Crossed Renal Ectopy Associated with Tuberous Sclerosis

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Crossed renal ectopy is a quite uncommon condition with an autopsy incidence of about 1 in 2000 and a slight male predominence (1,2). Tuberous sclerosis occurs 1 in every 150.000 births (3), and it is described generally as a part of a triad of epilepsy, mental retardation and adenoma sebaceum. Angiomyolipomas (hamartoma) occur in 40 % to 80 % of patients (3), and they can be identified in the kidneys, cerebrum or other organs by sonography and computerized tomography. Renal cysts may occur in about 43 % of patients, and renal cell carcinoma can occur in these cysts afterwards.

We present a case in which crossed renal ectopy without fusion was together with tuberous sclerosis.

Case report

R.B., a 6 years old girl was admitted to the hospital due to recurrent abdominal pain. She had a history of epileptic state beginning from the 2 nd month of life. Her family reported retardation in mental functions.

Intravenous pyelography, ultrasonography (US) and computerized tomography (CT) did not show the left kidney in the normal location. A cystic lesion of 4x3 cm diameters in the right iliac fossa was detected both by US and CT (Figure 1). Cystoscopic examination revealed a left ureterocele. Cranial CT scanning also demonstrated a cortical hypodense area (hamartoma?) in the supraventricular region of the right parietal cerebral lobe and subependimal calcified nodules in the lateral ventricules (Figure 2). In the surgical exploration, we found that the left kidney was localized in the right iliac fossa, and the right ureter ran over it (Figure 3). Total nephroureterectomy was performed after clamping its pedicule consisting of abnormal vessels.

Histopathologic examination showed dysplastic cystic kidney.

Discussion

Many patients with crossed renal ectopy remain entirely asymptomatic. However, because of the associated malrotation, a significant proportion of these kidneys display pelvicaliceal dysmorphism that simulates hydronephrosis. Also, reflux is commonly observed to be associated with these anomalies (4). In our case, there was only abdominal pain. Additionally, radiographic examinations revealed that the kidneys were hydronephrotic due to the stenotic ureteral orifice and ureterocele.

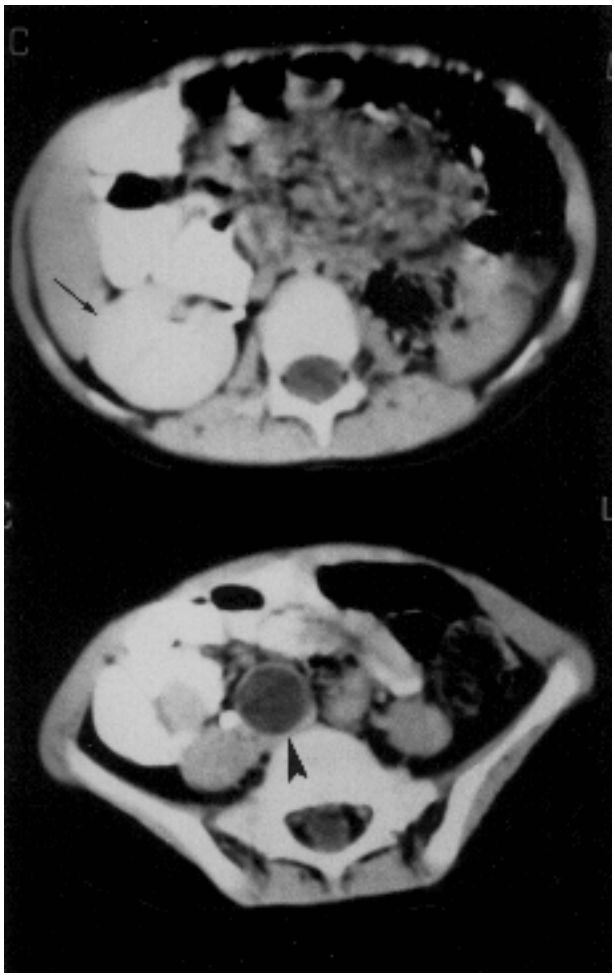

Figure 1. CT appearance of the crossed renal ectopia: Upper; right kidney (**arrow**), absence of the left kidney. Lower; cystic kidney in the right side (**arrow head**) .

Tuberous sclerosis may present as an autosomal dominant disorder (in 15 to 25 per cent of cases) which is characterized by the presence of adenoma sebaceum, epilepsy, and mental retardation. These findings may appear at different times and in different sequences in each patient. Thus, the diagnosis of the disease may be delayed until the initial symptoms emerge. Our examinations revealed that her father, mother and four brothers were healthy, and the patient had no adenoma sebaceum. That is, this case is either sporadic or an example of the genetic condition with variable or incomplete penetrance.

Neither hamartoma nor cyst was determined in the kid-

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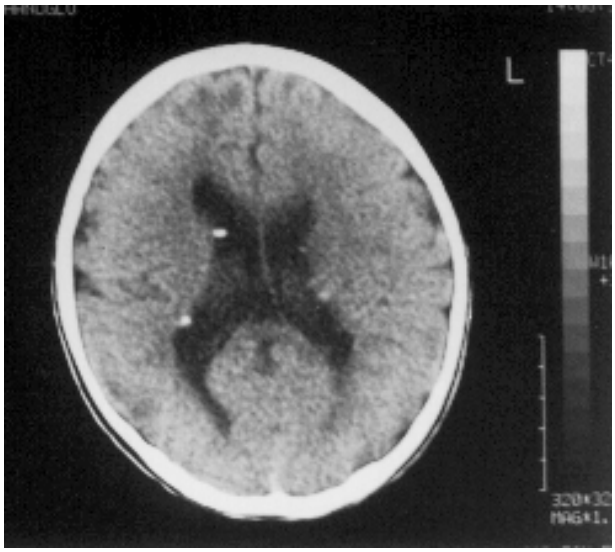

Figure 2. CT appearance of the subependimal calcified nodules.

neys. However, cystic dysplasia was the histopathological diagnosis of the ectopic kidney. This finding is not in agreement with the related literature, for hamartomas or cysts of the kidneys and other organs were reported in patients with tuberous sclerosis (3). Therefore, association of crossed renal ectopy, together with dysplastic cystic immaturation and tuberous sclerosis is very interesting, and according to our knowledge such association has not been reported before.

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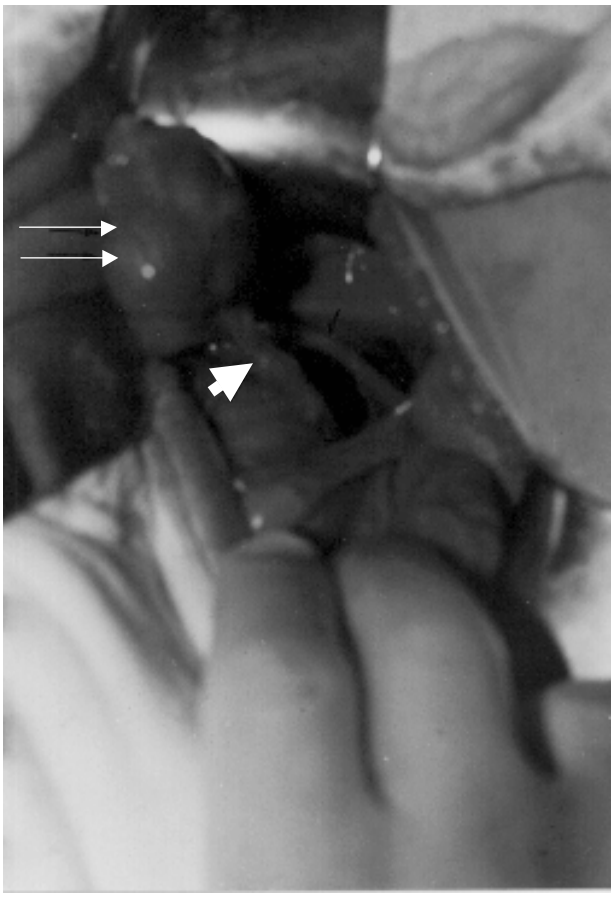

Figure 3. Appearance of the left cystic kidney in the right iliac fossa: non-functional cystic kidney after dissection (arrows), its dilated ureter (arrow head), ureter of the right kidney (small arrow).

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