Renal lymphangioma: a cause of neonatal nephromegaly

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Abstract. A newborn male presented with bilateral nephromegaly and mild hypertension. Function of the right kidney was reduced on excretory urography. Ultrasound showed bilaterally enlarged kidneys with increased echogenicity and poorly defined corticomedullary junctions. Areas of decreased medullary enhancement were seen on CT. Renal biopsy demonstrated lymphangioma, probably arising from the peripelvic renal tissues.

Bilateral nephromegaly in the newborn infant is usually caused by hydronephrosis or cystic renal disease. Rarely, neoplasia and infiltrative disorders such as leukemia and glycogen storage are implicated.

We recently observed a hypertensive neonate with bilateral nephromegaly in whom urographic, ultrasound and computed tomographic images of the kidneys did not conform to previously described causes of renal enlargement in this age group. The diagnosis of lymphangioma was eventually made by open renal biopsy. This entity has not been previously described in a neonate.

Case report

W.U. was a 3.08-kg white male born to a 39-year-old mother following a 36-week uncomplicated pregnancy and a normal delivery. Prior to delivery, ultrasonography and routine amniocentesis were normal. On initial physical examination, a 3-cm firm mass was palpated in the right upper abdomen. The infant had mild diastolic hypertension with a blood pressure of 80/70. The physical examination was otherwise normal and no peripheral edema or lymphatic abnormalities were identified. Initial urinalysis revealed a specific gravity of 1.019, 4+ protein, 2+ blood with 50–70 RBC/hpf and numerous granular casts and multiple subsequent urinalyses demonstrated continued proteinuria and hematuria. BUN was 8 mg%, creatinine 0.2 at age 2 days. Initial electrolytes were Na 134, K 3.8, chloride 98, CO₂ 12.

At 2 days of age, renal ultrasonography revealed bilaterally enlarged kidneys. The right measured 7 cm in length and the left 6 cm. There was increased echogenicity throughout both kidneys, with absence of normal cortico-medullary differentiation. The normal strong central echo complex [1] could not be identified on either side (Fig. 1).

An excretory urogram performed at 5 days of age revealed a normal left renal collecting system. The renal outlines were not visible and the right kidney did not opacify even on delayed films (Fig. 2).

A contrast-enhanced abdominal CT scan was performed at 6 days of age. The scan demonstrated bilaterally enlarged kidneys, more marked on the right, without evidence of hydronephrosis. There was marked bilateral enhancement of well-preserved renal...
Fig. 2. 6-min film from excretory urogram. The renal outlines are not visible. The left collecting system is somewhat attenuated but otherwise normal. The right kidney is not opacified.

Fig. 3. Enhanced CT scan. There is bilateral renal enlargement, the right more marked than the left. The renal cortices are enhanced but the medullary portions are poorly opacified. The collecting systems demonstrate reduced opacification, more marked on the right.

cortices. The medullary portions were less well opacified and showed areas of decreased attenuation within which irregular enhanced areas appeared to represent the columns of Bertin. Opacification of the renal collecting systems was also markedly reduced (Fig. 3).

To rule out intrarenal tumors, the patient underwent bilateral open renal biopsies at 8 days of age. At operation, there was generalized renal enlargement, more severe on the right, but the kidneys were otherwise grossly normal with fetal lobulation and no evidence of tumor masses. Nevertheless, multiple biopsies were obtained and frozen sections suggested a vascular anomaly. Permanent histologic sections revealed a lymphangioma located in

the renal peripelvic tissues. The tumor consisted of dilated lymphatic vessels which partially infiltrated the renal medullary tissues (Fig. 4). The renal cortex showed a poorly developed nephron mass, scattered subcapsular glomerular cysts and thickened arterioles (Fig. 5). The postoperative course was uneventful and the patient was discharged on antihypertensive medication. He was followed clinically and with intermittent renal ultrasound and CT. At the age of 13 months, the CT scan revealed the right kidney to be normal in size with normal opacification of the collecting system. The left kidney was unchanged in size but only a few areas of low attenuation remained. The renal ultrasound demonstrated a decrease in size of the right kidney. The echogenicity of both kidneys remained increased.

Discussion

While clinically and urographically this infant appeared to have unilateral renal enlargement, this impression was corrected by sonographic and CT examinations which revealed bilateral nephromegaly. This finding tended to exclude lesions which are usually unilateral such as congenital mesoblastic