

Malignant Hypertension in a Child with Solitary Functioning Hydronephrotic Kidney

Seema Kapoor
Urmila Jhamb
Santosh K. Mittal
Satish K. Aggarwal*

A child with solitary functioning hydronephrotic kidney and hypertensive encephalopathy is described. Removal of the contralateral dysplastic kidney failed to normalize his blood pressure.

Keywords: *Hydronephrosis, Hypertension, Solitary kidney.*

Children with a solitary hydronephrotic kidney are uncommon. We report a patient with severe hypertension associated with a normally functioning solitary kidney, which showed features of hydronephrosis.

Case Report

An 8-year-old boy presented with history of headache for the past eight days and vomiting, fever and neck stiffness for the past one day. There was no preceding history of seizures, diaphoresis, palpitations or blurring of vision. The patient was conscious with minimal neck stiffness, left hemiparesis and blood pressure of 240/160 mm Hg. Fundus

examination showed grade 4 hypertensive changes with papilledema. The blood counts were normal and differential revealed lymphocytosis. Blood urea level was 36 mg/dL and creatinine was 0.8 mg/dL. CSF revealed no pleocytosis, normal proteins and no hypoglychorrachia. Contrast enhanced computed tomography showed minimal meningeal enhancement with no exudates and a small infarct in the region of right external capsule. This infarct was a probable sequale of long standing hypertension.

Hypertension was treated with a combination of nifedipine (2 mg/kg/day), propranolol (2 mg/kg/day), clonidine (20 µg/kg/day) and furosemide (2 mg/kg/day). His chest skiagram was normal and ECG showed evidence of left ventricular hypertrophy. Abdominal ultrasound showed absent left kidney with mild hydronephrosis of the right kidney; Doppler study showed no evidence of renal artery stenosis. An intravenous urogram confirmed adequate function of the right kidney. Plasma renin estimation (4.8 ng/mL/hr) was within normal limits. Selective renal angiographic study with estimation of renal vein renin (4 ng/mL/hr) was essentially normal. A DTPA-renal scan showed normal renal function on the right side with no function on the left. Diuretic renography done simultaneously, confirmed the presence of nonobstructive hydronephrosis. Voiding cystourethrogram done did not show evidence of vesicouretral reflux.

DMSA scan done subsequently had shown no evidence of scarring consequent to a preexisting reflux. Twenty four hour urinary catecholamines and I¹³¹ MIBG (metiodobenzyl guanidine) scintigraphy were normal. CT of the abdomen failed to demonstrate the contralateral kidney. Cystoscopy done revealed ureteric orifices on both sides. On laprotomy a dysplastic atrophic kidney (*Fig. 1*)

*From the Departments of Pediatrics and *Pediatric Surgery, Maulana Azad Medical College and Lok Nayak Hospital, New Delhi 110 002, India.*

Correspondence to: Dr. Urmila Jhamb, Department of Pediatrics, Lok Nayak Hospital, New Delhi 110 002.

E-mail: ujhamb@hotmail.com

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was identified on the left side, which was removed. This however failed to ameliorate the hypertension. Immunoturbidometric examination of the urine showed microalbuminuria (24 mg/g creatinine).

Discussion

Severe hypertension posed a diagnostic dilemma. Ultrasound, Doppler study plasma renin levels, urinary catecholamines and MIBG scan ruled out renovascular hypertension and pheochromocytoma. Imaging modalities suggested a solitary functioning right kidney with agenesis of the left kidney. CT failed to pick up any renal tissue on the left side. Diuretic renogram ruled out pelvi-ureteric obstruction causing right hydronephrosis. Presumably, volume overload on the solitary functioning kidney resulted in

hydronephrosis. Vesico-ureteric reflux was ruled out on imaging studies. Therefore, the right kidney did not have a "surgically treatable" cause of hypertension. This left us with one important question - was the left kidney really absent or present but dysplastic? A faint suggestion came up on the angiography that showed a small beaking from the left side of the aorta at the usual location of the ostium of the left renal artery. Cystoscopy contributed by visualizing the left ureteric orifice. This settled the issue of agenesis *verses* dysplasia. On surgical exploration the lower part of the ureter was identified and followed upwards leading to the dysplastic kidney. Looking at the specimen grossly being just 1.5 cm and disorganized, it is not surprising that CT and ultrasound could not pick it up pre-operatively. The dysplastic left kidney was removed because hypertension, infection, hemorrhage and malignant change are known complications of renal dysplasia(1). There have been reports of improvement in hypertension following nephrectomy of the dysplastic kidney. The chances of complete regression of hypertension after nephrectomy are low, the possibility of reduction in the doses of antihypertensive drugs is a definitive goal.

Bachmann has presented case reports wherein blood pressure normalized after removal of the small kidney and in few where hypertension persisted(2). He emphasized that nephrectomy of the small kidney should only be performed, if the integrity of the contralateral kidney is certain.

Hypertension in patients with solitary kidney have appeared in literature. Lan, *et al.* collected data on 14 children with unilateral renal agenesis defined by cystoscopy of which 21% had hydronephrosis. They observed that this group had increased prevalence of hypertension, proteinuria and renal insufficiency to the tune of 29%, 43% and 36%, compared to

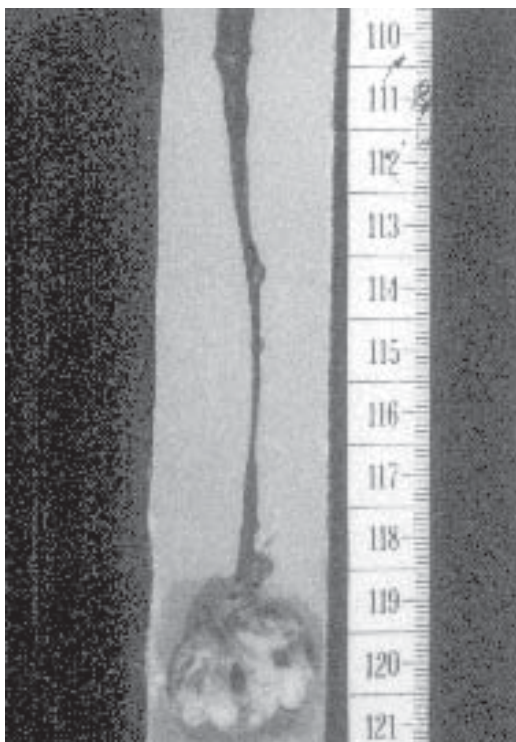


Fig. 1. Grossly atrophic kidney removed on laprotomy.

the uninephrectomized group. In their study the incidence of the congenital solitary kidney (CSK) was 1 in 1,496. They observed that these complications were common in the age group of 30 to 60 years, unlike our patient who presented in the first decade(3).

The cause of hypertension in a child with solitary kidney has been extrapolated from animal studies and later confirmed in humans. Laboratory studies show that a marked reduction (11/12) in renal mass results in progressive glomerulosclerosis of the residual tissue, manifested by azotemia, proteinuria and hypertension. Glomerular destruction appears to be due to augmented trans capillary hydraulic pressure difference (DP) and glomerular plasma flow, a process of chronic hyperfiltration that leads to altered charge and size selective permeability, increased trans capillary protein flux, mesangial overloading and sclerosis(4). CSK is an experiment of nature, in which the stimulus exists in fetal life, the remnant kidney being enlarged. The first suggestion that the solitary kidney might be prone to focal glomerulosclerosis (FGS) was provided by Kiproff, *et al.*(5). A forerunner of FSGS is microalbuminuria. The renal function may however remain stable for a long time.

It is also important to follow up these children, prevent protein overloading and retard the development of FGS(6). The incidence of finding a grossly atrophic kidney in a child is as high as 1 in 1400, therefore it is

important that such children are identified and followed up stringently.

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REFERENCES

1. Husmann DA. Renal dysplasia: the risks and consequences of leaving dysplastic tissue in situ. *Urology* 1998; 52: 533-536.
2. Bachmann H. Compensatory hypertrophy of the healthy kidney as an important prerequisite for the nephrectomy of the small kidney. *Urologe A* 1985; 24: 80-83.
3. RR Lan, JJ Huang, JM Sung, SN Lid. Congenital solitary kidney: Review of 14 cases and their outcome on comparison with uninephrectomized patients. *Acta Nephrol* 1996; 10: 26-31.
4. Riehle RA Jr, Vaughan RD Jr. Renin participation in hypertension associated with unilateral hydronephrosis. *J Urol* 1981; 126: 243-246.
5. Kiproff DD, Colvin RB, McCluskey RT. Focal and segmental glomerulosclerosis and proteinuria associated with unilateral renal agenesis. *Lab Invest* 1982; 46: 275-281.
6. Brenner BM. Hemodynamically mediated glomerular injury and the progressive nature of kidney disease. *Kidney Int* 1983; 23: 647-655.