Renal Segmental Hypoplasia, Ask-Upmark Kidney, in a Patient with Adult-onset Hypertension

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A 39-year-old Japanese woman was referred to our hospital because of a 3-year history of hypertension with a small right kidney identified by abdominal ultrasonography. Her blood pressure had remained normal during her first and second pregnancies, at age 24 and 26. Gestational hypertension had been noted during her third pregnancy, at age 30, but her blood pressure values had returned to normal values after delivery. Elevated blood pressure, around 140/90 mmHg, had been noted for the first time at age 36 at a regular medical examination. At age 39, blood pressure was elevated at 180/100 mmHg and renal dysfunction had been identified at a local hospital. On admission to our hospital, her blood pressure was 148/90 mmHg and physical examination was not remarkable except for a slight hypertensive change of the retina. Laboratory examination revealed mild renal impairment: serum creatinine was 1.59 mg/dl without proteinuria or any abnormalities on urinalysis. Contrast-enhanced abdominal computed tomography (CT) revealed a markedly small right kidney (7.5×4.0 cm), segmental cortical thinning in the upper part of the left kidney, and no urinary tract abnormalities (Fig. 1A). The renal scintigraphy and renography revealed markedly reduced accumulation or perfusion of technetium-99m-diethylenetriaminepentaacetic acid in the right kidney. Plasma renin activities before and at 60 minutes after captopril administration (25 mg) were respectively 2.1 and 6.5 ng/ml/hr; the increase was not significant (1). Moreover, renal arteriography revealed uniformly small renal arteries from the origin of the aorta in the right kidney (Fig. 1B), narrowed renal arteries with tiny renal arterial branches in the upper part of the left kidney (Fig. 1C), but no renal artery stenosis in either kidney. In the nephrographic phase, the normal parenchymal tissue remained segmentally only in the left kidney (Fig. 1D). The patient did not present any clinical or laboratory evidence of vasculitis, atherosclerosis, collagen diseases, or anti-phospholipid syndrome, and she had no history of frequent urinary tract infections. Because of her severe bilateral renal lesions and because her blood pressure was controlled at around 120/70 mmHg with mild sodium restriction (plasma renin activity, 1.1 ng/ml/hr), we did not perform any surgical procedures or pathological examination, and we discharged her.

For this patient with bilaterally asymmetrical renal anomaly with adult-onset hypertension and renal dysfunction, the radiological examination of the left kidney (i.e., contrast-enhanced CT and renal arteriography) revealed that the normal renal parenchymal tissue was separated from an atrophic lesion containing a thin cortex overlying dilated calices, characteristics consistent with segmental renal hypoplasia, i.e., Ask-Upmark kidney (2-4). This renal anomaly is frequently unilateral, but bilaterally asymmetrical segmental hypoplasia has also been reported (4). In the present case, the right kidney showed the diffuse anomaly; thus there is a possibility that the right kidney lesions have might have been due to other renal anomalies, such as unilateral renal hypoplasia or renal dysplasia (5). Ask-Upmark kidney has come to be regarded as a consequence of vesicoureteral reflux rather than a true congenital malformation (2). Although our patient showed no evidence of vesicoureteral reflux in adulthood, the possibility of intrarenal reflux in utero or in infancy can not be excluded.

Clinically, Ask-Upmark kidney usually presents as severe hypertension in pediatric or adolescent patients, and abnormal renin secretion is proposed as the cause of the hypertension (1-3). However, nephrectomy has been shown to normalize blood pressure regardless of plasma renin activities (2); thus, the role of renin in the pathogenesis of hypertension remains uncertain. As the plasma renin activities were not increased significantly with captopril and sodium restriction, sodium retention due to renal dysfunction might have been the main cause of the hypertension in our case; thus sodium restriction alone was sufficient to ameliorate her hypertension. This study indicates that a renal anomaly as seen in the present patient should be added to the list of diseases potentially associated with hypertension and renal dysfunction, even in adults.

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Figure 1. Contrast-enhanced abdominal computed tomography reveals a markedly small right kidney (7.5×4.0 cm) and segmental cortical thinning in the upper part of the left kidney (A). Renal arteriography discloses the arteries with a uniformly small caliber in the right kidney (B) and the upper part of the left kidney (C), but no renal arterial stenosis. Selective angiography shows the minimal cortex in the middle portion of the left kidney (D).

References


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