

## FIBROMUSCULAR DYSPLASIA OF THE RENAL ARTERIES TREATED WITH PERCUTANEOUS TRANSLUMINAL RENAL ANGIOPLASTY : A CASE REPORT\*

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**Summary:** Yalçinkaya F, Tümer N, Ekim M, Şanlıdilek U. (Pediatric Nephrology Unit, Department of Pediatrics, Ankara University Faculty of Medicine, Ankara, Turkey). Fibromuscular dysplasia of the renal arteries treated with percutaneous transluminal renal angioplasty: a case report. Turk J Pediatr 32: 265-271, 1990.

A nine-year-old girl with renovascular hypertension is presented. The diagnosis of fibromuscular dysplasia was established by selective renal angiography, and percutaneous transluminal renal angioplasty was performed. After successful dilation, the blood pressure returned to normal. The patient has been followed for a year and there is no evidence of hypertension. *Key words: fibromuscular dysplasia, percutaneous transluminal renal angioplasty.*

The incidence of secondary hypertension in the pediatric population has been reported to be high<sup>1</sup>. Renal and renovascular diseases account for nearly 80 percent of secondary hypertension in children, and approximately 70 percent of children with renovascular hypertension have fibromuscular dysplasia (FMD)<sup>2</sup>. In this report, we describe a patient with hyperreninemic hypertension and angiographically diagnosed FMD. Since the patient's hypertension could not be controlled with medication, a percutaneous transluminal renal angioplasty (PTRA) was performed.

### Case Report

A nine-year-old girl was admitted to the hospital with epistaxis. Physical examination revealed a blood pressure of 160/130 mmHg. She was a well developed child and had normal physical findings, including funduscopy examination.

Laboratory findings revealed a hemoglobin of 14 g/dl, hematocrit 46%, white blood cell count 12,000/m<sup>3</sup> with 60% neutrophils, 38% lymphocytes, 2%

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monocytes, and a normal platelet count. Urinalysis, BUN, serum creatinine, electrolytes, total protein, albumin and creatinine clearance were within normal limits. Urine cultures were negative. The chest radiograph, electrocardiogram, intravenous pyelography and abdominal ultrasound examinations were also normal. Urinary excretion of catecholamines and vanilmandelic acid (VMA) were found to be within the normal range. The peripheral plasma renin activity and aldosterone concentrations were high. In a supine position, the plasma renin activity was 4.5 ng/ml/hr (normal: 0.2-2.7 ng/ml/hr) and in an upright position it was 5.5 ng/ml/hr (normal: 0.4-3.2 ng/ml/hr). Plasma aldosterone concentration was 700 pg/ml (normal: 20-200 pg/ml).

Medical therapy was initiated which consisted of methyldopa (10 to 20 mg/kg/day), propranolol (2 to 5 mg/kg/day), captopril (1 to 1.5 mg/kg/day), prazosine (1 mg/kg/day) and hydrochlorothiazide (2 to 4 mg/kg/day) in increasing doses with various combinations for a year. But her blood pressure could not be controlled.

Intravenous digital subtraction angiography (DSA) was performed on the patient, and evidence of a possible stenosis was detected in the dorsal branch of the right main renal artery. To evaluate this stenosis further, selective renal angiography was performed which revealed a focal stenosis of the dorsal branch of the right main renal artery with its poststenotic dilatation (Fig. 1). The luminal diameter of this branch measured 2.4 mm, and the stenotic segment 0.4 mm, indicating high grade stenosis of the renal artery (Fig. 2). These angiographic findings were found to be characteristic of the focal type of FMD.

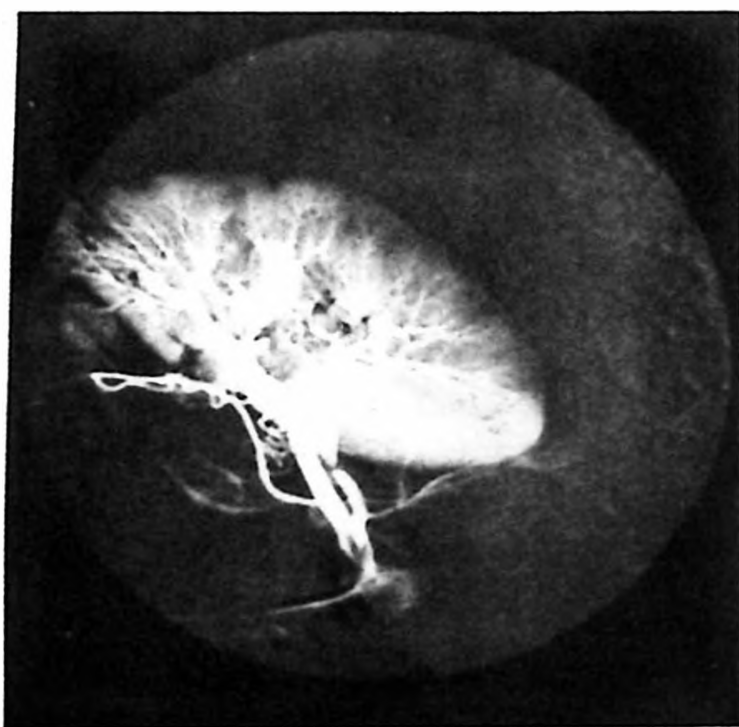


Fig. 1: Stenosis and post-stenotic dilatation of the right renal artery.



Fig. 2: High-grade stenosis of right renal artery.

Her right stenotic side ( $R_1$ ), left nonstenotic side ( $R_2$ ), inferior vena cava (IVC), and plasma renin activities ( $R_3$ ) were 3.9 ng/ml/hr, 1.3 ng/ml/hr, and 2.3 ng/ml/hr, respectively. The renal vein renin data were analyzed by using the plasma renin activity ratio (PRA) and suppression index. The PRA ratio is the ratio of PRA of the stenotic side to the nonstenotic side, and values equal to or greater than 1.5 are considered significant. The suppression index is the ratio of PRA of the nonstenotic side to PRA of IVC. If the values are equal to or less than one, it can be assumed that contralateral renin secretion is suppressible<sup>3,4</sup>. Thus, our patient's PRA ratio values which were ( $R_1/R_2 = 3.9/1.3$ ) greater than 1.5 and the suppression index which was ( $R_2/R_3 = 1.3/2.3$ ) less than one were considered significant (Table I).

Since there are various technical difficulties and risks involved in surgical intervention, PTRAs were performed on the patient. A successful dilation, using local anesthesia was carried out by introducing an inflatable balloon-tipped catheter over the guide wire percutaneously (Figs. 3, 4). After PTRAs, the blood pressure returned to normal, and anti-hypertensive therapy was discontinued. The patient has had normal blood pressure for a year without the use of any medication.

TABLE I: Plasma Renin Activity (PRA) and Suppression Index in Our Patient

| PRA Stenotic / PRA non-stenotic<br>R1                      R2<br>(PRA Ratio) | PRA non-stenotic / PRA IVC<br>R2                      R3<br>(Suppression index) |
|------------------------------------------------------------------------------|---------------------------------------------------------------------------------|
| 3.9 / 1.3 > 1.5                                                              | 1.3 / 2.3 < 1                                                                   |



Fig. 3: Inflatable balloon catheter in right stenotic artery.



Fig. 4: Dilatation of right stenotic artery after PTR and collaterals.

## Discussion

FMD is a well known angiopathy primarily involving the aortic branch arteries. The disease was originally described in the renal arteries in 1938<sup>5</sup>. But, later, it was reported that it could also affect carotid, celiac, mesenteric, iliac, splenic, hepatic, subclavian, and even the coronary arteries<sup>5-7</sup>. Recently, involvement of the aorta has also been documented<sup>5</sup>. Involvement of the renal arteries is the most common manifestation of FMD. It has been reported that half of the patients with bilateral renovascular disease show additional extrarenal FMD<sup>5</sup>.

Although FMD may be seen at any age, and in both sexes, it is known as the disease of young women, and the frequency in children remains low. The ages of patients range from five years to 69 years in large group studies<sup>6,8,9</sup>.

Usually the middle and distal third of the main stem of the renal artery is involved, but extension into the branches of the renal artery can occur. The disease may be bilateral or unilateral, and nearly 80 percent of unilateral lesions occurs on the right side<sup>6-9</sup>. Our patient had a high grade stenosis in the dorsal branch of the right main renal artery. Collaterals can be demonstrated in 17 to 93 percent of lesions<sup>6,9</sup>. The second angiography performed on our patient revealed renal arteries with collaterals (Fig. 4).

The etiology of FMD is still unknown. Many hypotheses have been proposed stressing the importance of genetic and humoral factors and the role of arteriolar spasm<sup>5-7</sup>. Renal ptosis is common among patients with renal artery lesions: It has been postulated that traction due to ptotic kidneys (the right kidney larger than the left) may also be an etiologic factor.

Arteriographic findings of the lesions are pathognomonic for the diagnosis. FMD is classified as focal, multifocal, tubular and mixed, according to the angiographic findings<sup>6,8,10</sup>. The most common form is the multifocal type, which is also known to have a "string of beads" appearance<sup>2,6,8,10</sup>. The angiographic findings of our patient revealed that she had the focal type of FMD.

Histopathologically, these lesions can be classified as medial, intimal and periarterial, according to the arterial wall layer in which they predominate<sup>7,8</sup>. In some cases, the angiographic appearance of the lesions shows an excellent correlation with the pathological findings. It is known that medial fibroplasia represents the most common dysplastic lesion (nearly 85%) found in pediatric patients<sup>7</sup>.

FMD is considered to be a persistent and often progressive abnormality<sup>8,10</sup>. Unilateral FMD may progress to involve the contralateral renal artery<sup>7</sup>. Recently, it has been reported that spontaneous improvement may occasionally occur<sup>11,12</sup>.

Three different therapeutic approaches are being used in patients with renovascular disease: medical therapy, surgical intervention, and PTR. It has been reported that more favorable results of curative interventions, other than pharmacotherapy, have been obtained in unilateral renal artery stenosis with a significant PRA ratio and, in particular, in those with suppression of renin secretion in the contralateral kidney<sup>3,4</sup>. Our patient's PRA ratio and suppression index were 3 and 0.5 respectively, and these results were considered significant.

Renal angioplasty is a balloon dilatation technique and reconstructive procedure. Following its first description in 1978, PTR soon became the established technique for the treatment of almost all forms of renal artery stenosis in adults, proving to be both effective and safe, since there is no need for general anesthesia, surgery or prolonged hospitalization<sup>13,14</sup>. This technique also has certain applications in the treatment of renovascular hypertension in children, and satisfactory results have been reported following the development of smaller catheters and proper patient selection<sup>15-17</sup>.

PTR has been reported to be technically successful in 66-87 percent of patients with FMD. After successful dilatation, the overall benefit (cure and improvement of hypertension) has been found to be 94 percent<sup>14,15</sup>. The results of many studies document the good, long-term effect of PTR on blood pressure and kidney function in patients with renal artery stenosis.<sup>14,17,18</sup> The occurrence of restenosis has been reported to be 22 percent in patients with FMD<sup>15</sup>. The frequency of complications has been reported to range between 3-10 percent, and only 2-4 percent of major complications has been found to be directly related to PTR<sup>19</sup>. Finally, the reported mortality rates have been found to be much lower with PTR (1-3%) than with surgery (11%)<sup>5,19</sup>.

Our patient was treated medically with combinations of various antihypertensive drugs for a year, but her hypertension did not respond to the therapy. After PTR, her blood pressure returned to normal.

We suggest that PTR can be the treatment of choice in renovascular FMD in children because of its high cure and low complication rates and its long-term beneficial effect on renal functions.

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