을 숙지하고, 신동맥협착을 보이는 환자에서 임상적으로 갈색세포종의 가능성을 항상 염두에 두는 것이 불필요한 신동맥 성형술이나 수술을 예방하는데 필수불가결하다.

ABSTRACT

In hypertensive patients it is very important to detect renal artery stenosis or pheochromocytoma, since both diseases are curable causes of hypertension.

However, renal artery stenosis can be induced by pheochromocytoma, when the diagnosis of the two simultaneous diseases is very difficult. We experienced two cases of pheo-

chromocytoma presented as renal artery stenosis. Pheochromocytoma was overlooked when renovascular hypertension was diagnosed. Pheochromocytoma was found during surgery in one patient and after angioplasty in the other. In both cases, BP returned to normal after surgical removal of pheochromocytoma without repair of the stenosis. Prevention of ineffective and unnecessary renal artery angioplasty or surgery requires knowledge of this unusual association between pheochromocytoma and renal artery stenosis and a high degree of clinical alertness for pheochromocytoma.

Introduction

Pheochromocytoma and renal artery stenosis are rare causes of hypertensive diseases. Moreover, simultaneous occurrence of these two entities in the same patient is distinctly unusual. The early recognition of possible association of pheochromocytoma is important, because pheochromocytoma is also a cause of "surgically curable" hypertension. It is known that unrecognized pheochromocytoma leads to potentially lethal condition since hypertensive crisis or shock or both leading to death have been precipitated by drugs, anesthetic agents, parturition, or surgery for an unrelated condition(1).

We report here two cases of pheochromocytoma presented as renal artery stenosis and renovascular hypertension.

Case I

A 35-year-old woman was admitted with headache and palpitation of 20 days duration. The blood pressure was 190/120. Grade Ⅱ-Ⅲ hypertensive retinopathy was discovered. Grade Ⅲ-Ⅳ/Ⅵ systolic bruit was audible at right upper quadrant of abdomen. The chest roentgenogram, complete blood count, urinalysis, blood urea nitrogen, creatinine, and glucose were normal.

Vanillylmandelic acid (VMA) was 12.5mg/24hr (normal : 0.7-6.9mg/dl/24hr). Basal renin level was 3.1ng/ml (normal : 1-2.5ng/ml).

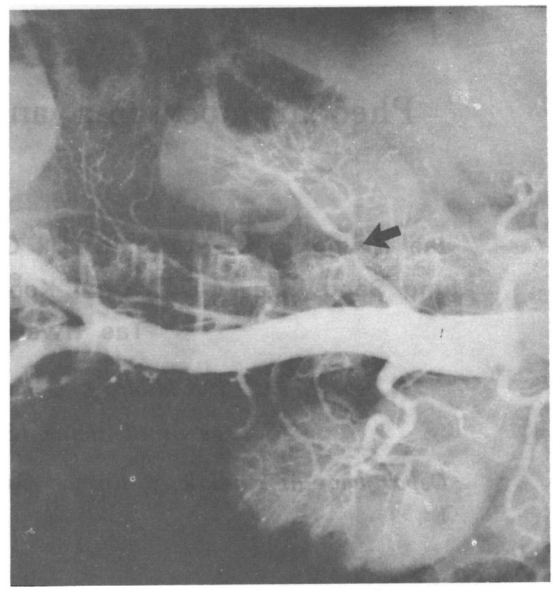

Fig. 1. Preoperative aortogram demonostrates stenosis (arrow) of right main renal artery and its dorsal segmental branch.

Abdominal aortogram demonstrated moderate stenosis of right main renal artery and severe stenosis of posterior branch (Fig. 1). At that time, selective renal vein renin sampling was done, and the ratio from the left side (3.6) to the right side (6.5) was one to 1.8, suggesting that the right renal artery stenosis was producing renovascular hypertension. It was thought that elevated VMA level was false positive finding.

Surgery was carried out for renal artery stenosis to perform autogenous saphenous patch angioplasty. At surgery, a well-encapsulated soft ovoid tumor, approximately 4×3×2cm sized, was found incidentally. It was located at medial side of upper pole of right kidney compressing the right main renal artery and its anterior branch. Manipulation of the tumor precipitated a sharp rise in systolic BP up to 240mmHg. Right adrenal gland was intact. After removal of this tumor, the renal artery looked to presume its normal contour.

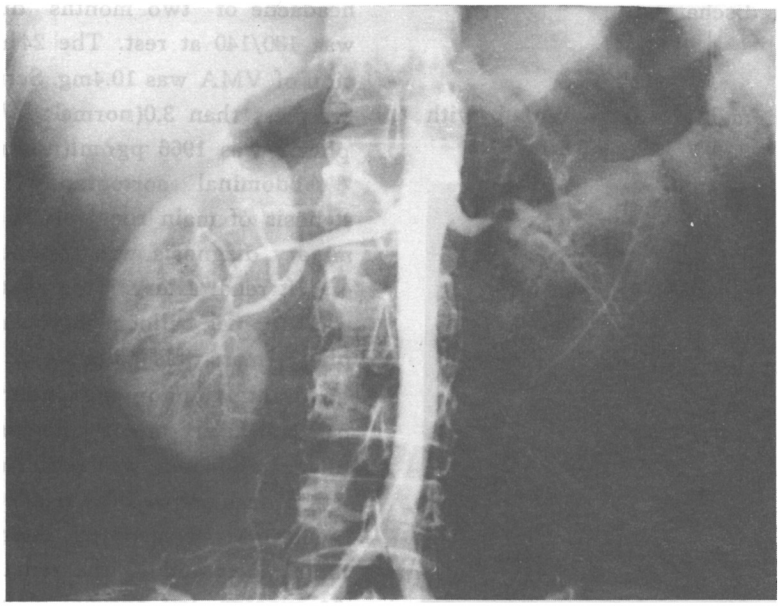

Fig. 2. Postoperative aortogram demonstrates normal right renal artery.

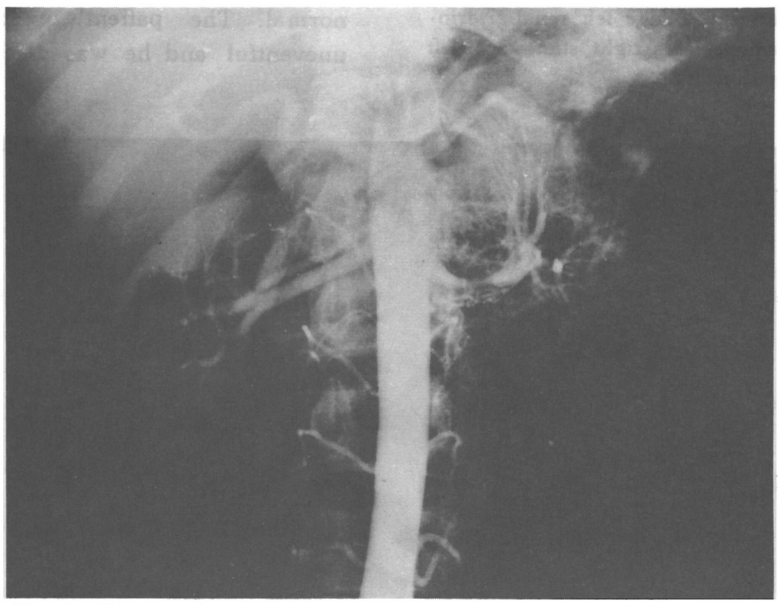

Fig. 3. Preoperative aortogram shows narrowing of left renal artery with fine vascularities around the lesion.

Microscopic examination of the tumor revealed extraadrenal pheochromocytoma. Postoperatively, patient's symptom was subsided and the BP stabilized at 120/80 mmHg. Urinary VMA was normal. Follow-up angiography 2wk later revealed normalized renal artery

(Fig. 2). He was discharged.

Case II

A 22-year old male was admitted with headache of two months duration. The BP was 180/140 at rest. The 24hr urinary excretion of VMA was 10.4mg. Serum epinephrine was less than 3.0(normal; 〈375)and norepinephrine was 1966 pg/ml(normal : 〈1050 pg/ml).

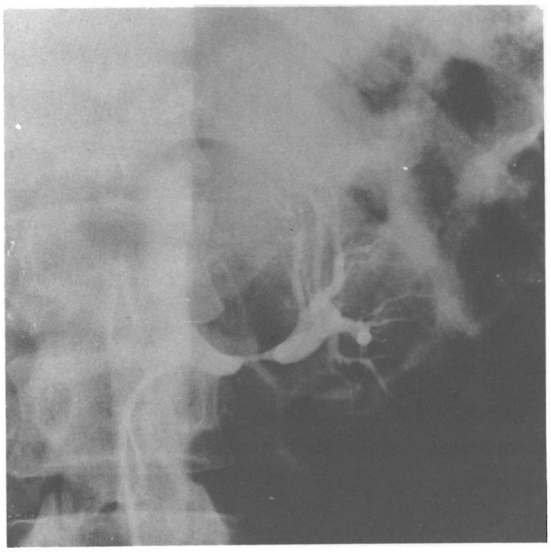

Fig. 4. Preoperative selective left renal arteriogram demonstrates tight stenosis of left main renal artery.

Abdominal aortography revealed severe stenosis of main renal artery (Fig. 3, 4). Preliminary diagnosis was fibromuscular dysplasia of left renal artery. This patient underwent transluminal balloon angioplasty. After angioplasty, mass was palpable at the left upper quadrant of abdomen. Under the impression of renal artery aneurysm, computed tomography of abdomen was performed.

CT scan showed the relatively well-defined ovoid mass located just lateral to the abdominal aorta and in the renal hilum (Fig. 5). At surgery, left adrenal gland was removed, and 5×5×4cm sized fixed mass was found.

Pathologic diagnosis was pheochromocytoma. During the postoperative period, the BP was normal. The patient's convalescence was uneventful and he was discharged.

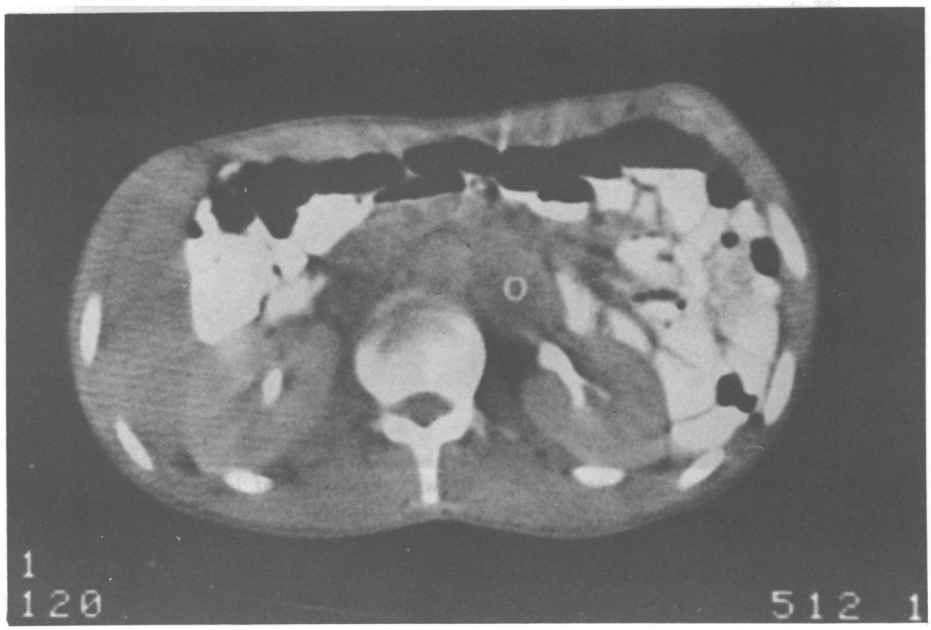

Fig. 5. Abdominal CT scan performed after angioplasty demostrates mass in the left renal hilum.

Discussion

The coexistence of renal artery stenosis and pheochromocytoma has been recognized since 1958 when Harrison first reported this unusual occurrence(2). Hill et al. in 1983 reviewed 36 patients with this association recognized since 1958(3). Extensive review of English literature revealed 5 further cases (4, 5, 6, 7) thereafter including the case reported by Hill himself. Recently there was also a case report, in which correct diagnosis of the coexistence of hypervascular pheochromocytoma and renal artery stenosis was made preoperatively in this country(8)

Previous literatures (3, 9, 10) suggested that pheochromocytoma may be associated with renal artery stenosis by several mechanisms; one is direct compression of the renal artery by a tumor in the renal hilum or angulation and constriction of the renal artery by fibrous band from the tumor. In this case, pheochromocytoma are usually of extraadrenal origin arising in the sympathetic ganglia adjacent to the renal hilum as in case I. Less often an adrenal pheochromocytoma may compress the renal artery by direct extension as in case II.

Another is renal artery spasm secndary to secreted catecholamine from the tumor. This is supported by the experimental work of Abrams et al(11), who found that the administration of epinephrine intravenously or intraarterially produces a local zone of narrowing in the main renal artery proximal to its bifurcation in dogs. The unilateral occurrence of renal artery stenosis in previously reported cases was immediately adjacent to the tumor, which suggested the "local leak" of catecholamines was also an important factor(4, 7).

Other minor causes are concurrent fibromuscular dysplasia, arteriosclerotic plaques of renal artery, or neurofibromatosis involving renal artery(9, 12).

For recently percutaneous transluminal angioplasty is widely accepted as a primary procedure for the treatment of renal artery stenosis it became more important to recognize the possible association of the two conditions early.

The suspicion of the possible association with review of laboratory findings, sonography or computed tomography might be helpful for the correct diagnosis of this unusual condition.

Otherwise, we may perform unnecessary angioplasty inadvertently or be faced with the risk of overlooking pheochromocytoma preoperatively.

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