

A Case with Cerebral Aneurysm in Renovascular Hypertension due to Fibromuscular Dysplasia

Mohammed E. Ghaidari, MD

Abstract

A case with renovascular hypertension due to fibromuscular dysplasia (FMD) was associated with cerebral aneurysm.

This patient was a twelve-year-old girl with cerebral aneurysm and complete obstruction of the right renal artery. The patient's cerebral aneurysm was successfully clipped with successful right side nephrectomy, and pathologic changes corresponded to FMD.

After nephrectomy, the patient's blood pressure became normal without need for medical therapy.

This report suggests that FMD should be considered to be a systemic angiopathy, including the cerebral artery as well as the renal artery (*Iranian Heart Journal 2004; 5(3):70-73*).

Key words: cerebral aneurysm ■ renovascular hypertension ■ fibromuscular dysplasia

Fibromuscular dysplasia (FMD) is an arterial disease of unknown etiology typically affecting the medium and large arteries of young to middle aged women. FMD first was described by Leadbetter and Burkland in 1938. FMD is an arterial disease typically affecting the renal arteries in 85% of patients and often presenting as renovascular hypertension. FMD has been associated with intracranial aneurysms in as many as 30% of patients. One third of patients with carotid FMD also have renal artery FMD. The reported incidence of FMD in adults is 0.5% via angiography and 1.1% via autopsy. Male to female ratio is 1:3 and most typically presents in persons aged 25-50 years. FMD has been associated with intracranial aneurysm and should be considered in patients who present with intracranial hemorrhage. Angiography is the criterion standard for diagnosing FMD.¹

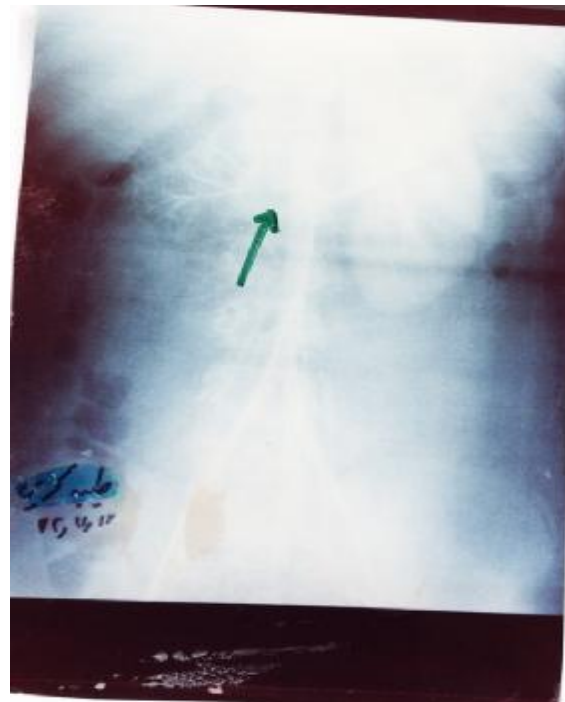
Fibromuscular dysplasia (FMD) of the renal artery is one of the leading causes of renovascular hypertension. FMD of the cerebral, internal carotid and vertebral arteries is clinically important as a cause of

cerebrovascular accidents in patients with renovascular hypertension due to FMD, which has not been stressed²; furthermore, successful cases of elective surgical intervention for cerebral FMD in renovascular hypertensive patients has been reported only in one report from Tokyo.² In this report, we describe one case of renovascular hypertension due to FMD with cerebral aneurysm that was successfully operated on for a cerebral aneurysm.

Case Report

A twelve-year-old woman was admitted to Rafsanjan Hospital in Iran. Her problem started 20 days before with headaches and vomiting. On the morning of her admission and following severe headaches and vomiting, she experienced tonic-clonic seizure and then a semi-comatose state. Her blood pressure was 220/140mmHg in the upper extremities. Her BP was controlled temporarily with IV sodium nitroprusside and then oral nifedipine, captopril, atenolol and triamterene-H, but still she had hypertensive crisis with a

severe headache and vomiting which was controlled with sublingual nifedipine. MRI was performed for assessing the intracranial events, and it showed intracranial hemorrhage. As a result, the patient was referred to our hospital in Tehran. At the time, she complained of continuous diplopia and intermittent headaches and vomiting associated with an increased blood pressure crisis. Prior to this, she had never had her blood pressure measured and had no family history of hypertension. Blood pressure was 130/90 in both upper extremities and 150/100 in both lower extremities; still, she sometimes had systolic blood pressure more than 200mmHg during admission. Her right eye had an internal deviation, but the pupil size and pupil reaction to light were normal. She had no carotid murmur or audible murmur over the abdominal region. In ophthalmoscopy, she had pupilledema in both eyes. On neurologic examination, she had positive Babinski sign; nonetheless, other findings of physical examination were normal. In the laboratory findings, CBC; ESR; FBS; urea; creatinine; uric acid; Na; K; Ca; P; thyroid and liver functional tests were normal. Antinuclear antibody, LE cell and RF were negative. Aldosterone level was normal, and plasma renin activity was high (she was under treatment with captopril). VMA level in 24 hr. urine was normal. In ECG, she had increased voltage. In abdominal ultrasonography, she had a small right kidney, and her left kidney was hypertrophied. Abdominal scan showed delayed excretion in the right kidney. In rapid sequence IVP, the right kidney size in the longitudinal axis was 10 cm; and in the left kidney, it was 13 cm with delay in excretion (Fig. 1). Kidney veins catheterization was done, revealing that renin level in kidney veins was high (she was under treatment with anti-hypertensive drugs, which could not be stopped).



A



B

Fig. 1. A, IVP depicting a small right kidney and hypertrophy of the left kidney. B, delayed excretion of the right kidney.

Radioisotope scans (DMSA and DTPA) with and without captopril, and VCUG with isotope were performed. In the

DMSA report, the right kidney was small and the left one was hypertrophic.

DTPA scan with and without captopril showed normal function with left kidney hypertrophy and low functional excretion with decreased perfusion and small size in the right kidney.

In VCUG, the urinary bladder was normal without tumoral lesions.

MRI was performed and suggested the possibility of middle cerebral artery aneurysm with bleeding in the left temporal lobe. Cerebral artery angiography revealed the presence of a saccular aneurysm in the middle cerebral artery (Fig 2).



Fig. 2. Cerebral angiography revealing a saccular aneurysm of the middle cerebral artery.

Kidney angiography was performed and revealed complete obstruction of the right main renal artery with collateral artery development. Left kidney arteries were normal.

The cerebral aneurysm was successfully clipped with successful right-sided nephrectomy. Pathologic findings in

different levels of kidney tissue and its veins and arteries showed:

- No lesion in the venous system
- Normal glomerular and kidney tissue
- Presence of organized and recanalized of thrombus in the renal artery
- Thickening of vessel walls in the affected region
- Proliferation and irregular interruption of the intima and inner elastic layer
- Severe thickening of the medial layer with collagen replacement in muscles.

These pathologic changes corresponded to FMD (Fig. 3).

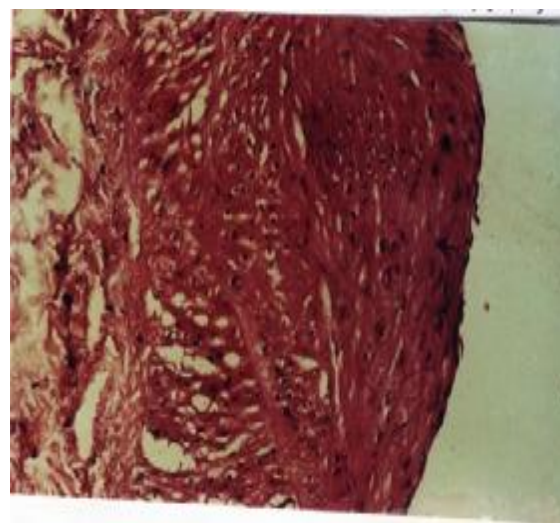


Fig. 3. Photomicrograph of renal pathology depicting typical features of FMD.

After nephrectomy, blood pressure became normal (without anti-hypertensive drugs).

In later follow-up, her blood pressure was normal and eye lesions and diplopia were gone.

Discussion

FMD is a disseminated vascular lesion affecting both vessels in the kidney and brain, and the same pathology in kidney vessels may be presented in cerebral

arteries. High blood pressure and weakness of the vascular wall in cerebral arteries might accelerate aneurysm formation in the cerebral arteries. This, with bleeding in cerebral parenchyma, accelerates increased intracranial pressure and blood pressure.

This case had a hypertensive crisis, accompanied with headaches and diaphoresis. She did not have orthostatic hypertension; nevertheless, in order to rule out pheochromocytoma, VMA in 24 hr urine collection was measured, but it was normal. Based on symmetric pulses and identical blood pressure in the extremities, coarctation of the aorta was ruled out.

IVP and abdominal scan findings revealed a small kidney with decreased functional excretion and this suggested renal artery stenosis, which was confirmed with angiogram.

This case had chronic hypertension (according to ophthalmoscopy findings and left ventricle hypertrophy). She presented with a hypertensive crisis that could not be controlled with medical therapy.

Angioplasty was impossible due to total obstruction, and renal artery surgery was also not possible owing to the obstruction and small size of the kidney; therefore, nephrectomy was required for blood pressure control, and her blood pressure became normal. After coarctation of the aorta, this is the second type of hypertension correctable with surgery.

Cases of FMD complicated with cerebral aneurysms have been reported by Houser et al. and Handa et al. Among the cases diagnosed as FMD of the renal artery or other arteries by angiography or by autopsy, cerebral aneurysmal

complications were noted in 19 of 37 cases (51%) by Mettinger et al.; in 9 of 19 (47%) by Wylie et al.; in 4 of 16 cases (25%) by Houser et al.; in 5 of 70 (7%) by Palubiskas et al.; and in 8 of 152 (5%) by Stanely et al. The possible reason for such a considerable variation in the incidence of cerebral aneurysm could be the fact that these studies were not prospective.

Only one successful case of elective surgical intervention for cerebral FMD in renovascular hypertensive patients was reported in 1989, and our case is the second case of successful surgical intervention for cerebral FMD in renovascular hypertensive patients.²

Conclusion

In light of the aforementioned observation, it is suggested that FMD be considered not as a disease localized in the renal artery but as a systemic angiopathy. Since the rupture of an aneurysm could result in a fatal event, cerebral angiography should be performed at least in those patients with multiple abdominal lesions of FMD so as to detect an association with cerebral aneurysm.

References

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