Journal of Korean Medical Science
Vol. 3, No. 1, P 35-39, March 1988

Primary Intimal Fibroplasia of the Renal Artery

Yeon-Lim Suh, Je G. Chi and Joon Ryang Roh*

Departments of Pathology and Thoracic Surgery, College of Medicine, Seoul National University, Seoul, Korea*

Two cases of primary intimal fibroplasia of the renal artery with renovascular hypertension are described. Case 1 was 27 year old female who was incidentally found to have blood pressure of 210/130 mmHg on routine physical examination. Renal arteriogram revealed tubular narrowing of the mid protion of the left renal artery. Both patients showed lateralization in renin activity at the involved side renal vein. Case 2 was a 10 year old girl who was first noted to have a hypertension of 180/120mmHg after a sudden attack of seizure, vomiting and altered consciousness. Renal arteriogram showed concentric narrowing of the proximal half of the right renal artery.

Histopathologic examination of the affected arterial segments from both cases showed essentially same findings, i.e., diffuse fibrous thickening of the intima occluding the lumen, focal fragmentation, duplication and disappearance of the internal elastic membranes. There were no deposit of lipid and inflammatory cells. The media and adventitia remained intact. The blood pressure of both patients became normal, after the surgery and the patients are in good health up to this time

Key Words: *Fibromuscular dysplasia, intimal fibroplasia, renal artery stenosis, renovascular hypertension, childhood hypertension*

INTRODUCTION

Primary intimal fibroplasia is an infrequent type (1-2%) of the fibromuscular dysplasia of the renal artery that are characterized by fibrous or fibromuscular thickening of the layers of the artery involved (Harrison et al., 1971). The renal artery with primary intimal fibroplasia shows circumferential or eccentric accumulation of loose fibrous tissue without lipid or inflammatory cells. This type of the fibromuscular dysplasia is not common cause of renovascular hypertension in adults, but it is not uncommon type in pediatric age group (Novick et al., 1978).

This report concerns two cases of primary intimal fibroplasia of the renal artery in a 10 year and a 27 year old female patients with renovascular hypertension treated surgically.

Address for Correspondence: *Je G. Chi, M.D., Department of Pathology, College of Medicine, Seoul National University, #28 Yunkun-Dong, Chongno-ku, Seoul 110-744, Korea (Tel. 02) 7601-3540)*

CASE REPORT

Case 1. This patient was twenty years of age when hypertension of 210/130 mmHg was discovered on routine physical examination. She had been healthy except for intermittent headache that annoyed her for the last 6 months. There was no family history of hypertension. She had a mass in the right thyroid. Chest X-ray and EKG revealed no

abnormal findings. Under the impression of the renovascular hypertension diagnostic work-ups were done and included renogram, renal arteriogram and renal vein renin assay. Renogram demonstrated delayed function of the left kidney. Selective renal arteriogram revealed long segment of tubular constriction in the mid portion of the left renal artery together with abundant collateral vessels. Renal vein renin assay showed 8.0 ng/ml per hour on the left compared with 4.8 ng/ml per hour on the right, and 6.2 ng/ml/hour in the inferior vena cava. Plasma renin was 8.0 ng/ml per hour. The patient was treated with multiple antihypertensive drugs without satisfactory control of blood pressure.

She was readmitted to OB & GY department 7 years after the first hospitalization because of vaginal bleeding at 27 weeks of pregnancy. Her pregnancy was complicated by the placenta previa, and premature delivery of 520 gm fetus was resulted.

She was hospitalized again for the surgical management of known renovascular hypertension 6 months after the second admission. The physical findings were unchanged except for the hypertensive retinopathy, grade I. Renal arteriography revealed no progression of the stenotic lesion which was noted at the first admission. She underwent a nephrectomy after failure of an attempt of bypass graft for the stenosis. At operation it was found that there was discrepancy in size of both kidneys. The pathologic examination was not performed from the opposite kidney.

Removed kidney was decreased in size, measuring 8×4.5×2cm. It weighed 60gm. The renal artery measured 2cm in length and 0.2cm in diameter. The kidney capsule was stripped off easily. The cortex was smooth and markedly congested. Pelvocalyceal system remained intact. Histologic examination of the renal artery showed areas where intimal fibroblastic proliferation and collagen deposit was quite marked circumferentially (Fig. 2). The lumen was definitely narrowed by the circumferential intimal thickening, which was associated with fragmentation, duplication and disappearance of internal elastic membrane (Fig. 1). The media was focally deficient, but showed little alteration. There were marked tubular atrophy, patchy interstitial inflammatory cell infiltration and fibrosis. Global obsolescence of glomeruli were less than 10% of total glomeruli. The interlobar arteries revealed similar histologic alteration seen in the renal artery. Juxtaglomerular apparatus was generally prominent. One year later she became pregnant and delivered a normal infant. The blood pressue subsequently became normal and has remained so up to this time of report without medication.

Case 2. A 10 year old girl was transferred to Seoul Natioanl University Children's Hospital for the evaluation and management of known renovascular hypertension. She first came to medical attention after attacks of seizure, altered consciousness, severe headache and vomiting. These symptoms were preceded by the episodes of headache, abdominal pain and general weakness of 1 month duration. She was found to have hypertension of 180/120 mmHg, for which investigation was performed at other hospital. Physical examination revealed grade II systolic murmur heard on the apex. Left ventricular hypertrophy was traced on electrocardiogram. Plasma renin was 35.1 ng/ml/hour, aldosterone 116.2 ng/dl, and 17-hydroxycorticosterone 13.5 mg/24 hr. Spot urine vanil mandelic acid was negative. Right renal arteriogram showed segmental concentric narrowing of the proximal half of the right renal artery. To control the renovascular hypertension, local angioplasty was attempted without success.

On admission to SNUCH physical examination revealed an acutely ill-looking girl with marked emaciation. Blood pessure were 150/90 mmHg. Pulse rate was 100 per minute, respiration rate 24 per minute, and body temperature 36.5°C. Neither cardiac murmur nor rales was heard. On the abdomen aortic pulsation was found. Laboratory findings were as follows; Hemoglobin 12.8 mg/dl, Hematocrit 38.4%, WBC 11,000/mm^3 with differential counts of 89% seg, 8% lympho and 35% mono. Erythrocyte sedimentation rate was 45 mm/hr. Plasma protein, cholesterol,SGOT and SGPT were all remained within normal limits. Blood urea nitrogen and creatinine were 11 mg/dl and 1.3 mg/dl, respectively, Serum sodium, potassium and chloride were 163 mEq/L, 3.9 mEq/L and 108 mEq/L, respectively, Urinalysis showed 2 positive proteinuria. 24 hour urine contained total protein of 124 mg/dl, creatinine of 0.4 ng/dl and creatinine crearance of 50ml/min. Plasma renin and aldosterone were 20 ng/ml/hr and 80 ng/dl. Renal vein renin assay showed 20 ng/ml/hr on the right compared with 12.4 ng/ml/hr on the left. Inferior vena cava renin was 15.1 ng/ml/hr. Mantoux test revealed 2×2cm, erythematous lesion. 99mTc-DTPA kidney scan revealed nonfunctioning right kidney. Aortography showed nonvisualization of the right kidney and the renal artery. Clinically she was diagnosed to have Takayasu's arteritis, for which

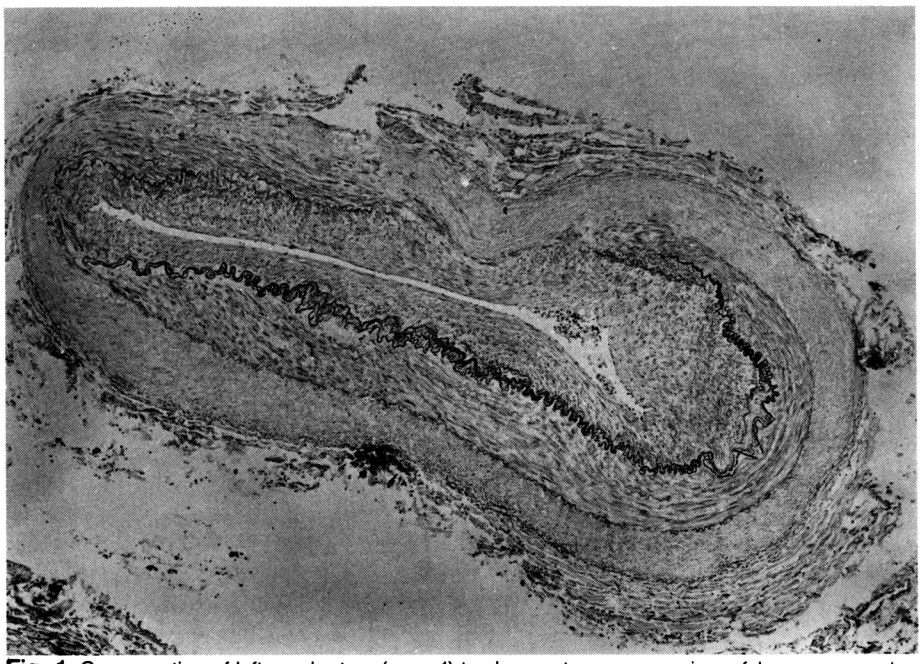

Fig. 1. Cross section of left renal artery (case 1) to show extreme narrowing of lumen caused by diffuse intimal thickening. Note focal duplication and loss of the internal elastic lamina (elastic, X10).

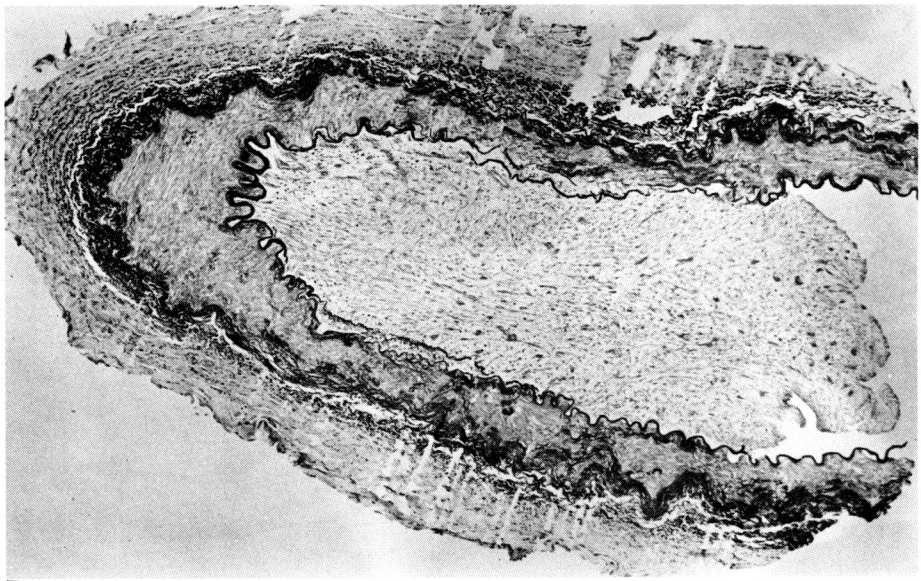

Fig. 2. The intimal thickening of the renal artery (case 1) consisting of collagen deposit and fibroblast proliferation (Masson-trichrome, X200).

revascularization using the great saphenous vein graft was done.

Histologic examination of the stenotic segment of the right renal artery showed intimal fibrous proliferation occluding the lumen. There were focal areas with fragmentation, duplication and loss of internal elastic lamina (Fig. 3). The media and adventitia showed little alteration. Follow-up DTPA renal scan done at the 7th postoperative day revealed that the perfusion of the right kidney improved. Plasma renin was 17.4 ng/ml/hr. Now 5 months after the operation the child is well, with blood pressure of 120/80 mmHg on no medication.

DISCUSSION

Fibromuscular hyperplasia may affect any of the muscular arteries and the branches of the aorta, but appears to have a particular affinity for renal arteries (Fry et al., 1973; Horwitz et al., 1985).

These lesions are considered to be congenital dysplasias with maldevelopment of the fibrous, muscular, and elastic tissue of the renal artery. The pathologic classification of fibrous and muscular renal artery diseases is based upon the layer of the arterial wall involved. Harrison and McCormack (1971) have classified the fibromuscular dysplasias into three groups; Intimal fibroplsia, medial fibromuscular dysplasia and adventitial fibroplasia. The first group, medial fibromuscular dysplasia is the most common and again consists of four types; (1) medial fibroplasia; (2) perimedial fibroplasia; (3) medial hyperplasia; (4) medial dissection. The medial fibroplasia with mural aneurysm is the type generally referred to as fibromuscular hyperplasia, and is the most frequently encountered type of fibromuscular disease (60-70%) in adults. It characteristically affects the distal two-thirds of the main renal artery, producing "string of beads" sign in arteriogram. Perimedial fibroplasia is the second most frequent variety of the medial dysplasias (15 to 20%) and is the most frequent lesion seen in pediatric age group.

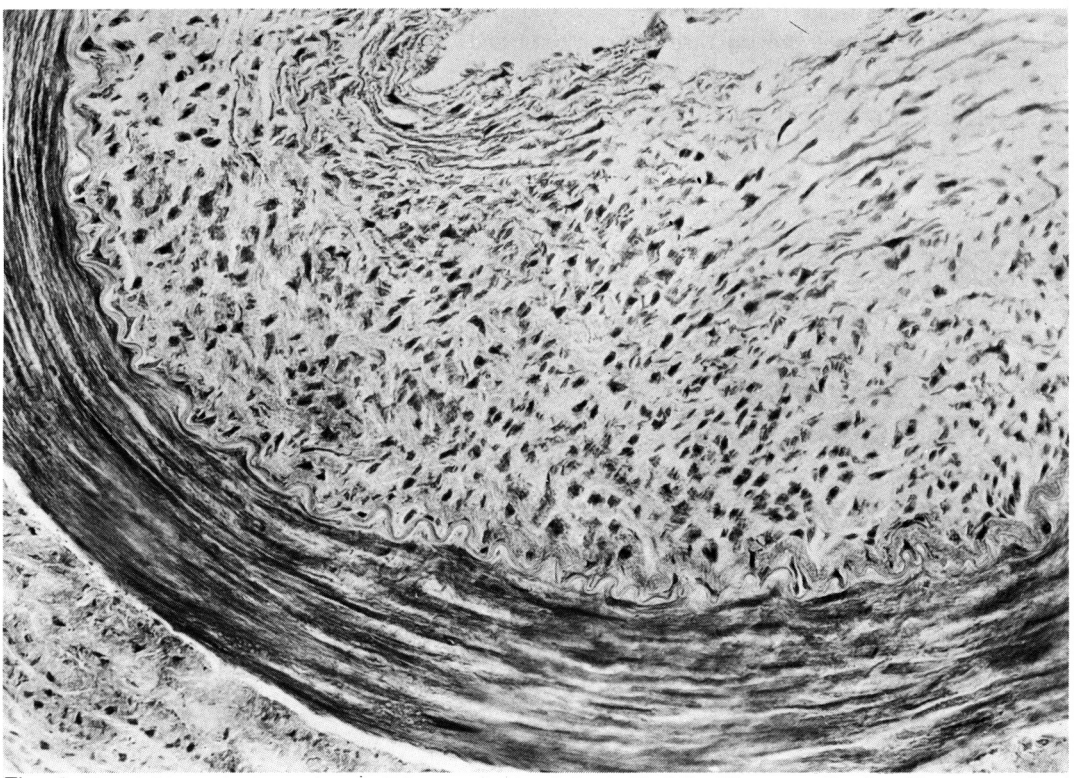

Fig. 3. The right renal artery of case 2 is completely obstructed by intimal fibrous proliferation. Also seen are focal duplication and loss of internal elastic lamina (elastic, X10).

It is characterized by fibromuscular thickening of the outer half of the media, replacing external elastic lamina. The third group, intimal fibroplasia, as seen in the present cases is characterized by a circumferential or eccentric accumulation of loose, moderately cellular, fibrous tissue involving the intima without lipid deposit or inflammatory exudate. Histologically, primary intimal fibroplasia must be differentiated from the secondary intimal fibroplasia that were found in a variety of renal parenchymal diseases such as cured malignant hypertension, chronic pyelonephritis and end-stage renal disease. The distinguishing point is that the primary intimal fibroplasia affects branches of renal artery proximal to the interlobar arterial level as opposed to others that affect smaller vascular structures. Of course other findings in the renal parenchyma should also be distinguishing ones.

Since the advent of arteriography, this disease is being found more frequently than before and is a relatively common cause of hypertension in the young (Kaufman et al., 1972; Hendren et al., 1982). There is a strong predominance in the female (about 75%), and it often is discovered in the second and third decades of life (Maxwell et al., 1972; Dean O'Neill, 1983; Braunwald et al., 1987). Bilateral involvement is in 40 to 50% of the patients, particularly if they are monitored periodically (Alterbarmakian et al., 1980).

There is no report on the ethnic difference in the incidence of certain type of fibromuscular dysplasia. However, it certainly seems to be extremely rare in Korea. We could not come up case reports on this disease in the Korean literature. The literature available to us contains only one case of medial fibromuscular dysplasia of the renal artery occurring in a 6 year old boy (Lee et al., 1971). At Seoul National University Hospital we have experienced only two cases of this entity. This paucity of record may not represent the actual status in this country. Some of the cases might not have been throughly examined in terms of renal artery, and some might have been classified into Takayasu's disease based on radiographic features.

REFERENCES

Altebarmakian VK, Rabinowitz R, Linke CA, Caldamone AA, Cockett ATK: *Surgical treatment of renovascular hypertension in children. J Urol. 124:877-881, 1980.*

Braunwald E, Isselbacher KJ, Petersdorf RG, Wilson JD, Martin JB, Fauci AS: *Harrison's principles of internal medicine. Eleventh edition, McGraw-Hill book company, pp 1201-1202, 1987.*

Dean RH, O'Neill JA: *Vascular disorders of childhoold. Lea & Febiger, Philadelphia, pp 77-95, 1983.*

Fry WJ, Ernst CB, Stanley JG, Brink B, Arbor A: *Renovascular hypertension in the pediatric patient. Arch Surg 107:692-698, 1973.*

Harrison EG, Mccormark LJ: *Pathologic classification of renal arterial disease in renovascular hypertension. Mayo Clin Proc 46:161-167, 1971.*

Hendren WH, Kim SH, Herrin JT, Crawford JD: *Surgical correctable hypertension of renal origin in childhood. Amer J Surg 143:432-442, 1982.*

Horwitz O, McCombs PR, Roberts B: *Disease of blood vessels. Les & Febiger, Philadelphia, pp 177-195, 1985.*

Kaufman JJ, Goodwin WE, Waisman J, Gyepes MT: *Renovascular hypertension in children. Amer J Surg 124:149-157, 1972.*

Lee CK, Oh JS, Lee HS, Kang JM, Lee SW: *A case of hypertension caused by fibromuscular hyperplasia of renal artery. J Kor Pediatr Assoc 14:393-397, 1971.*

Maxwell MH, Bleifer KH, Franklin SS, Varady PD: *Demographic analysis of the study. J Amer Med Asssc 220: 1105-1204, 1972.*

Novick AC, Benjamin S, Straffon RA: *Stenosing renal artery disease in children. Clinicopathologic correlation and results of surgical treatment. Nephron 22;182-195, 1978*